La richiesta di competenza neurologica nel prossimo futuro

Malattie della sostanza bianca: diagnosi neuroradiologica e contributo alla diagnosi differenziale

Roberto Floris

22-24 ottobre 2021

Sin SOCIETÁ ITALIANA DI NEUROLOGIA

FONDAZIONE SOCIETÀ ITALIANA DI NEUROLOGIA Sin

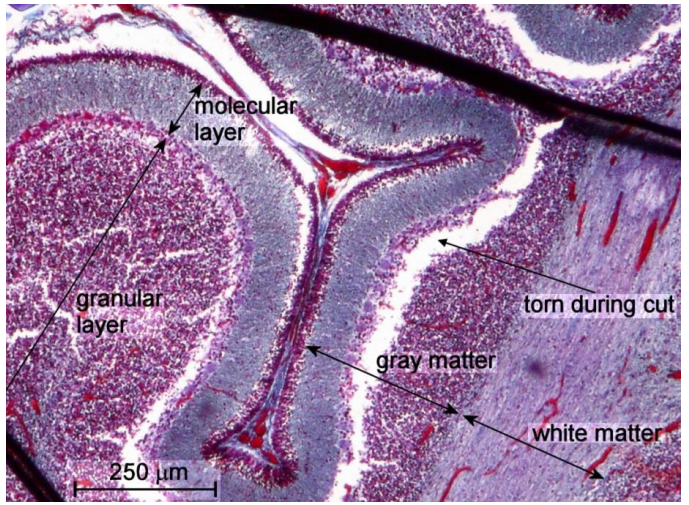
Sign SEZIONE ITALIANA GIOVANI NEUROLOGI

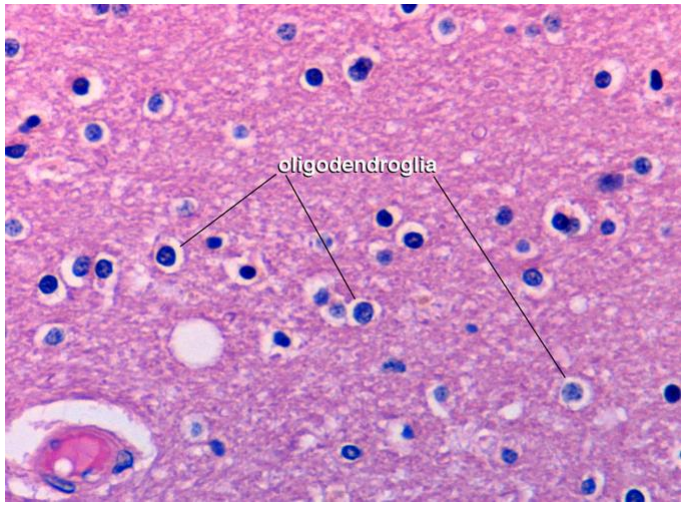
Sostanza bianca

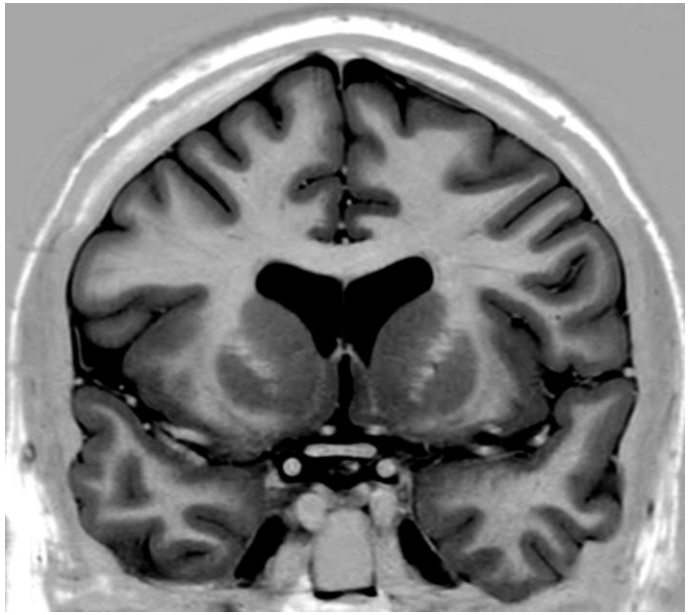
- Costituita da **assoni mielinizzati** e **cellule gliali**
- Mielina prodotta da **oligodendrociti** è responsabile del colore e delle caratteristiche RM
- Mielina composta da 40% di acqua, lipidi (cerebrosidi, lecitina) e proteine (proteolipidi, **proteine basiche della mielina*** e **glicoproteina mielinica degli oligodendrociti***)

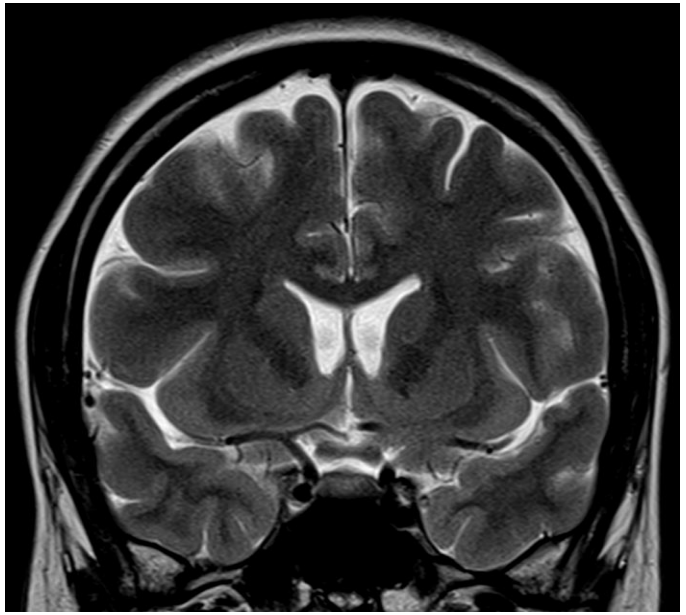
* Localizzate nello strato più esterno della mielina ed è un potenziale target di autoimmunità

Colorazione con "Luxol Fast Blue"

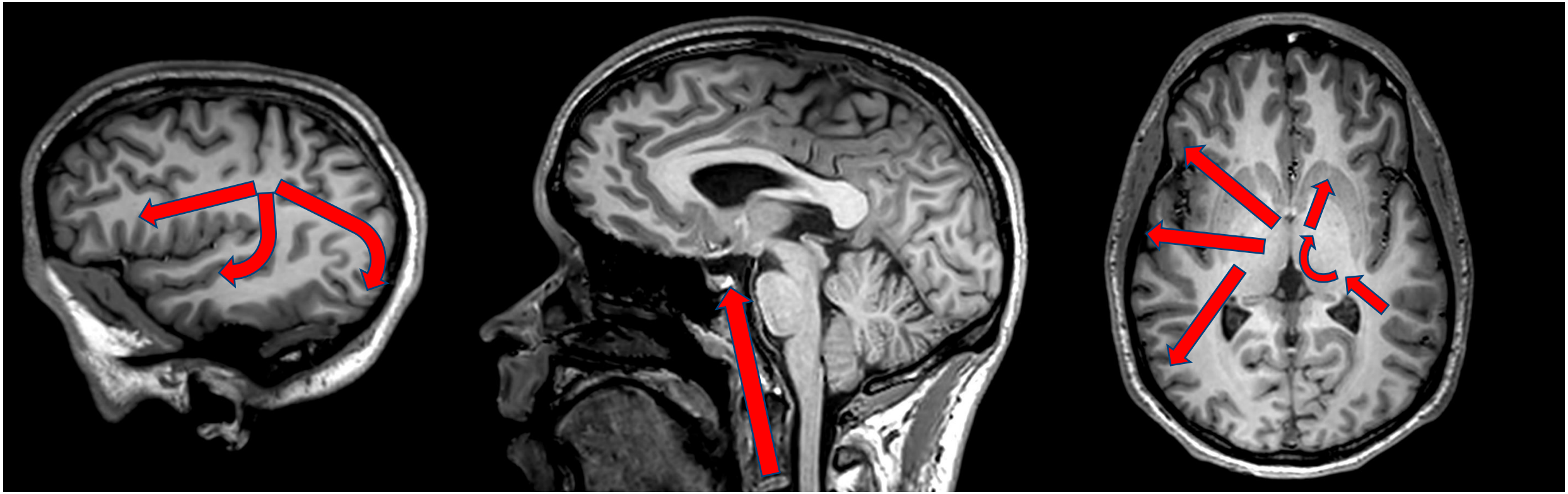








Mielinizzazione



Mielinizzazione delle regioni sensitive avviene prima delle regioni motorie

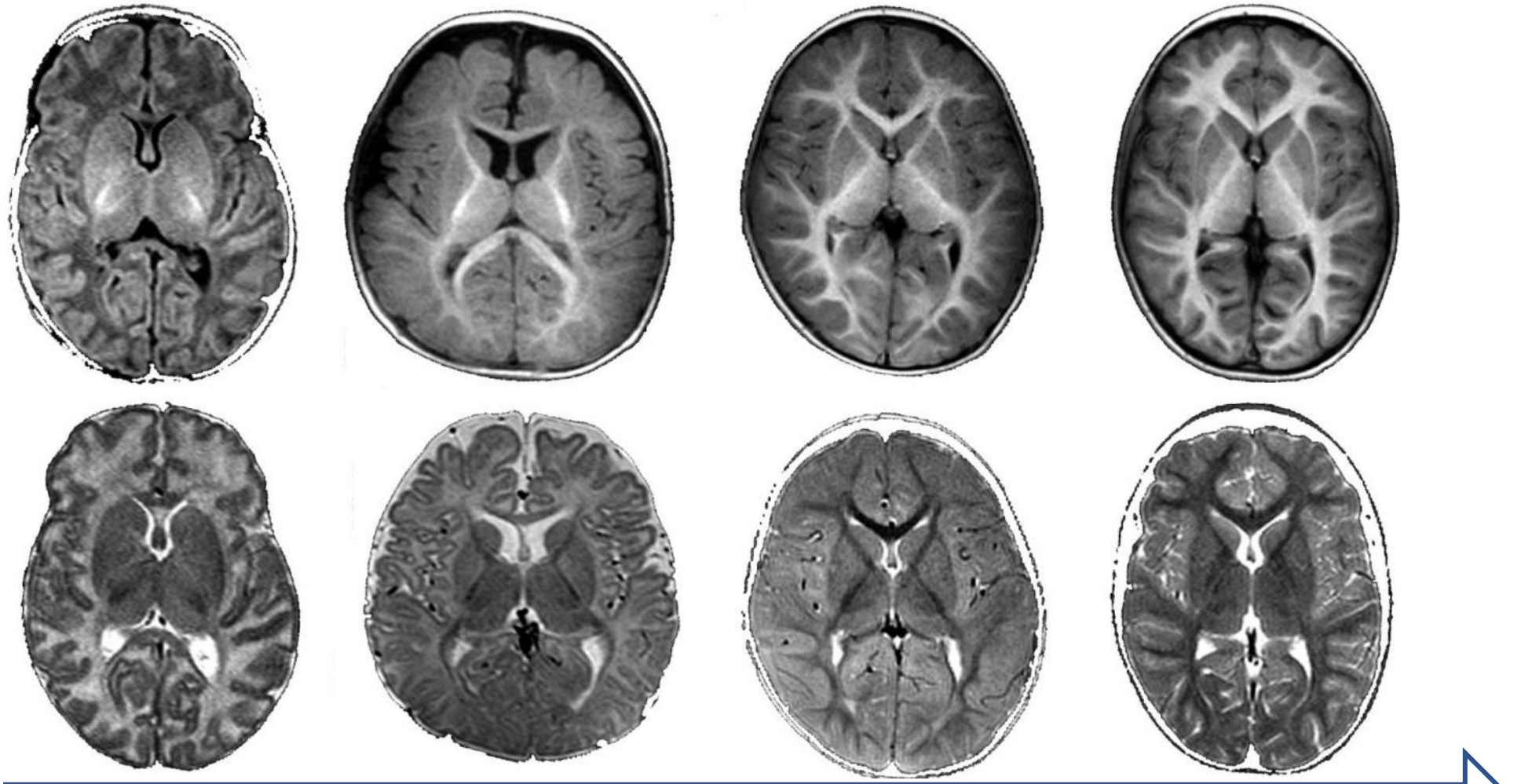
Mielinizzazione dalla sostanza bianca centrale alla periferia

La mielinizzazione delle regioni posteriori antecede le regioni temporali e frontali

Mielinizzazione

T1

T2



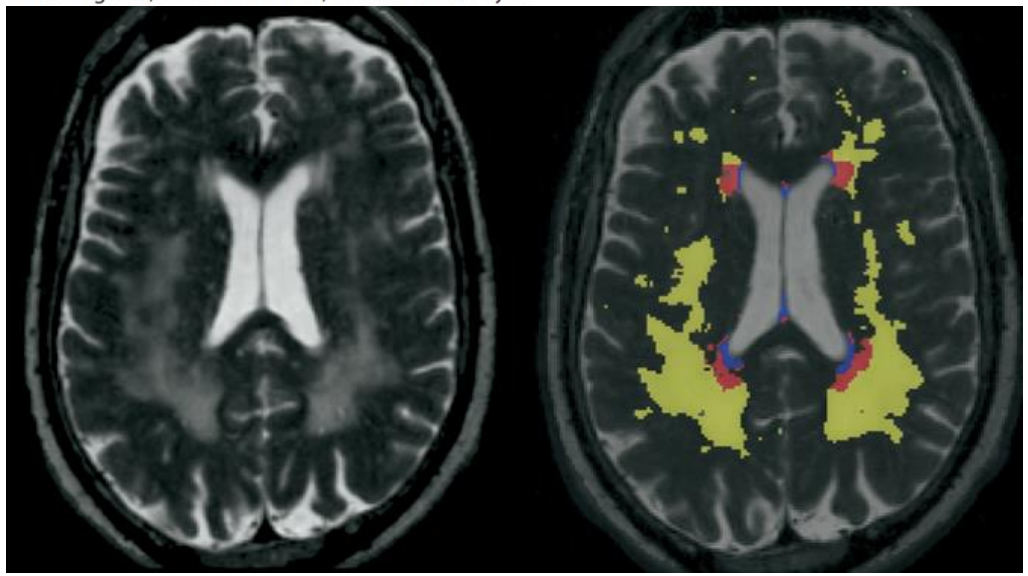
Processo realizzato da **Oligodendrociti** nel sistema nervoso centrale dal **5° mese** di vita sino ai **2 anni** di vita

Classificazione topografica sostanza bianca

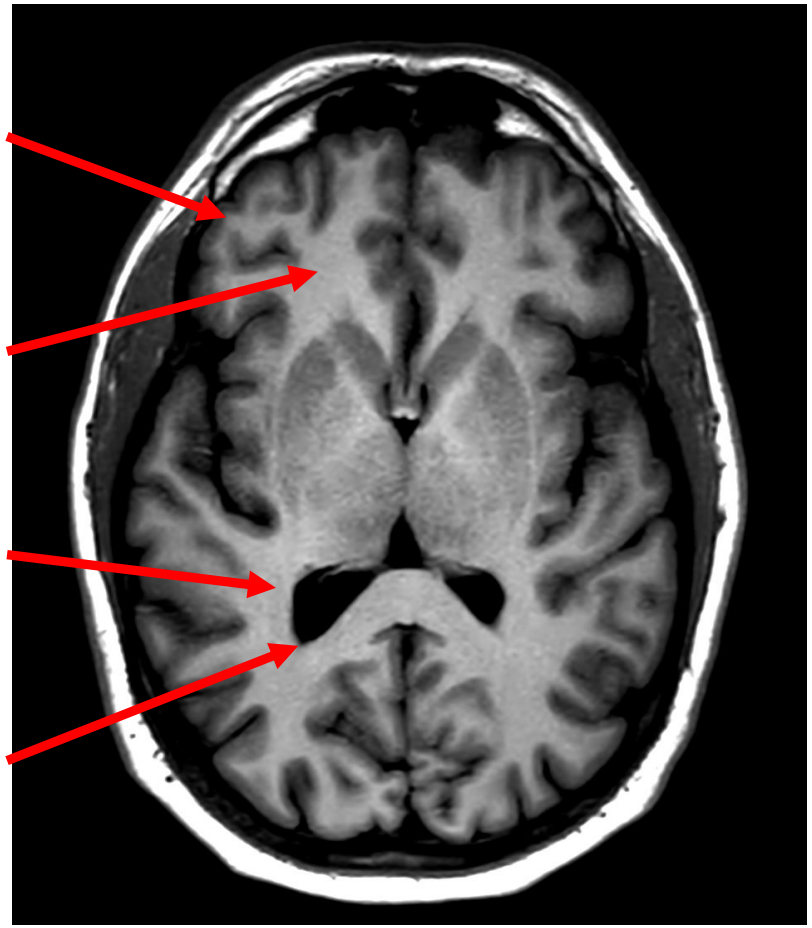
REVIEW BIOL PSYCHIATRY 2008;64:273–280

Classification of White Matter Lesions on Magnetic Resonance Imaging in Elderly Persons

Ki Woong Kim, James R. MacFall, and Martha E. Payne



Sostanza bianca iuxtaventricolare
Sostanza bianca periventricolare
Sostanza bianca profonda
Sostanza bianca iuxtacorticale



Sostanza bianca iuxtacorticale

Sostanza bianca profonda

Sostanza bianca periventricolare

Sostanza bianca iuxtaventricolare

Table 1. Proposed Subclassification of WM Lesions

	Juxtaventricular	Periventricular	Deep	Juxtacortical
Locations	Within 3 mm from ventricular surface	Periventricular watershed zone 3–13 mm from ventricular surface	Between periventricular WM and juxtacortical WM	Within 4 mm from corticomedullary junction

Malattie sostanza bianca

Comprendono disordini che colpiscono esclusivamente o prevalentemente la sostanza bianca encefalica

Genetiche o **acquisite**

Progressive o **statiche**

Possono insorgere ovvero manifestarsi in qualsiasi età

DEMIELINIZZANTI	**DISMIELINIZZANTI**	**IPOMIELINIZZANTI**
Anomalie secondarie a distruzione di mielina	*Anomalie primarie della mielinizzazione*	*Difettiva mielinizzazione*

Malattie demielinizzanti

Malattie demielinizzanti

Malattie autoimmuni

- Sclerosi multipla e varianti (Balo, Marburg, Schilder)
- Neuromielite ottica (m. di Devic)
- Encefalomielite acuta disseminata (ADEM)
- Leucoencefalopatia acuta emorragica (m. di Hurst)

Malattie infettive

- Leucoencefalopatia multifocale progressiva (PML)
- Leucoencefalopatia HIV correlata

Malattie vascolari

- Arteriolosclerosi
- Angiopatia amiloide cerebrale
- CADASIL
- Sindrome di Susac
- Neurolupus

Malattie tossico-metaboliche

- Mielinolisi pontina
- Leucoencefalopatia da Metotrexate
- Sindrome dell'encefalopatia posteriore reversibile
- Patologie alcol-relate

Sclerosi Multipla

The NEW ENGLAND JOURNAL of MEDICINE
2018

REVIEW ARTICLE

Dan L. Longo, M.D., *Editor*

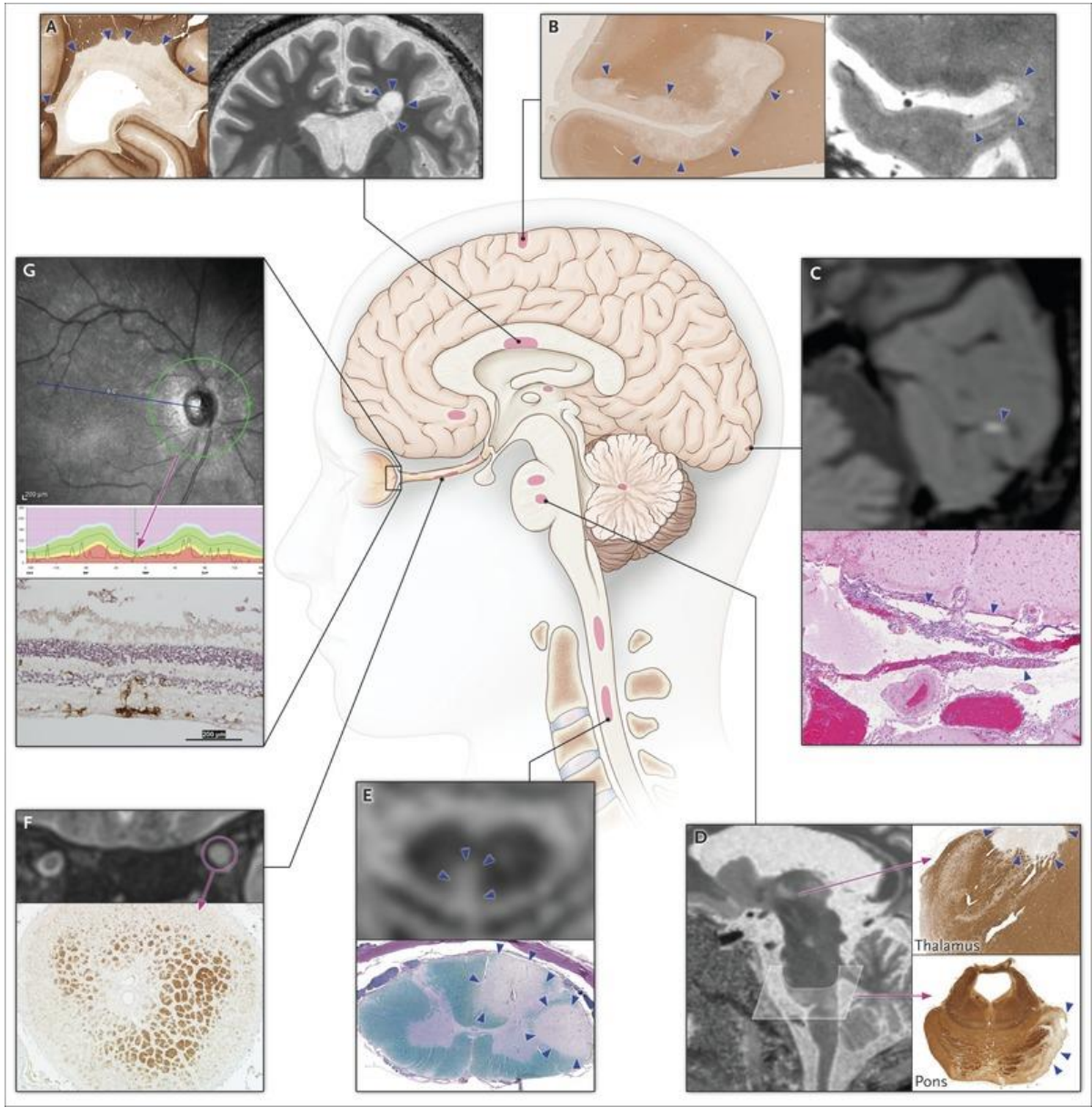
Multiple Sclerosis

Daniel S. Reich, M.D., Ph.D., Claudia F. Lucchinetti, M.D.,
and Peter A. Calabresi, M.D.

- Malattia demielinizzante primitiva ad etiologia sconosciuta

- Istologia:
  - -infiammazione perivenulare
  - -demielinizzazione con risparmio assonale
  - -infiltrazione macrofagica e linfocitaria perivenulare
  - -attivazione microglia e deposito di fibrina
  - -possibili depositi emosiderinici

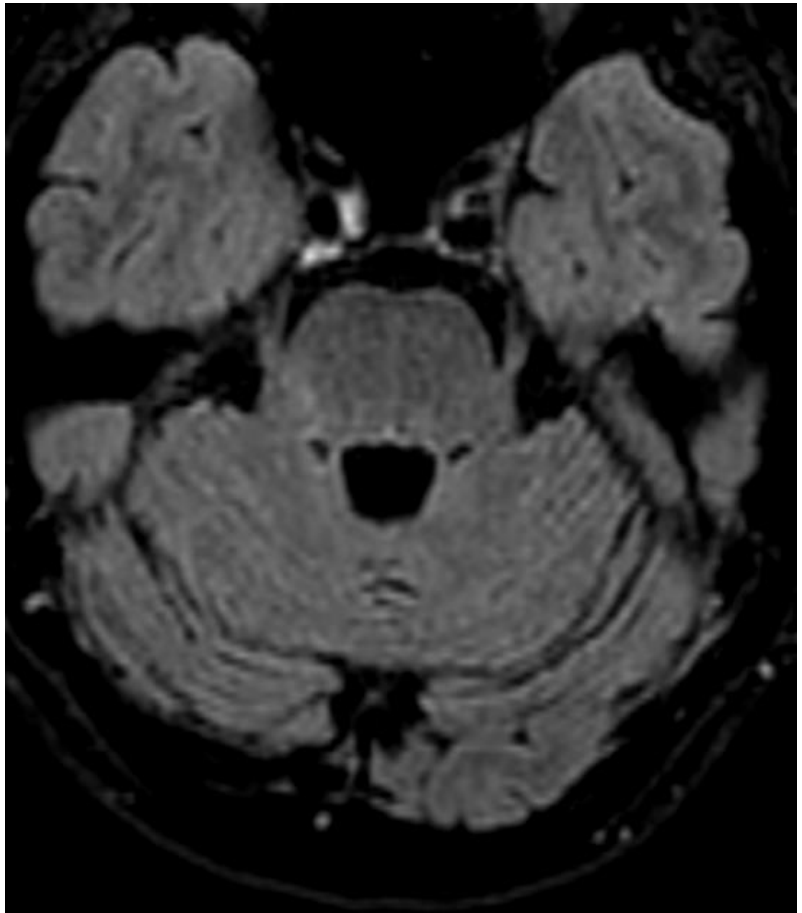
- Imaging: lesioni della sostanza bianca periventricolare, iuxtacorticale, infratentoriale, corpo calloso e del midollo spinale



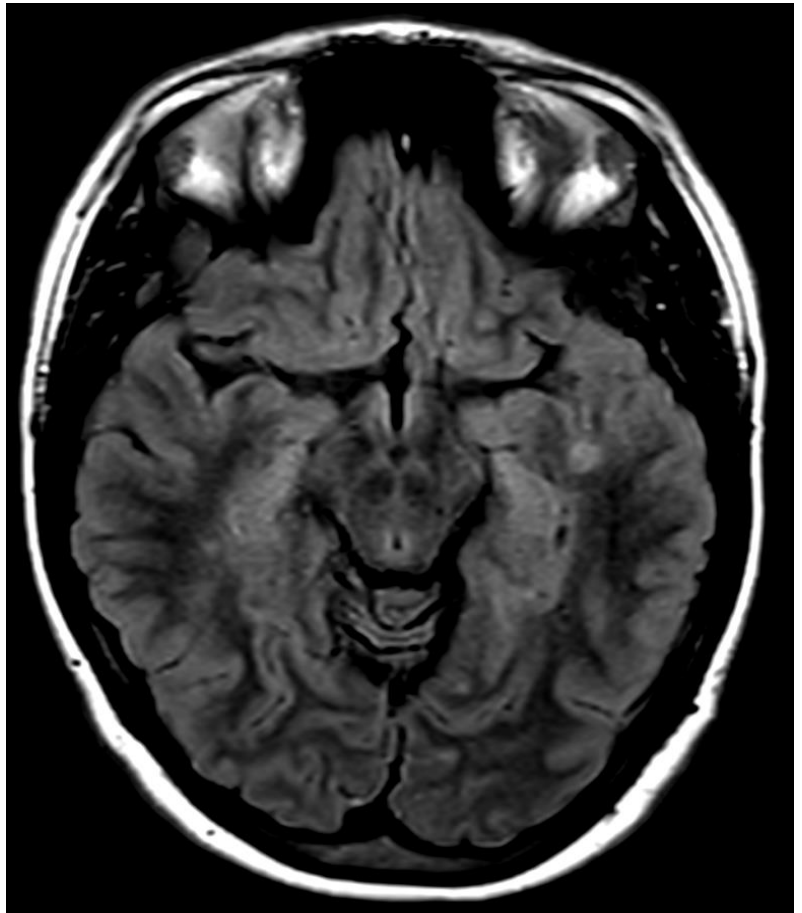
Sclerosi Multipla

Aspetti tipici

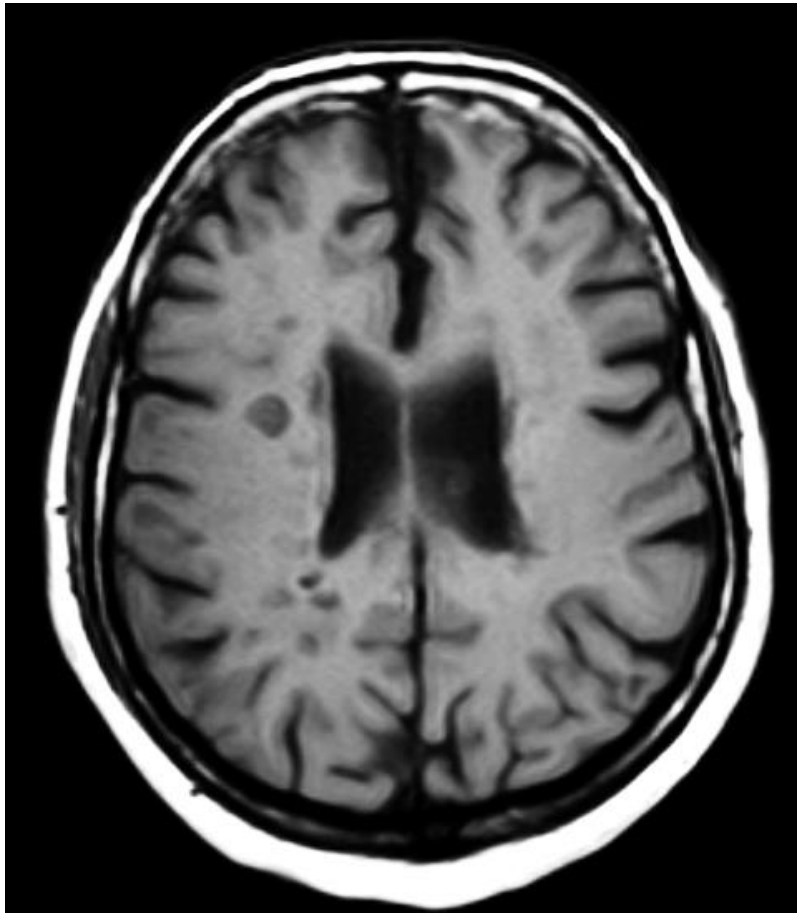
In sede **truncale lesioni**
spesso **periferiche**



Lesioni **periventricolari**
adiacenti i corni **temporali**



Black holes
Volume correla con scale cliniche

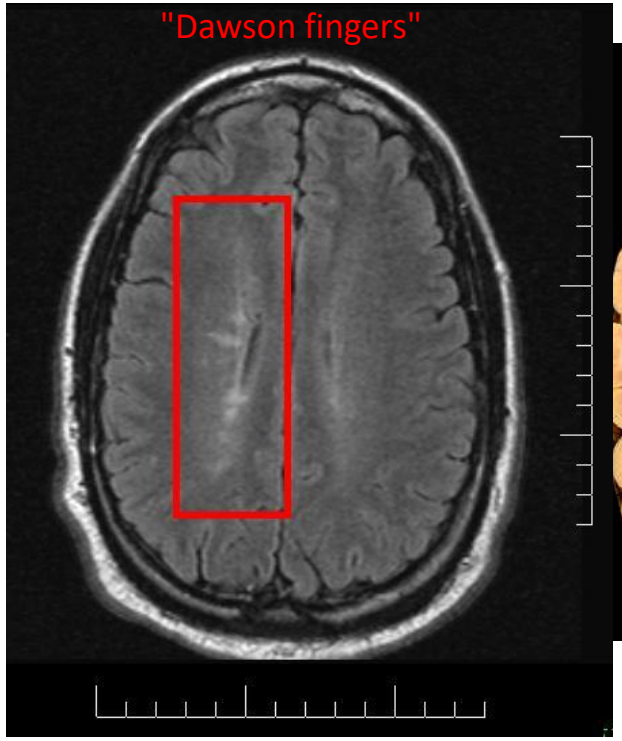


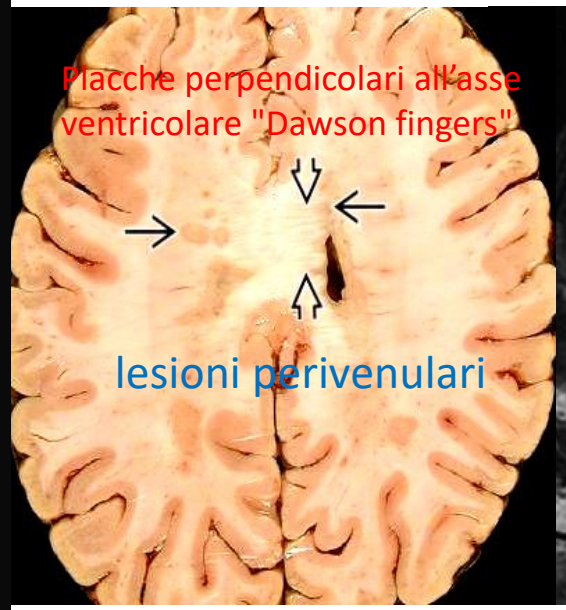
Sclerosi Multipla

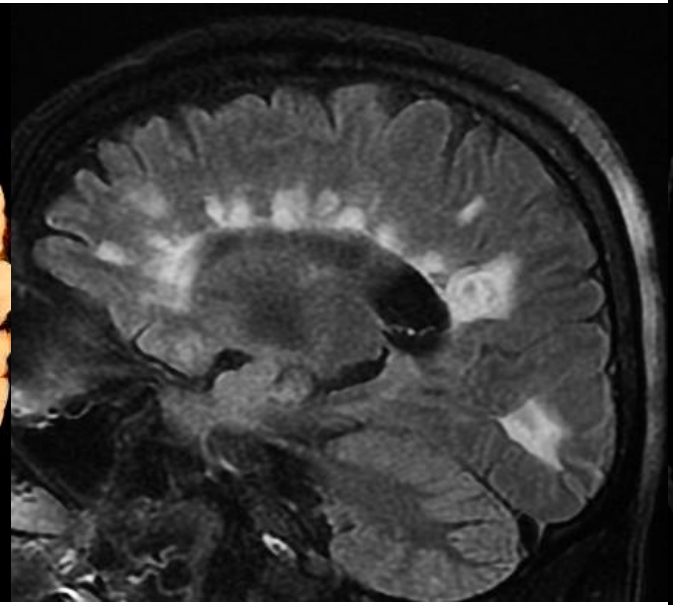
Dawson fingers

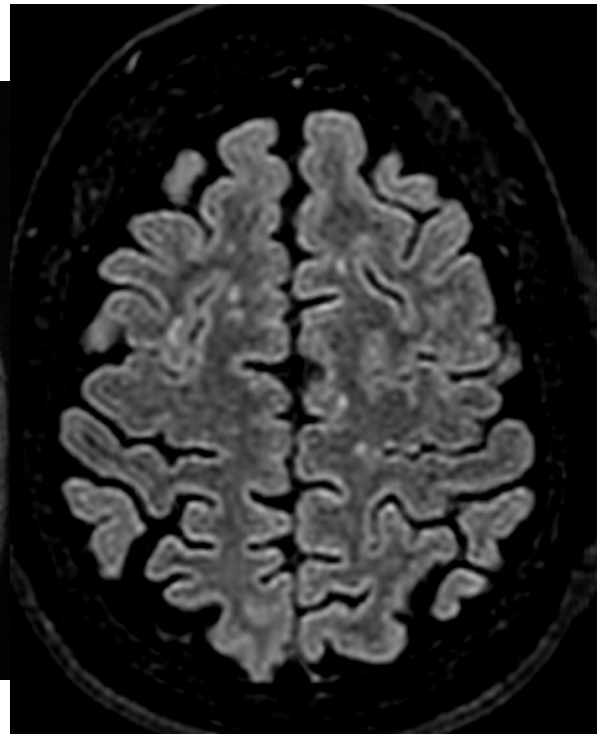
multiple lesioni ovalari o lineari iperintense in T2 perpendicolari all'asse ventricolare-ependimale ed a livello della giunzione calloso marginale

Lesioni possono coinvolgere
fibre sottocorticale associative brevi
(Fibre a «U»)



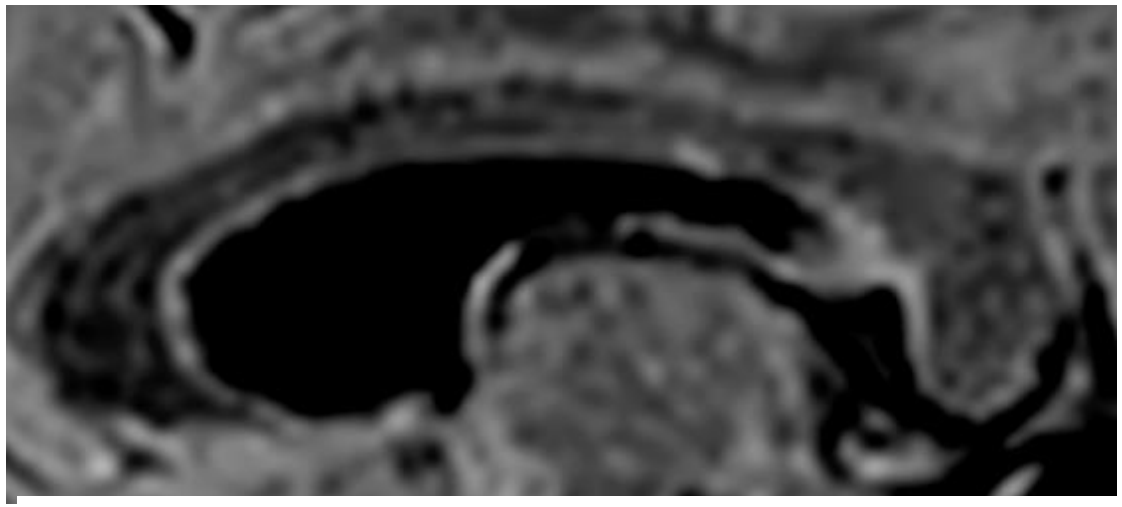






Sclerosi Multipla

Dot-dash sign



European Journal of Radiology 81 (2012) 3491–3495

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European Journal of Radiology

journal homepage: www.elsevier.com/locate/ejrad

MR imaging findings of the corpus callosum region in the differentiation between multiple sclerosis and neuromyelitis optica

Zhiye Chen[a], Feng Feng[b], Yang Yang[c], Jinfeng Li[a], Lin Ma[a,*]

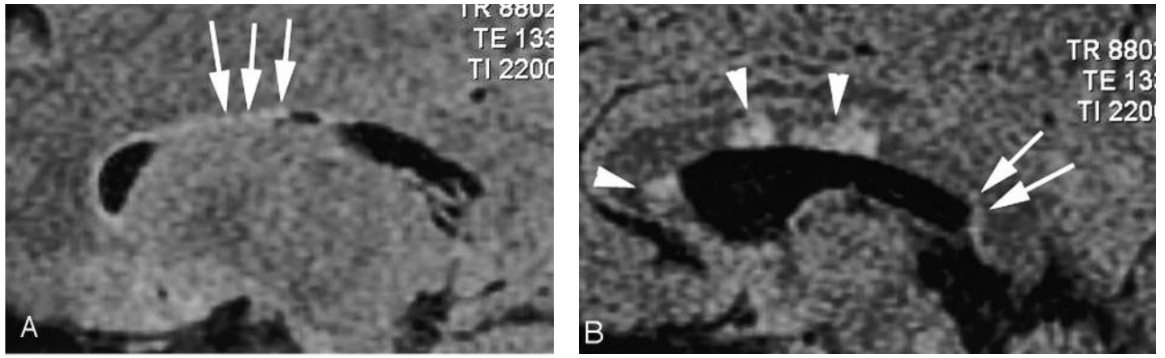
*Dot-dash sign subcallosale può essere utile per **distinguere SM** (segno frequente) dalla **neuromielite ottica** (segno meno frequente)*

The Ependymal "Dot-Dash" Sign: An MR Imaging Finding of Early Multiple Sclerosis

Christopher J. Lisanti, Patrick Asbach, and William G. Bradley, Jr.

AJNR Am J Neuroradiol 26:2033–2036, September 2005

Tempo 0 → 7 mesi

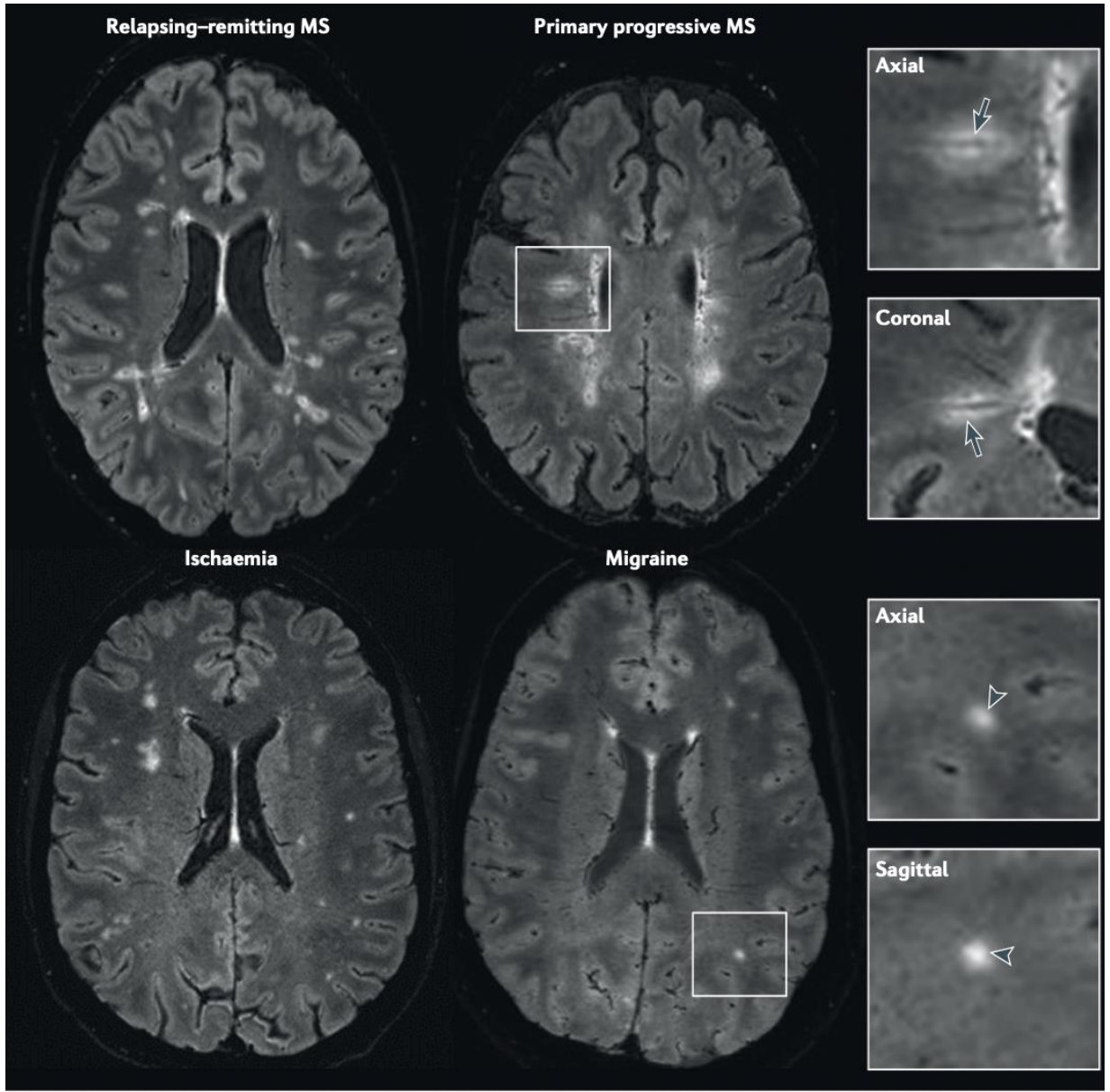


CONCLUSION: **The Dot-Dash sign of ependymal irregularity on thin-section sagittal fluid-attenuated inversion recovery images is an early marker for multiple sclerosis, which is particularly useful in the younger patient. This finding appears to be more sensitive for early lesion detection than any other multiple sclerosis imaging finding yet described in the literature.**

Nei giovani (<50 anni) : sensibilità >95% e specificità (>70%)

Negli soggetti > 50anni: ridotta sensibilità e specificità

Sclerosi Multipla



*The **central vein sign***

- ✓ Sottile ipointensità linere o puntiforme
- ✓ Visualizzati in due piani RM
- ✓ Diametro trasverso massimo <2mm
- ✓ Attraversa parzialmente o totalmente la lesione
- ✓ Posizionata centralemente la lesione

L'**incidenza** complessiva: **74%** (95% CI, 65-82%)

La **sensibilità** aggregata: **98%** (95% CI, 92-100%)

La **specificità** aggregata: **97%** (95% CI, 91-99%)

2016

CONSENSUS STATEMENT

OPEN

EXPERT CONSENSUS DOCUMENT

The central vein sign and its clinical evaluation for the diagnosis of multiple sclerosis: a consensus statement from the North American Imaging in Multiple Sclerosis Cooperative

Pascal Sati[1], Jiwon Oh[2,3], R. Todd Constable[4], Nikos Evangelou[5], Charles R. G. Guttmann[6],

The "Central Vein Sign" on T2- weighted Images as a Diagnostic Tool in Multiple Sclerosis: A Systematic Review and Metaanalysis using Individual Patient Data*

Chong Hyun Suh 2019

ORIGINAL CONTRIBUTION

Imaging Cortical Lesions in Multiple Sclerosis With Ultra–High-Field Magnetic Resonance Imaging

David Pitt, MD; Aaron Boster, MD; Wei Pei, MD; Eric Wohleb, BS; Adam Jasne, BS; Cherian R. Zachariah, BS; Kottil Rammohan, MD; Michael V. Knopp, MD, PhD; Petra Schmalbrock, PhD

Arch Neurol. 2010;67(7):812-818. **7 Tesla**

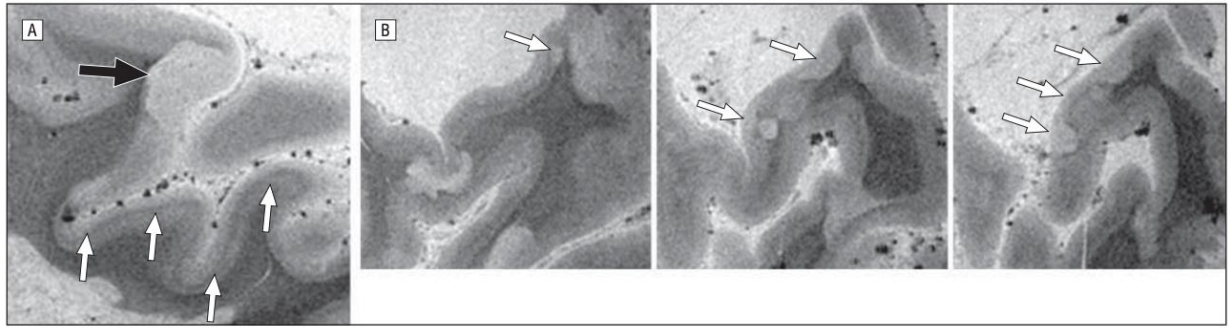


Figure 2. T2*-weighted gradient-echo magnetic resonance images. A, A type 4 cortical lesion (large arrow) and the line of Gennari (small arrows) in the occipital lobe. B, A cortical lesion morphs on consecutive magnetic resonance imaging cuts from a round, small type 2 lesion into a type 4 lesion (extending throughout the cortex) and fuses with a second lesion through a subpial bridge (arrows).

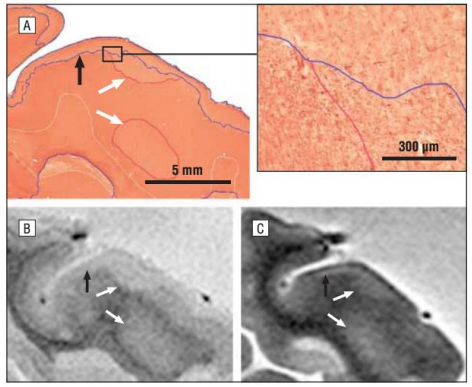


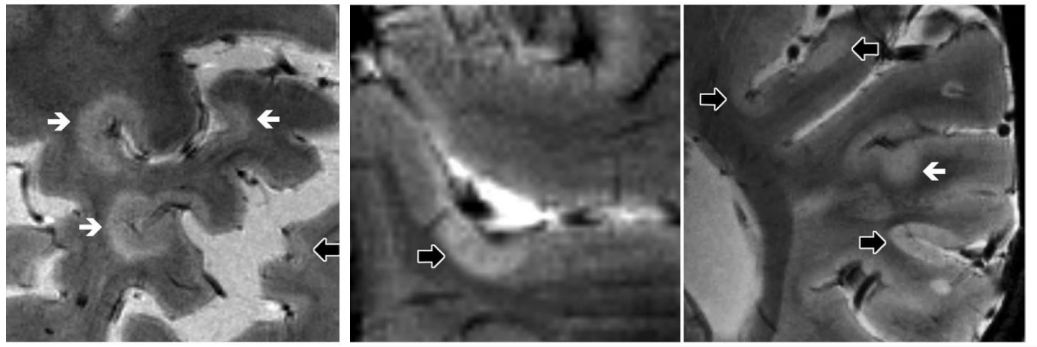
Figure 4. Cortical lesions. A, Myelin basic protein–labeled section containing demyelinated (black arrow) and thinly myelinated (white arrows) cortical lesions. Thinly myelinated lesions were seen on T2*-weighted gradient-echo (B) and white matter–attenuated turbo-field-echo (C) sequences. These lesions may be remyelinated; however, this has not be established by electron microscopy. Black arrows indicate demyelination; white arrows, thin myelination.

Conclusions: Three-dimensional T2*GRE and white matter–attenuated TFE sequences at a 7-T field strength detect most cortical lesions in postmortem multiple sclerosis tissue. This study indicates the potential of T2*GRE and white matter–attenuated TFE sequences in ultra–high-field magnetic resonance imaging for cortical lesion detection in patients with multiple sclerosis.

Longitudinal Characterization of Cortical Lesion Development and Evolution in Multiple Sclerosis with 7.0-T MRI

Constantina A. Treaba, MD, PhD • Tobias E. Granberg, MD, PhD • Maria Pia Sormani, PhD • Elena Herranz, PhD • Russell A. Ouellette, BS • Céline Louapre, MD, PhD • Jacob A. Sloane, MD, PhD • Revere P. Kinkel, MD • Caterina Mainero, MD, PhD

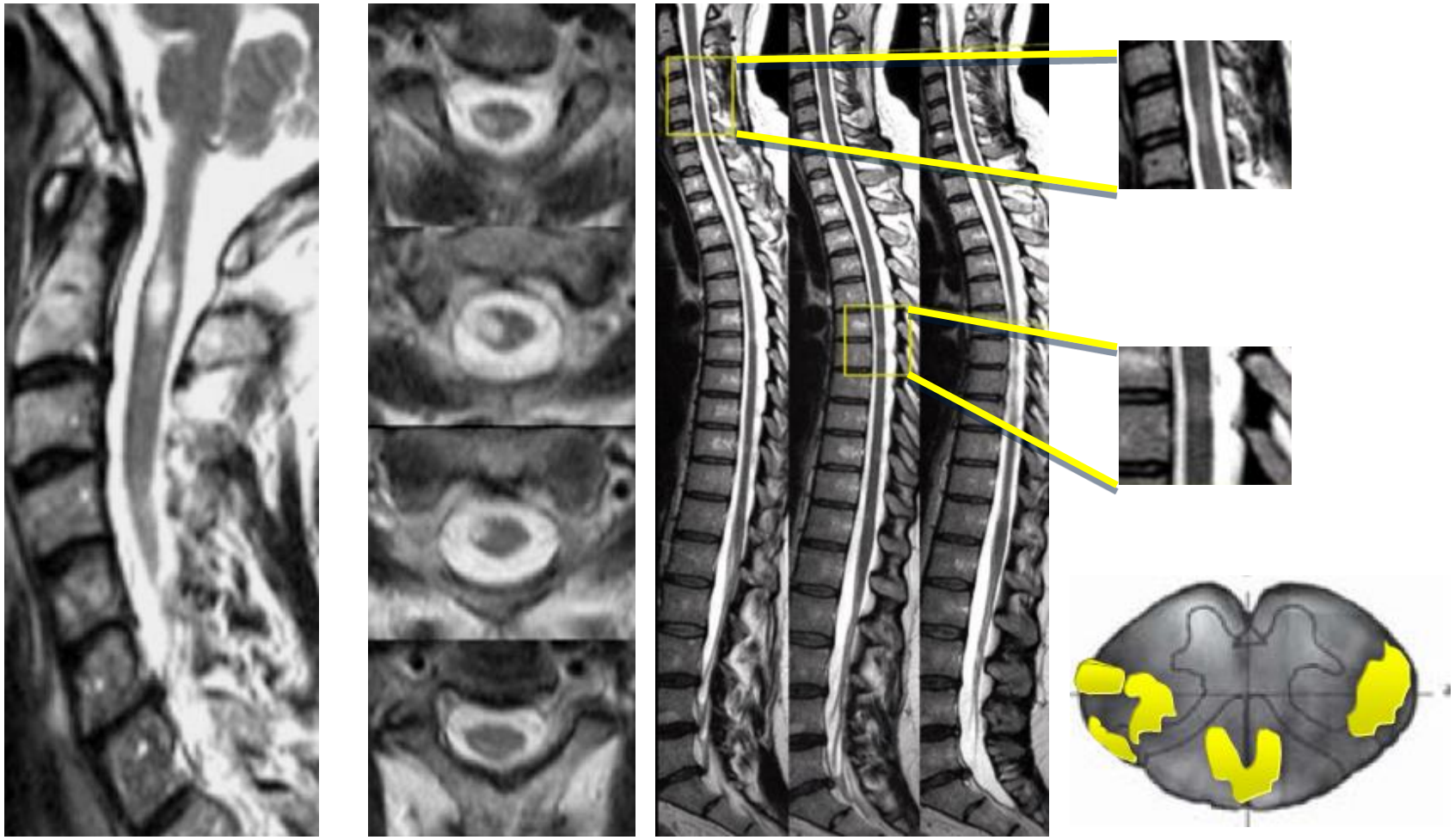
Radiology 2019; 291:740–749



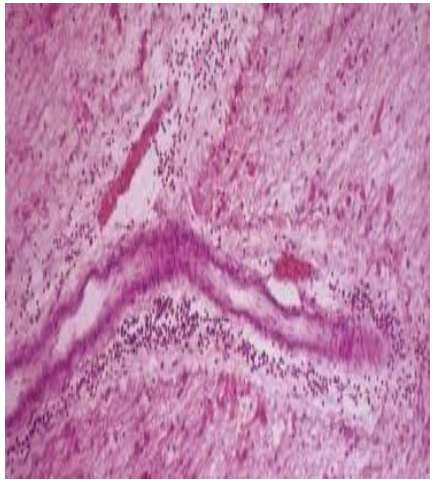
- Cortical lesions preferentially develop in cortical sulci, indicating a possible link with an ongoing cerebrospinal fluid–mediated neuroinflammatory process.
- Assessment of cortical lesions should represent a main component in the evaluation of progression of disease burden in multiple sclerosis.

Sclerosi Multipla

La presenza di lesioni midollari è un reperto altamente specifico per la diagnosi di Sclerosi Multipla.
La frequenza dell'interessamento midollare varia dal 75% al 90% nei Pazienti con diagnosi di MS



Sclerosi Multipla

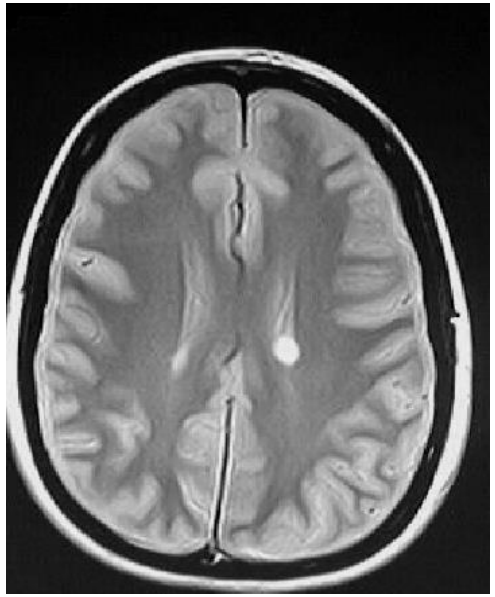


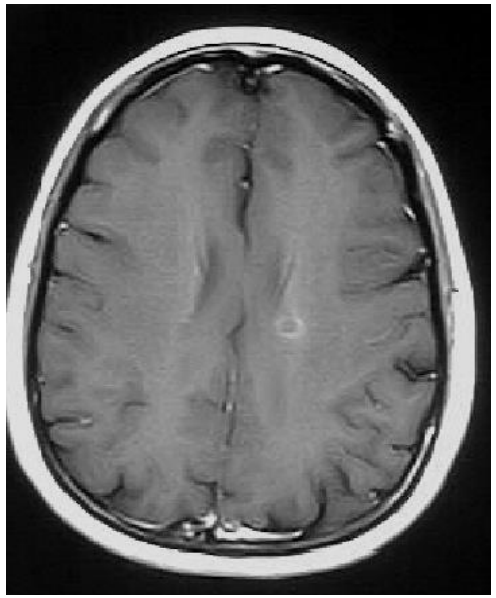
Risposta infiammatoria perivenulare

Danno barriera emato-encefalica

nelle prime 4 settimane ➔ RM potenziamento dopo mdc

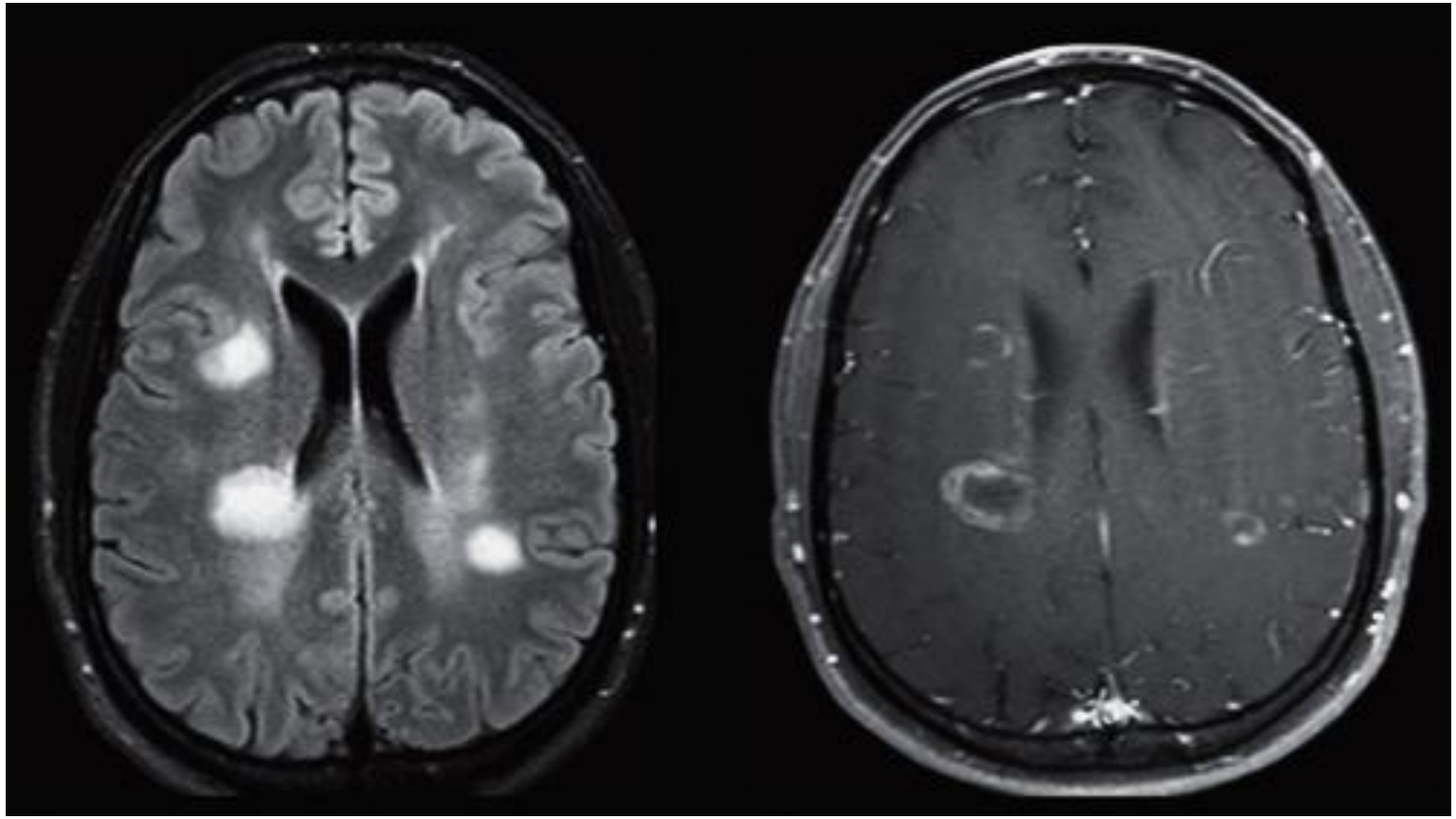
Leakage passivo di contrasto dallo spazio intravascolare-interstiziale



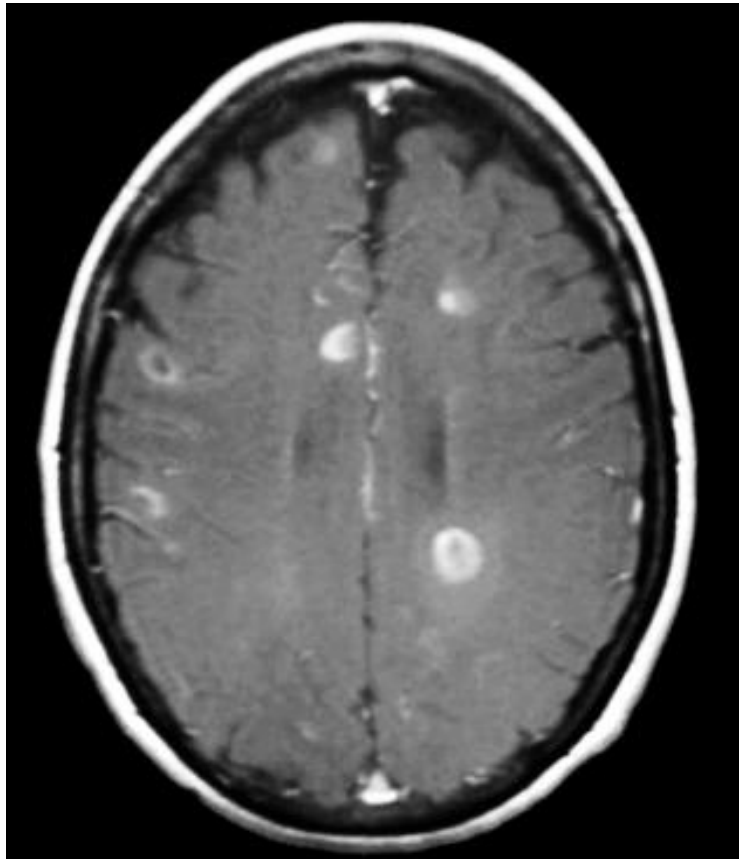


Sclerosi Multipla

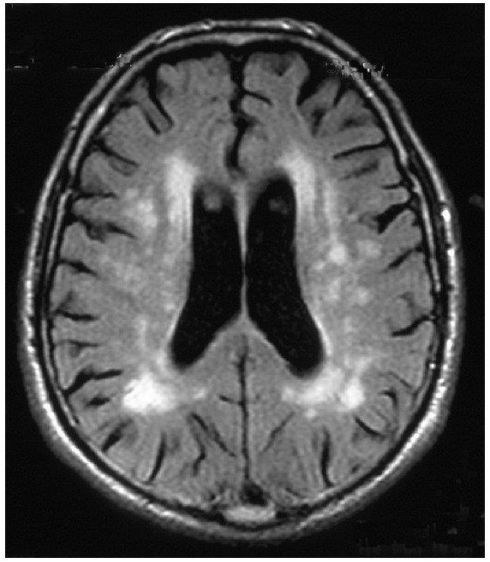
Placche che **potenziano** = demielinizzazione attiva



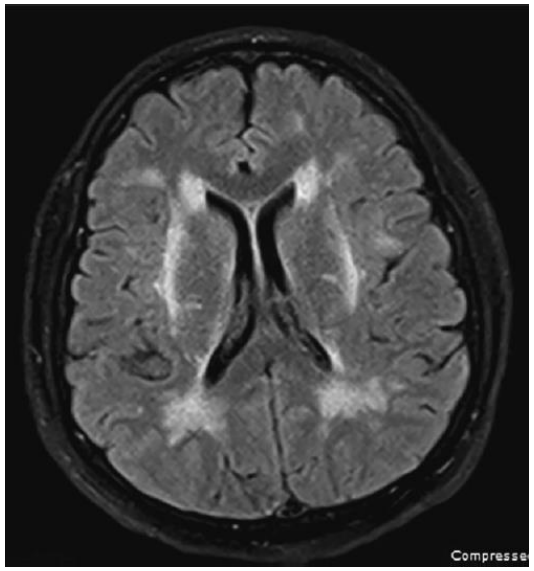
Pattern di **potenziamento**
(frequentemente ad anello incompleto)



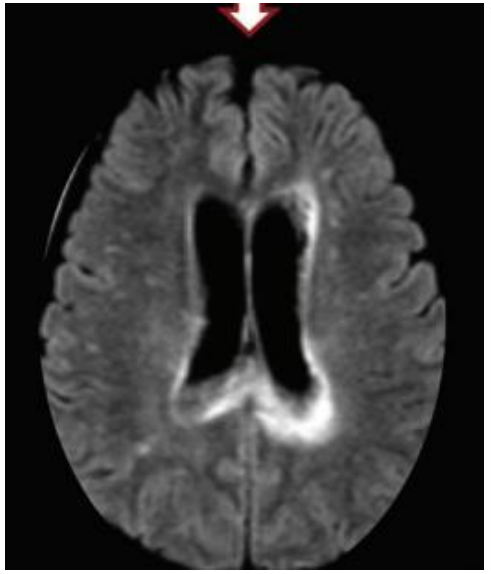
Sclerosi Multipla - ddx



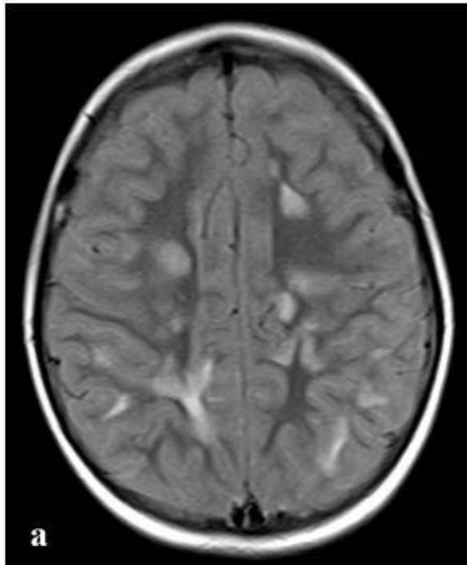
Cerebrovascolare



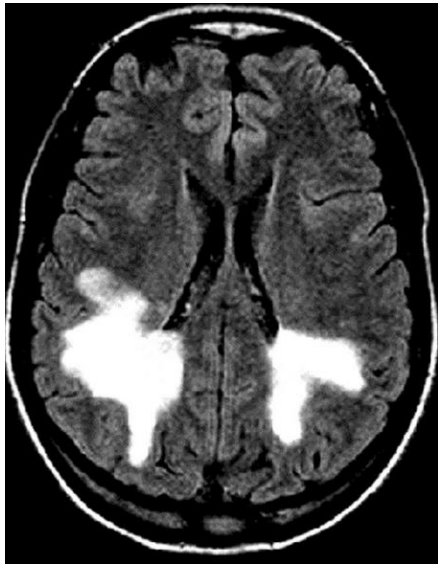
CADASIL



NMO



ADEM



Schilder

Tips

-stretta contiguità delle placche con il profilo ependimale

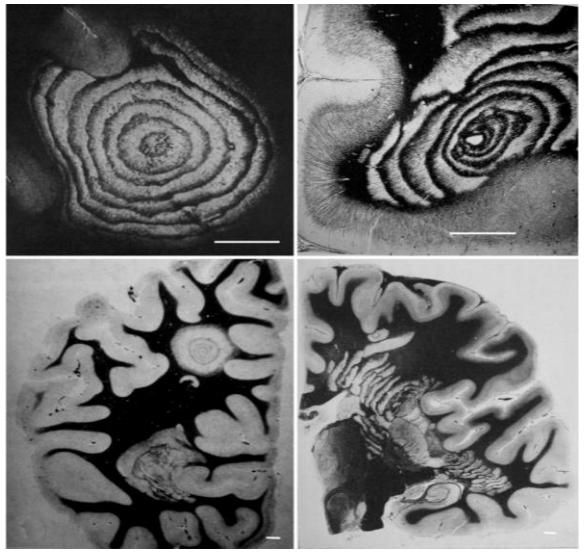
-Coinvolta la sostanza bianca attigua ai corni temporali dei ventricoli laterali

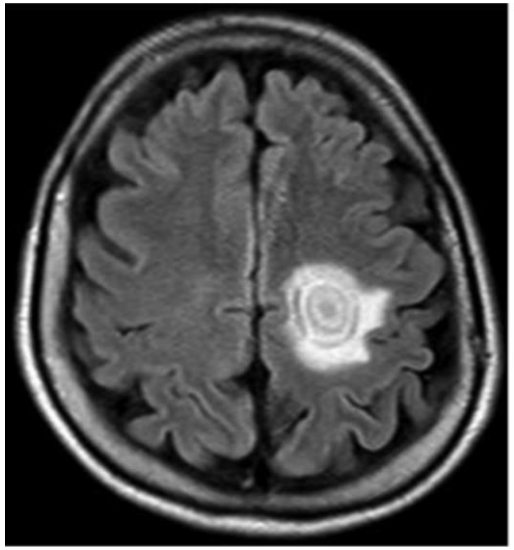
-Segno della vena centrale e del Dawson's finger

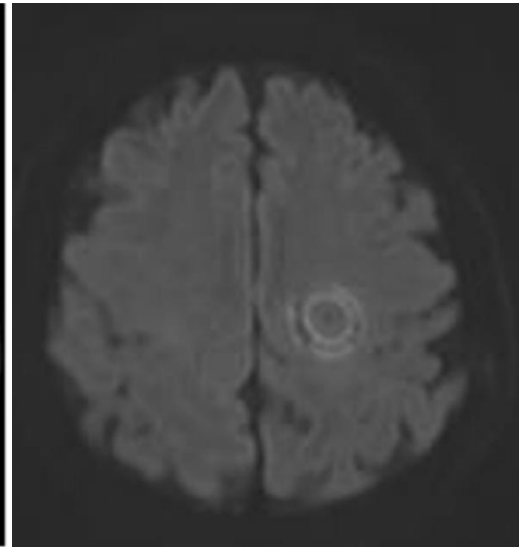
-Dot-dash sign

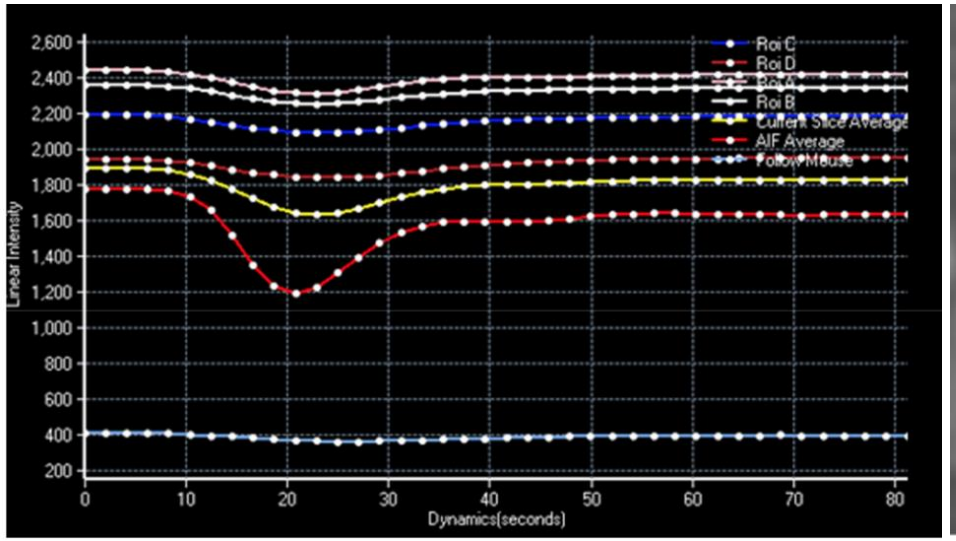
Sclerosi concentrica di Balò

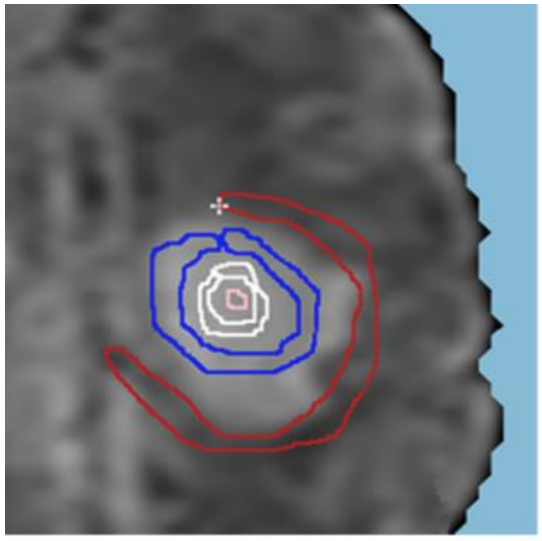
- Malattia demielinizzante rara e severa a decorso **monofasico**
- Istologia: bande di demielinizzazione e di sostanza bianca normale a **sviluppo concentrico**
- Imaging: bande di **alternato segnale**
  - -T1: aree concentriche ed irregolari di iso o iposegnale
  - -T2: aree concentriche ed irregolari di alternata iso/ipointensità e iperintensità
  - -DWI: diffusività ristretta nell'anello più esterno (infiammazione attiva – fronte)
  - -T1 Gd: potenziamento ad anello periferico (attiva demielinizzazione)





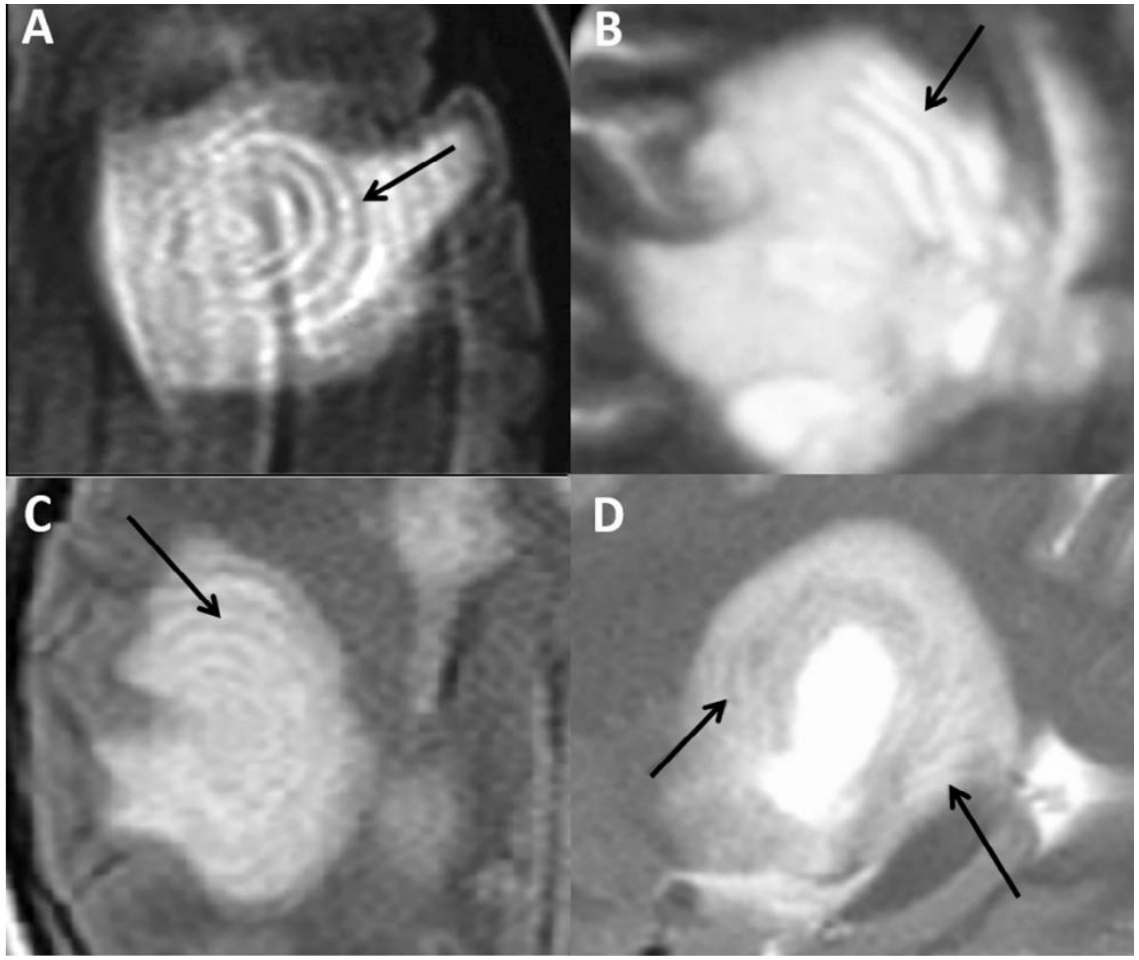


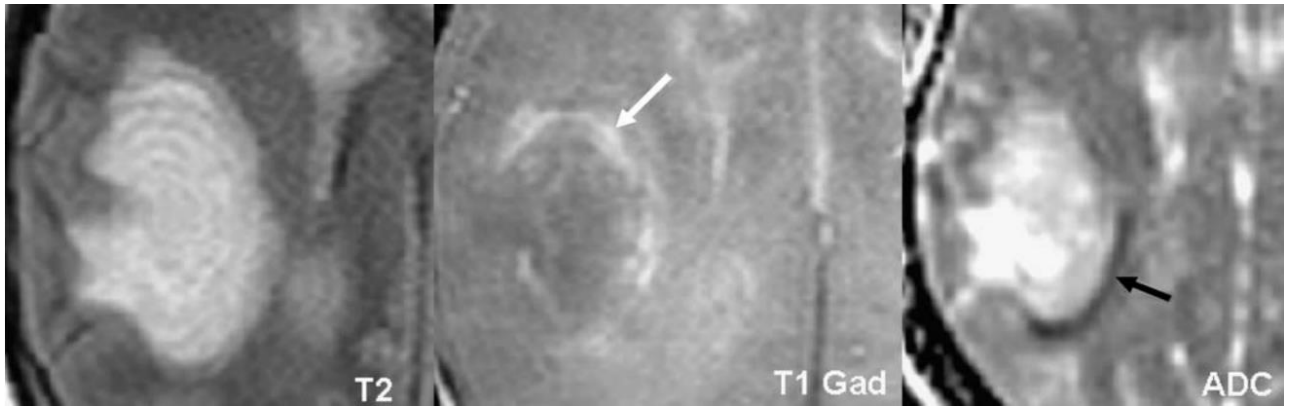




Maggior vascolarizzazione della componente centrale

Sclerosi concentrica di Balò





Porzione centrale: no lamelle per marcata demielinizzazione

Porzione periferica: pattern lamellato-stratificato

ADC: fronte di demielinizzazione (altamente cellulare)

Potenziamento: stratificato, ad anello incompleto

Neuroradiology (2007) 49:393–409
DOI 10.1007/s00234-007-0216-2

REVIEW

Idiopathic inflammatory-demyelinating diseases of the central nervous system

A. Rovira Cañellas · A. Rovira Gols · J. Río Izquierdo · M. Tintoré Subirana · X. Montalban Gairin

Chapter 21

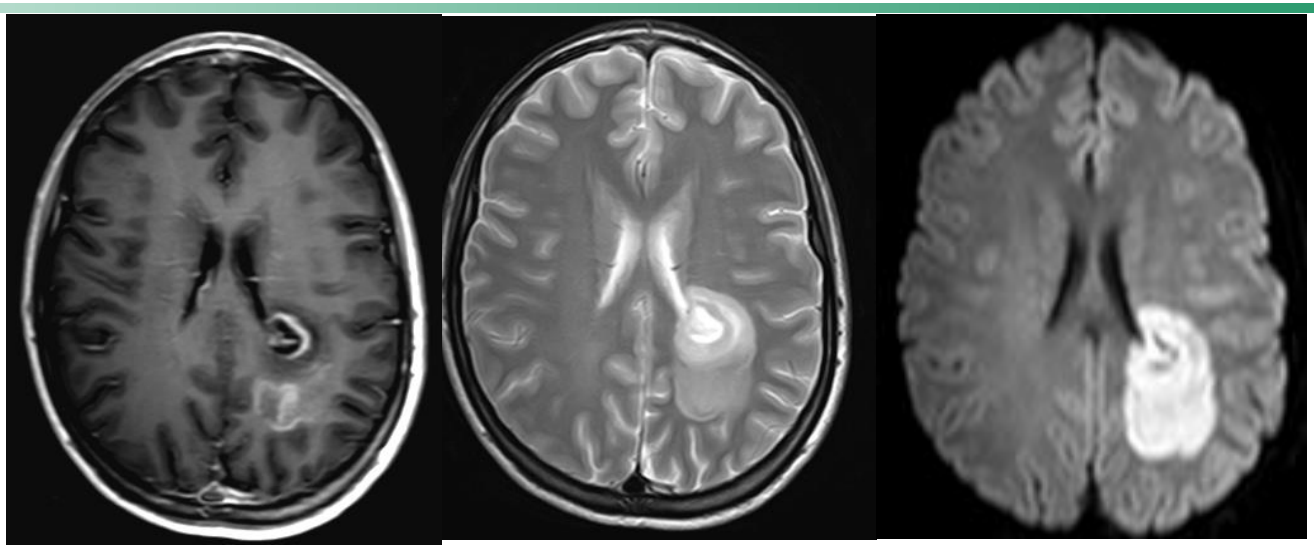
Other noninfectious inflammatory disorders

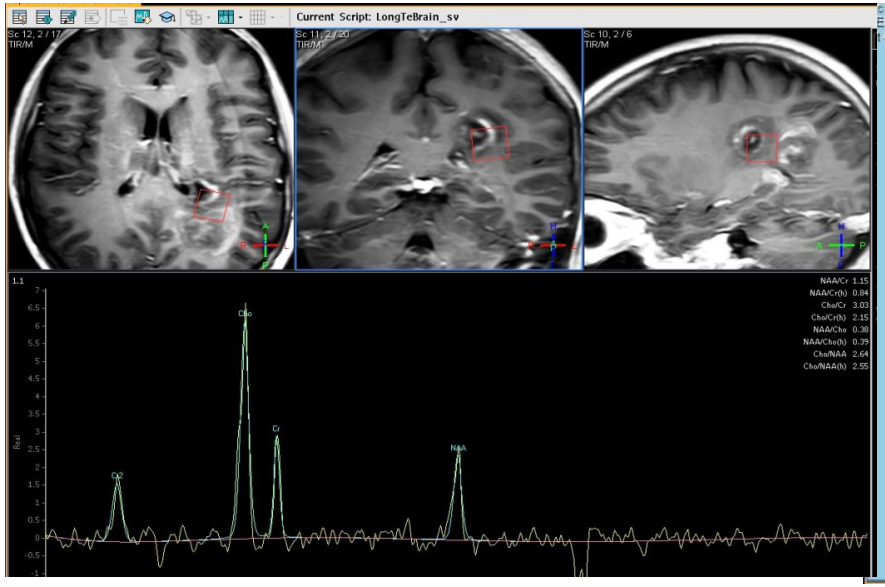
In: Handobbok of Clinical Neurology, vol. 135 (3° series) 2016

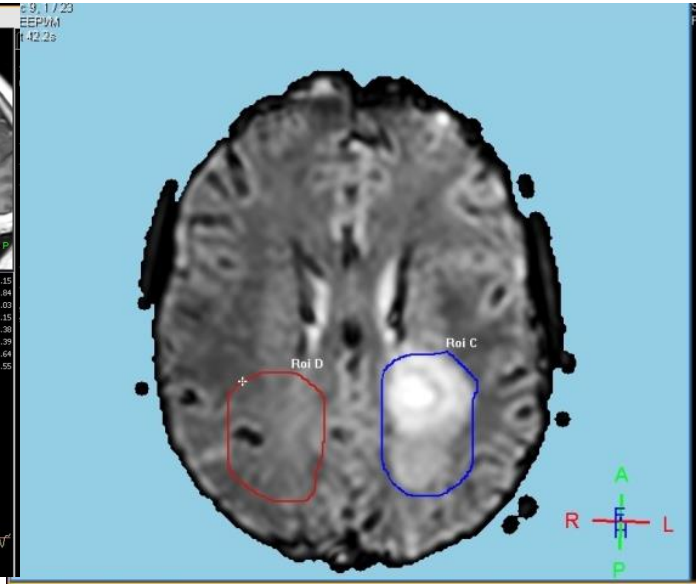
ÁLEX ROVIRA[1]*, CRISTINA AUGER[1], AND ANTONI ROVIRA[2]

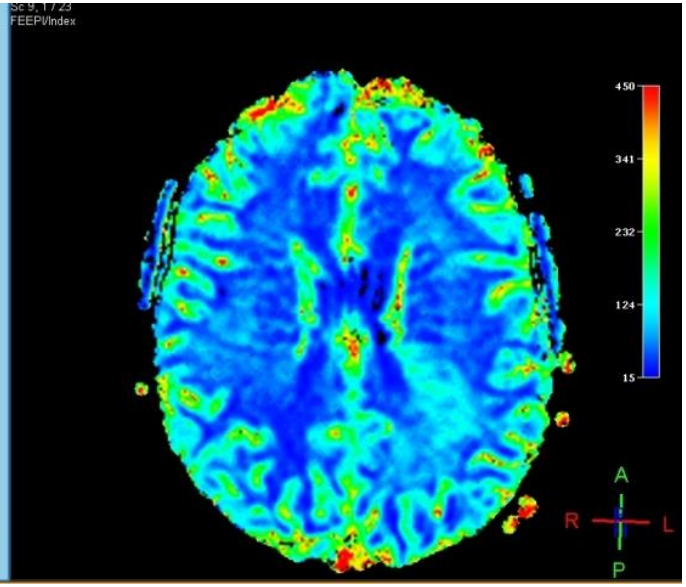
Sclerosi concentrica di Balò

Paziente: I.C.
Età: 18 anni, F
Disturbi cognitivi,
parestesie,
deficit visivi









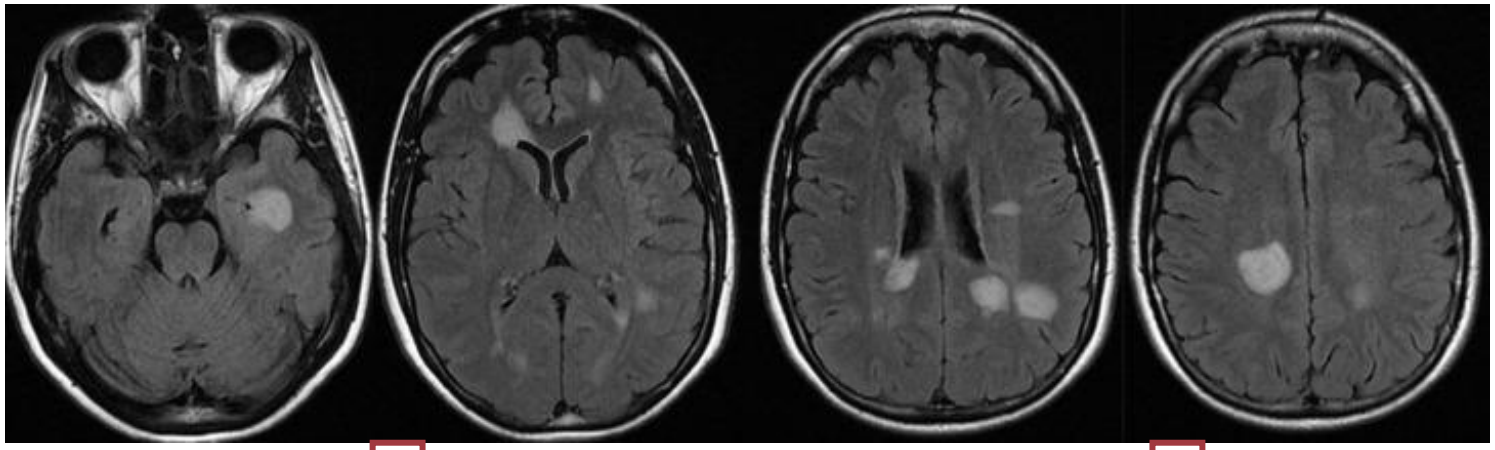
Sclerosi multipla – var. Marburg

- Malattia **acuta fulminante** - forma maligna

- Istologia: estensiva e fulminante demielinizzazione con elevata letalità

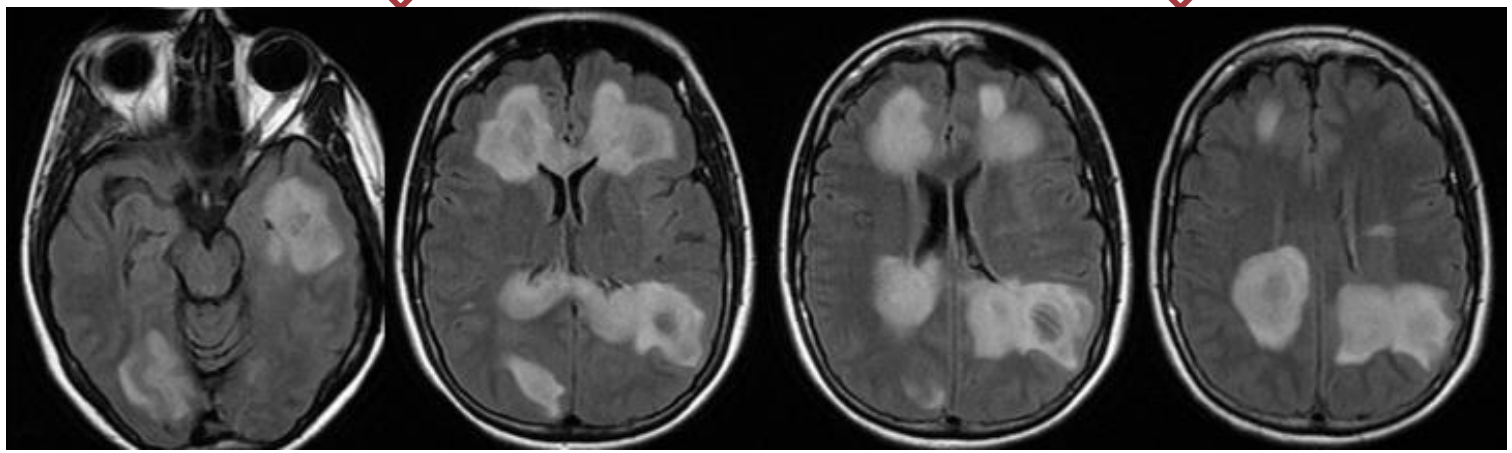
- Imaging:
  - **estese e confluenti aree tumefattive**;
  - effetto **massa**;
  - potenziamento ad **anello incompleto**;
  - Raramente possono presentarsi come lesioni più piccole e disseminate nell'encefalo.

Donna, 34aa con rapido declino cognitivo – RM tempo 0



Cortisone ad alte dosi

RM a 2 settimane



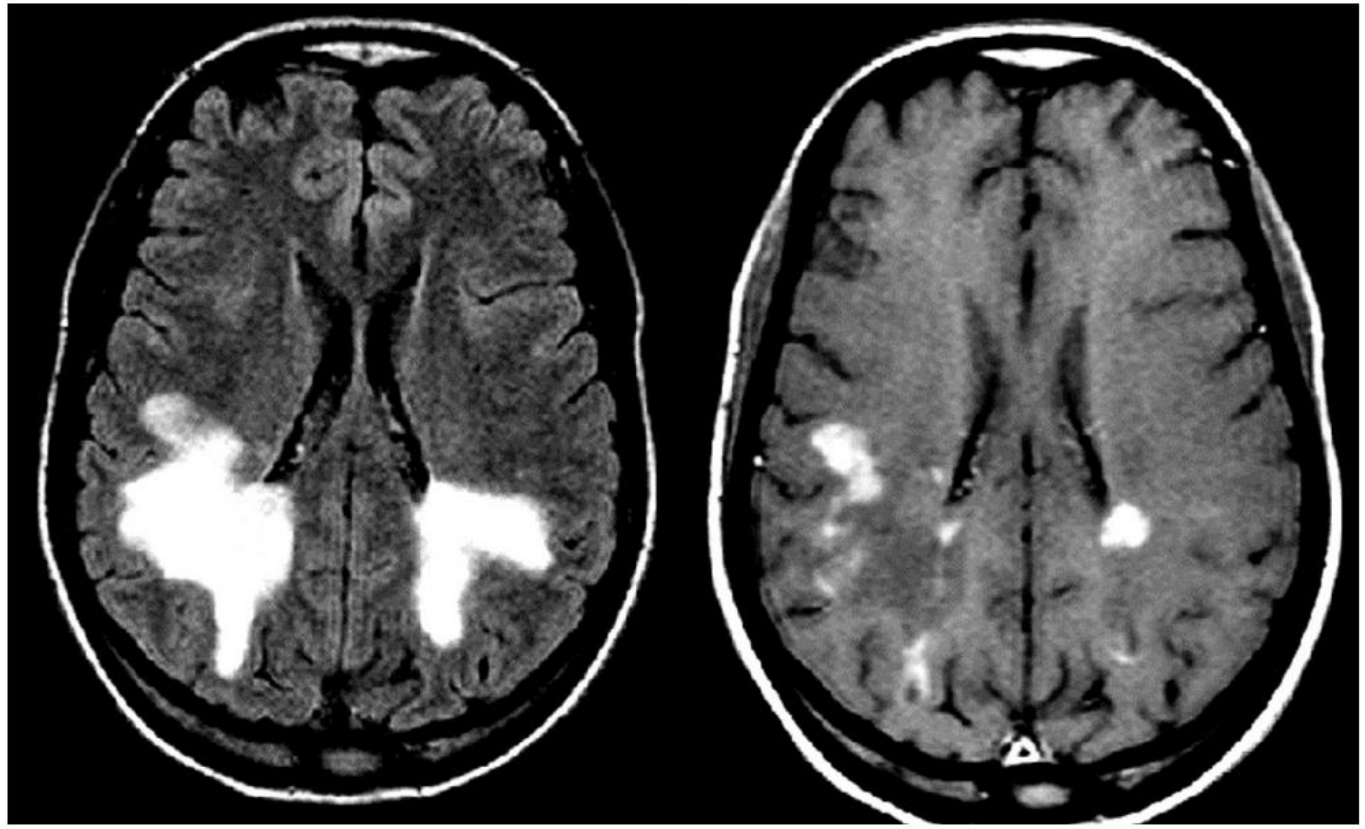
Malattia di Schilder

- Specifica presentazione clinico-radiologica di SM
- Decorso acuto – subacuto

- Istologia:
  - aree ben marcate di demielinizzazione e gliosi reattiva
  - degenerazione microcistica (con isolate cavitazioni)

- Imaging:
  - **esteso** e relativamente **simmetrico** coinvolgimento di **entrambi** gli **emisferi** cerebrali (prevalentemente parieto - occipitali)
  - potenziamento ad **anello incompleto o patchy**
  - modesto **effetto massa**
  - **diffusione ristretta**
  - **risparmio del tronco encefalo**

- Dd: tumore, ascesso e leucodistrofie



Chapter 21

Other noninfectious inflammatory disorders

In: Handobbok of Clinical Neurology, vol. 135 (3° series) 2016

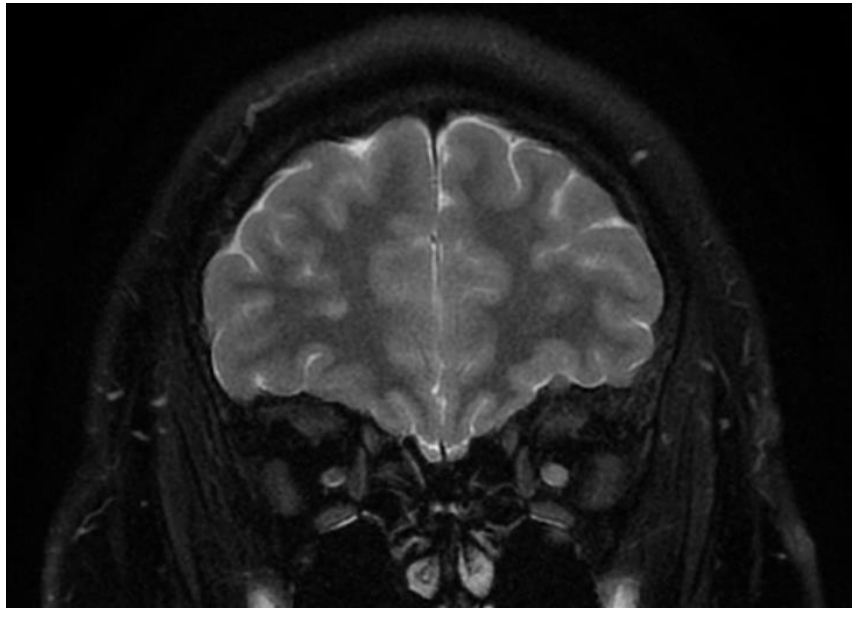
ÁLEX ROVIRA[1*], CRISTINA AUGER[1], AND ANTONI ROVIRA[2]

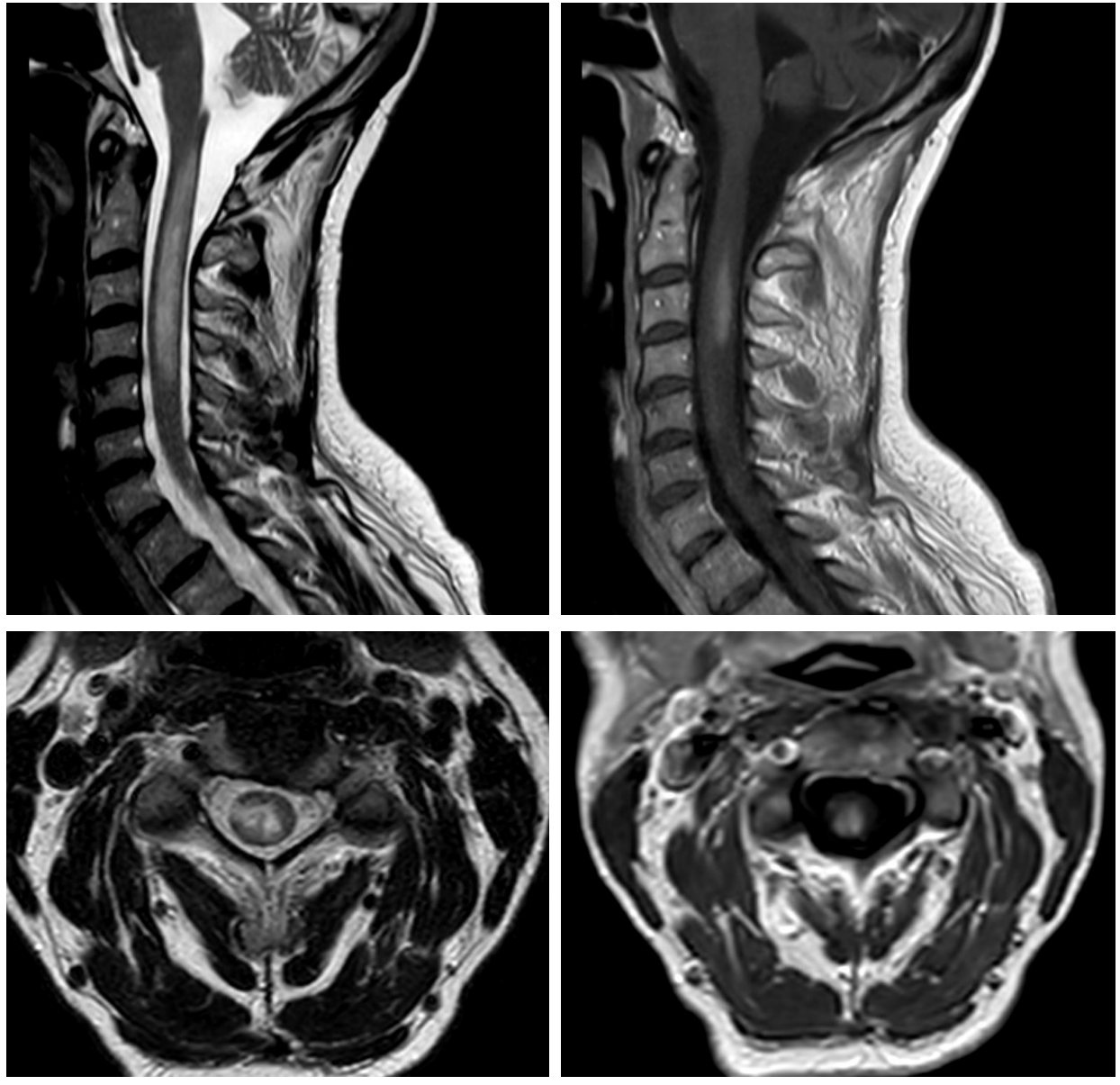
Neuromielite ottica – m. di Devic

- Caratterizzata da **neurite ottica** ed **estesa mielite longitudinale**

- Positività ad IgG anti AQP4

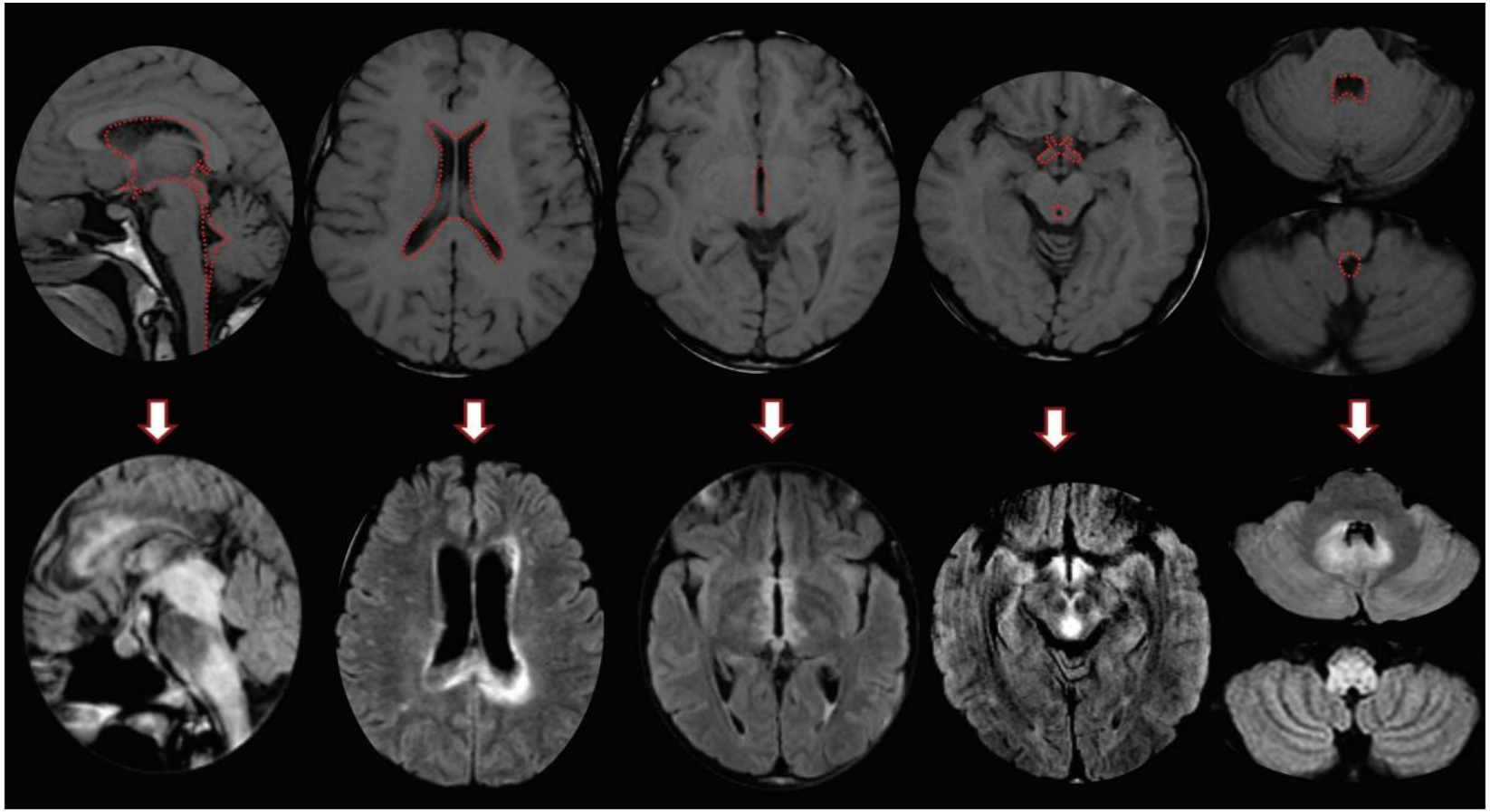
- Le lesioni encefaliche possono comparire successivamente nella malattia





Neuromielite ottica – m. di Devic

Distribuzione antigeni AQP4



Superficie ependimale corpo calloso

Ipotalamo

Area periacqueduttale

Area postrema

Canale midollare

Superficie ependimale ventricoli laterali

Regione diencefalica (III ventricolo)

Chiasma ottico

Superficie peri-ependimale del IV ventricolo

Aree circumventricolari: aree altamente vascolarizzate, con capillari fenestrati senza barriera emato-encefalica restrittiva

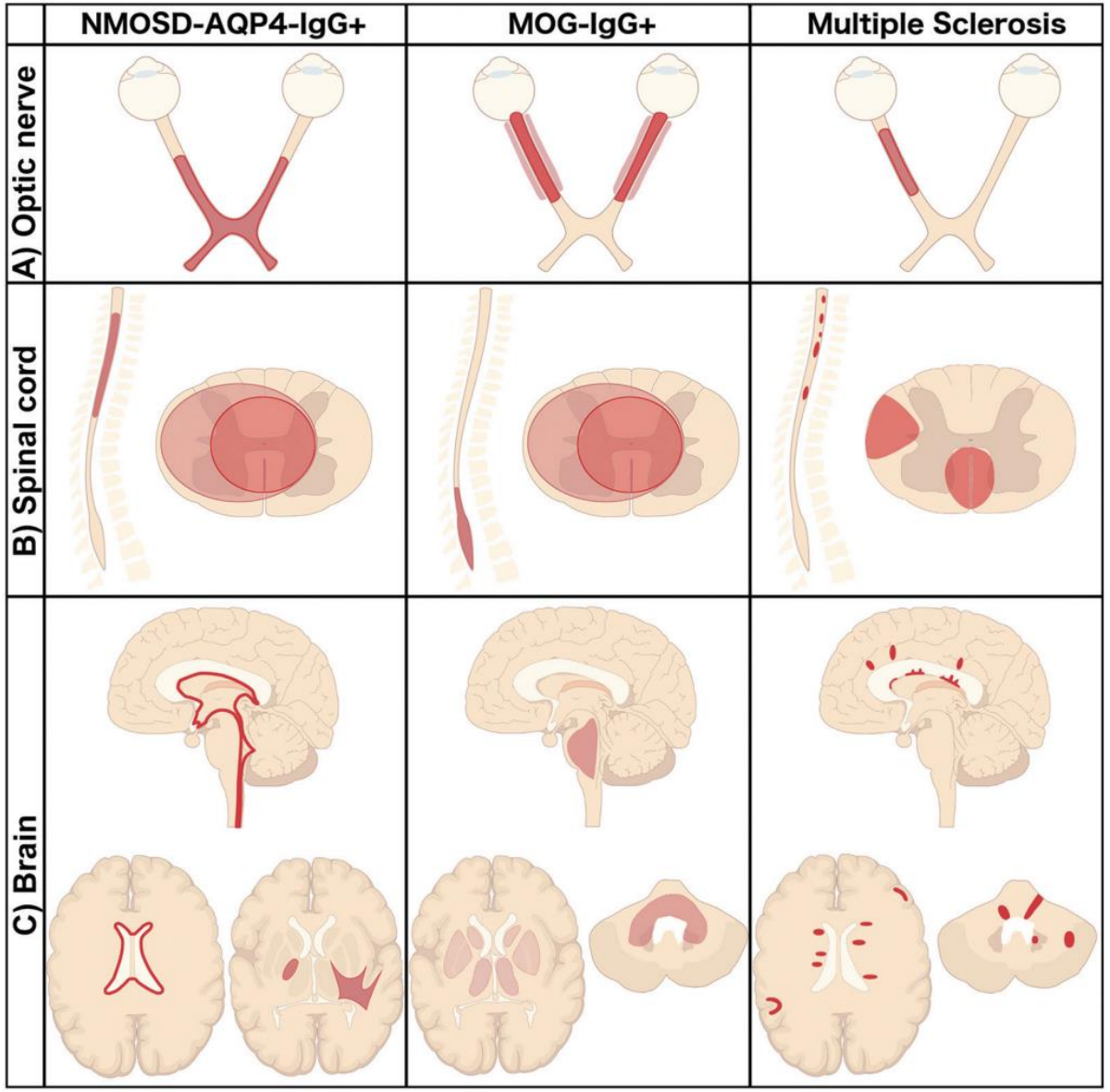
M. di Devic vs Sclerosi Multipla

Neuromyelitis Optica Spectrum Disorders: Spectrum of MR Imaging Findings and Their Differential Diagnosis[1]

Dutra et al 2018 - Radiographics

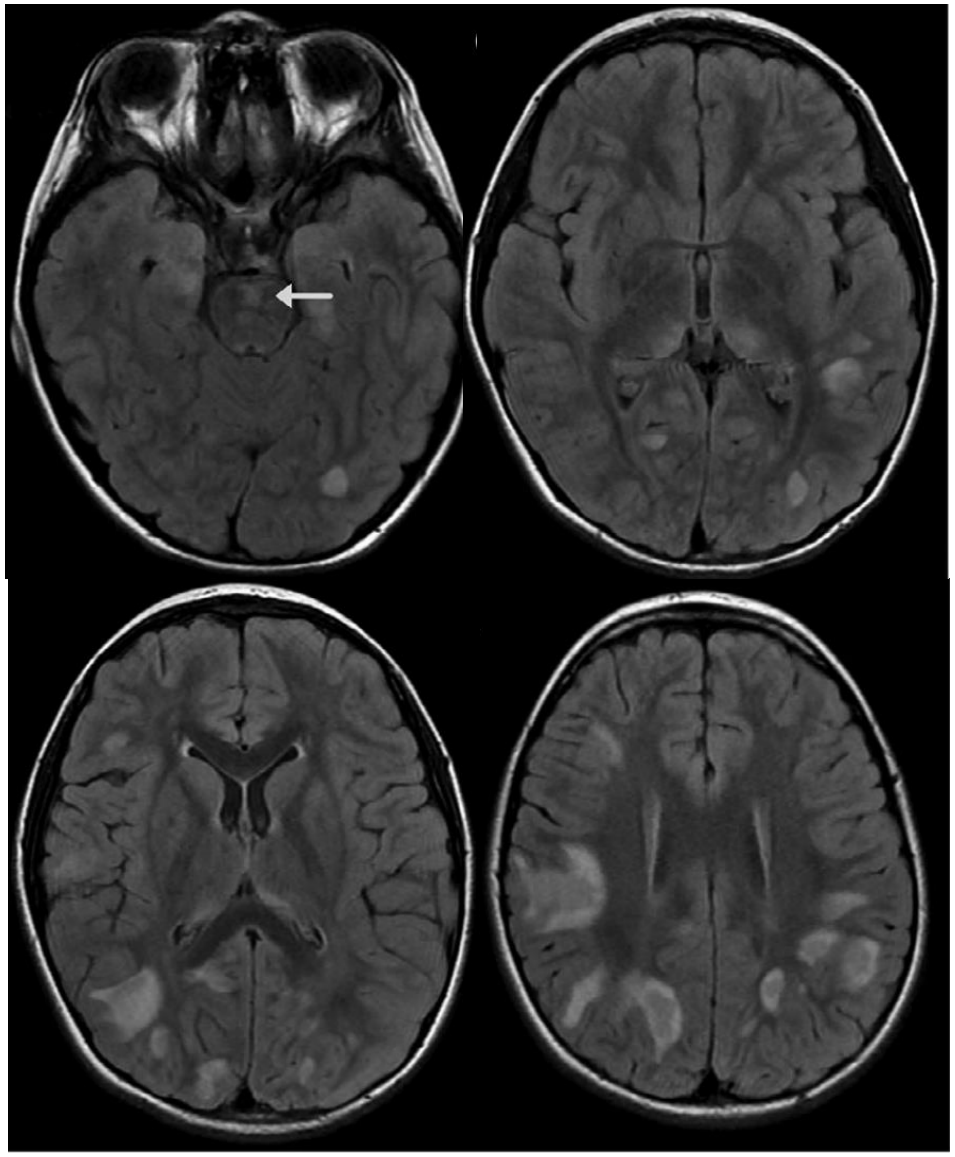
Diverse caratteristiche permettono di differenziare NMO dalla sclerosi multipla, ma nessuna è patognomonica

NMO	MS
Spinal cord	**Spinal cord**
Usually one big lesion.	Multiple lesions.
> 3 segments.	1-2 segments.
Central.	Posterolateral
T1 hypointense.	T1 isointense.
Ring enhancement.	Homogeneous enhancement.
Brain	**Brain**
Uncommon and late.	Common and early.
Periventricular.	Periventricular.
Perivenular, ovoid.	Following ependymus.
Thalamus/hypothalamus.	Leukocortical.



ADEM

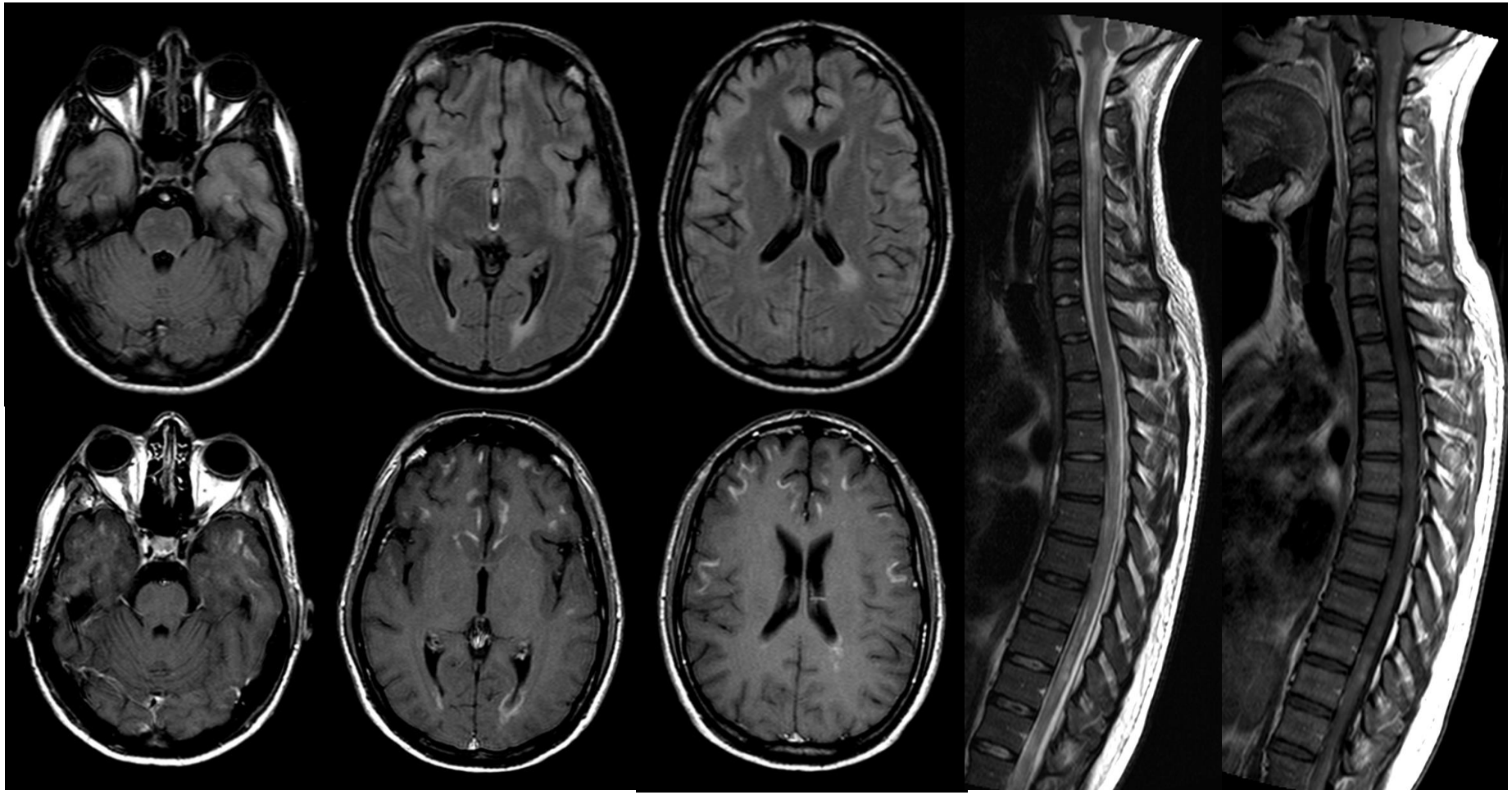
- Malattia infiammatoria acuta monofasica demielinizzante
- Frequente nei bambini e giovani adulti
- Recente **infezione** o **vaccinazione**!!!
- Nel 50% dei casi sono presenti **IgG anti-MOG**
- Possibile coinvolgimento dei gangli della base e della sostanza grigia



Imaging

- **Multiple** iperintensità **T2** con edema periferico nella sostanza bianca **sottocorticale** (possibile coinvolgimento talamo e tronco encefalo), **bilaterali** e **asimmetriche**
- **Potenziamento** puntato o ad anello incompleto lungo il margine di infiammazione
- **Diffusione ristretta in periferica** e facilitata centralmente
- Lesioni **midollari confluenti** con potenziamento variabile in 1/3 dei casi

ADEM



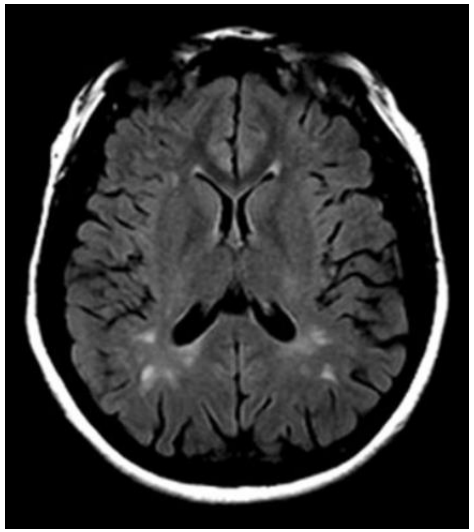
Acute disseminated encephalomyelitis following *Campylobacter jejuni* gastroenteritis: Case report and review of the literature

Floris R et al 2016

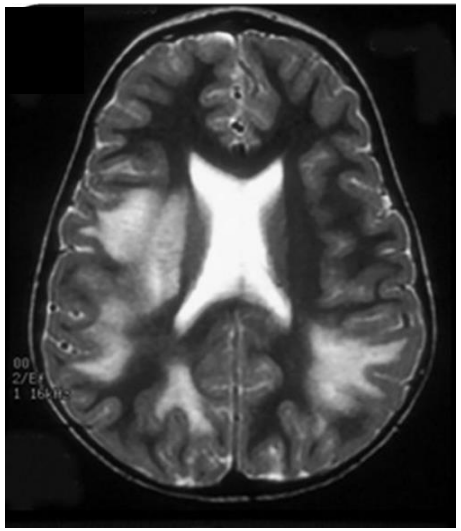
T2: iperintensità focali, bilaterali, asimmetriche, giunzionali, coinvolti anche i gangli della base

T1WI+C: puntato, ad anello, ad anello incompleto, periferico. Potrebbe non potenziare

SM vs ADEM



Sclerosi Multipla		Encefalomielite acuta disseminata
Sostanza bianca periventricolare Sostanza bianca calloso-marginale Sostanza bianca sotto e iuxta-corticale	**SEDE**	Sostanza bianca sottocorticale Tronco encefalo Nuclei di grigia e corteccia
Lesioni bilaterali simmetriche	**PATTERN**	Lesioni bilaterali asimmetriche
Sì	**BLACK HOLES (T1)**	No
Ben definiti	**MARGINI**	Mal definiti
Lesioni non confluenti	**ESTENSIONE**	Lesioni prevalentemente confluenti
Puntato – ad anello incompleto	**POTENZIAMENTO**	Ad anello incompleto



Criteri di Callen SM-ADEM

Applicabili alla **popolazione pediatrica** per distinguere ADEM da SM all'esordio;

almeno **due** dei seguenti criteri per avere una diagnosi di **SM**:

- ✓ Assenza di pattern diffuso e bilaterale delle lesioni
- ✓ Presenza di black holes
- ✓ Presenza di due lesioni periventricolari

A comparison of MRI criteria for diagnosing pediatric ADEM and MS

Neurology 2010 - Ketelslegers et al

Sensibilità: **75%** Specificità: **95%**

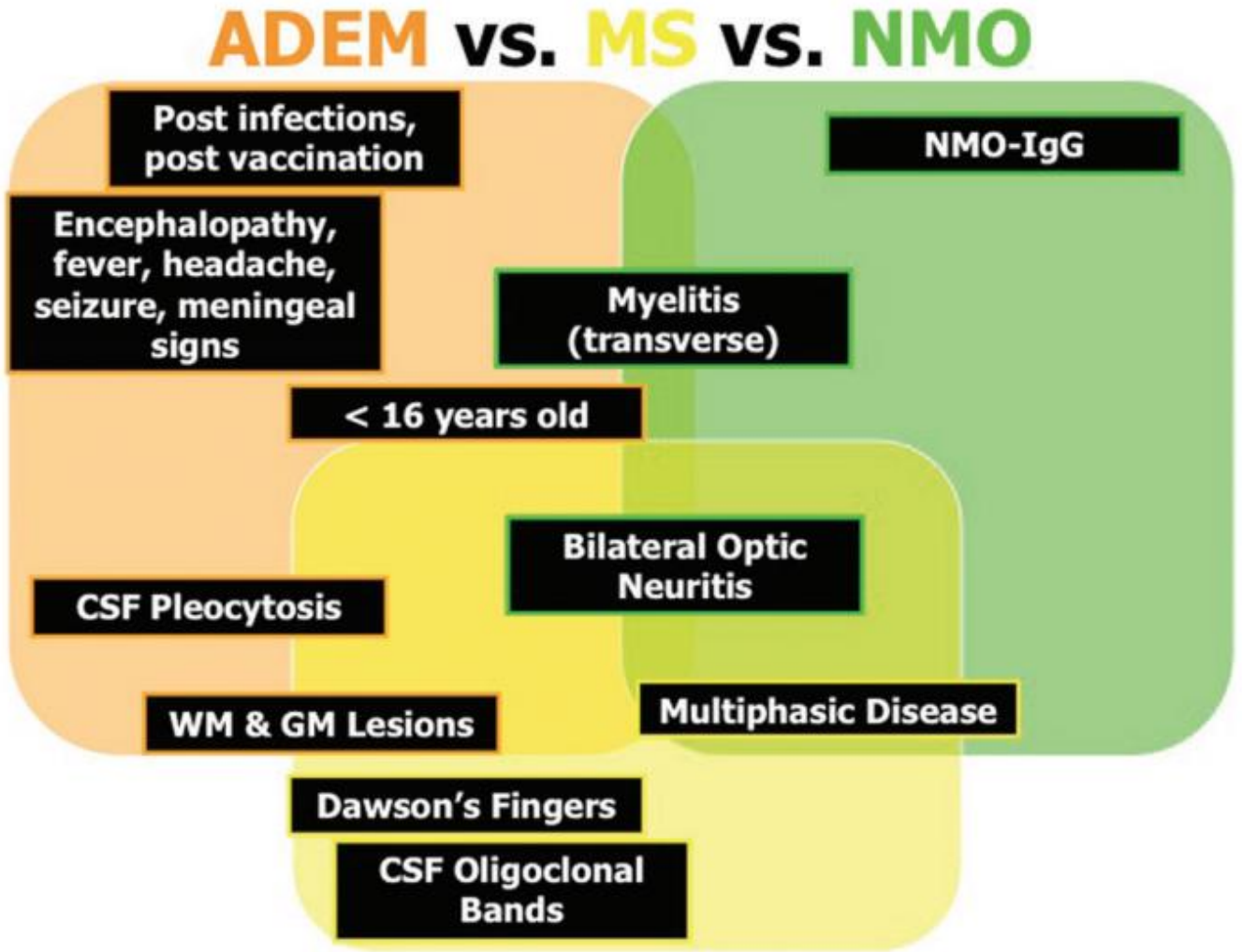
SM vs ADEM vs NMO

MedPix®

WM Lesion: Differential Diagnosis

	ADEM	MS	NMO
Clinical	79-95% Monophasic	Multiphasic	80-90% relapsing
Brain MR	Large (>1-2cm) WM lesions, deep GM involved	Smaller (≤ 1.5cm) WM lesions, ± cortex	ON lesions
Cord MR	Often confluent	Short segments (<3)	Long segments (≥3)
CSF Pleocytosis	> 49 WBCs/ml	uncommon	
Myelin Basic Protein	11%	75%	
Oligoclonal Bands	0-29% (5%)	64-92%	15-30%
NMO Protein	50-90% sensitive and 90-100% specific for NMO		

f08171e6-c5bc-42e6-b745-f4b344b141e8,Uploader : James G. Smirniotopoulos, M.D.,Source : James G. Smirniotopoulos, M.D.,11-05-2009



White Matter Diseases with Radiologic-Pathologic Correlation[1]

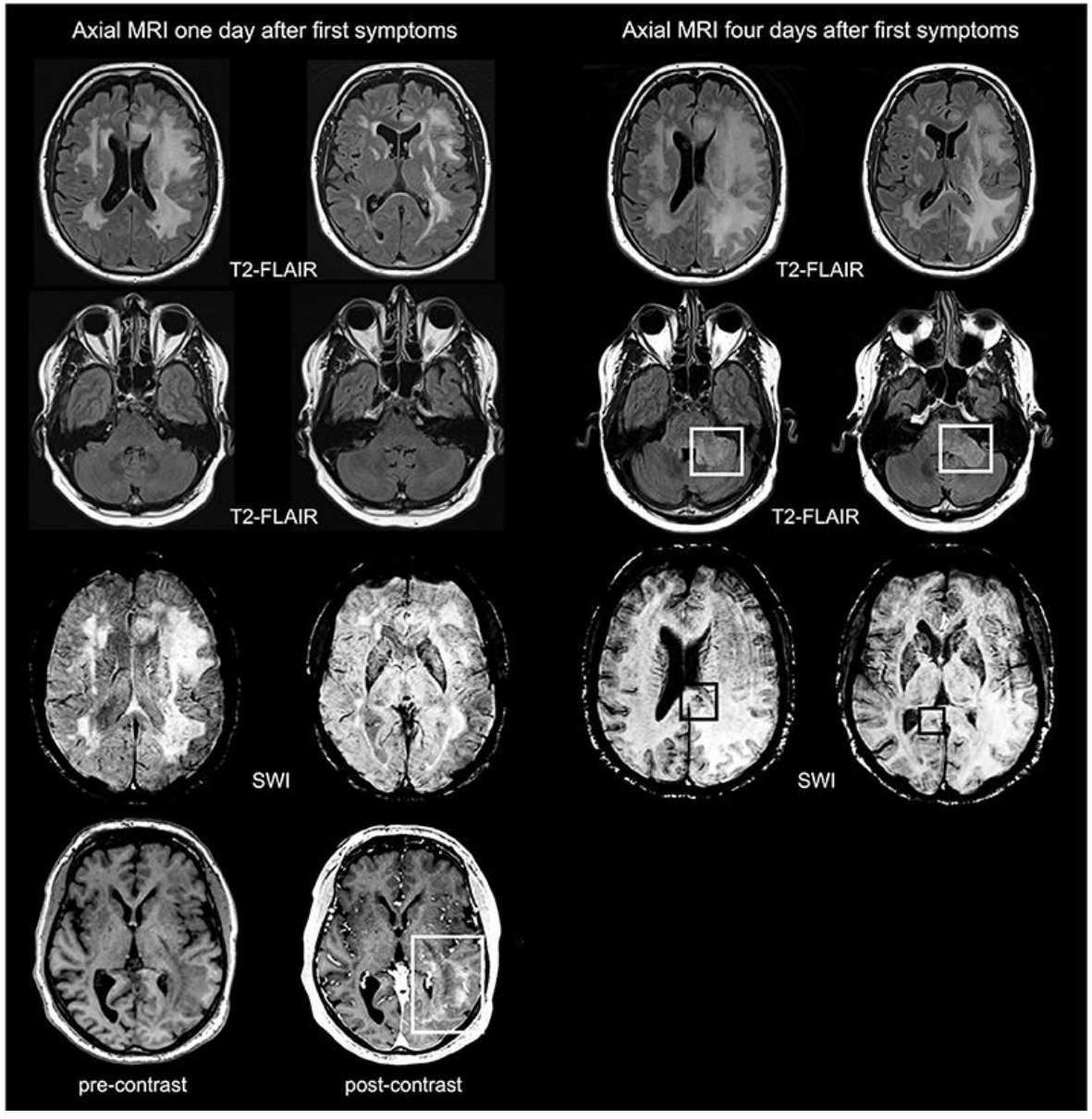
Sarbu et al. 2016 - Radiographics

Malattia di Hurst

- Encefalomielite acuta emorragica
  Leucoencefalite acuta emorragica
- Variante severa di ADEM, rapidamente progressiva
- Frequente nei giovani adulti
- Imaging
  - -lesioni **tumefattive** di grandi dimensioni
  - -**esteso** coinvolgimento della **bianca**
  - -risparmio della corteccia
  - -effetto **massa**
  - -**edema** perilesionale
  - -possibile coinvolgimento **talami** e **gangli della base**

DD: ADEM
La presenza di micro-emorragie è tipica della M. di Hurst

Utilizzare SWI>GRE



Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids

- Malattia infiammatoria del SNC descritta nel 2010
- Lieve predominanza M, età: 40-50 aa
- Etiologia : sconosciuta, probabilmente infiammazione immuno-mediata
- CLinica: atassia, diplopia, parestesie faciali, nistagmo
- Sede: **ponte** ++; può estendersi ai **peduncoli cerebellari**, al **mesencefalo**, al **bulbo**.

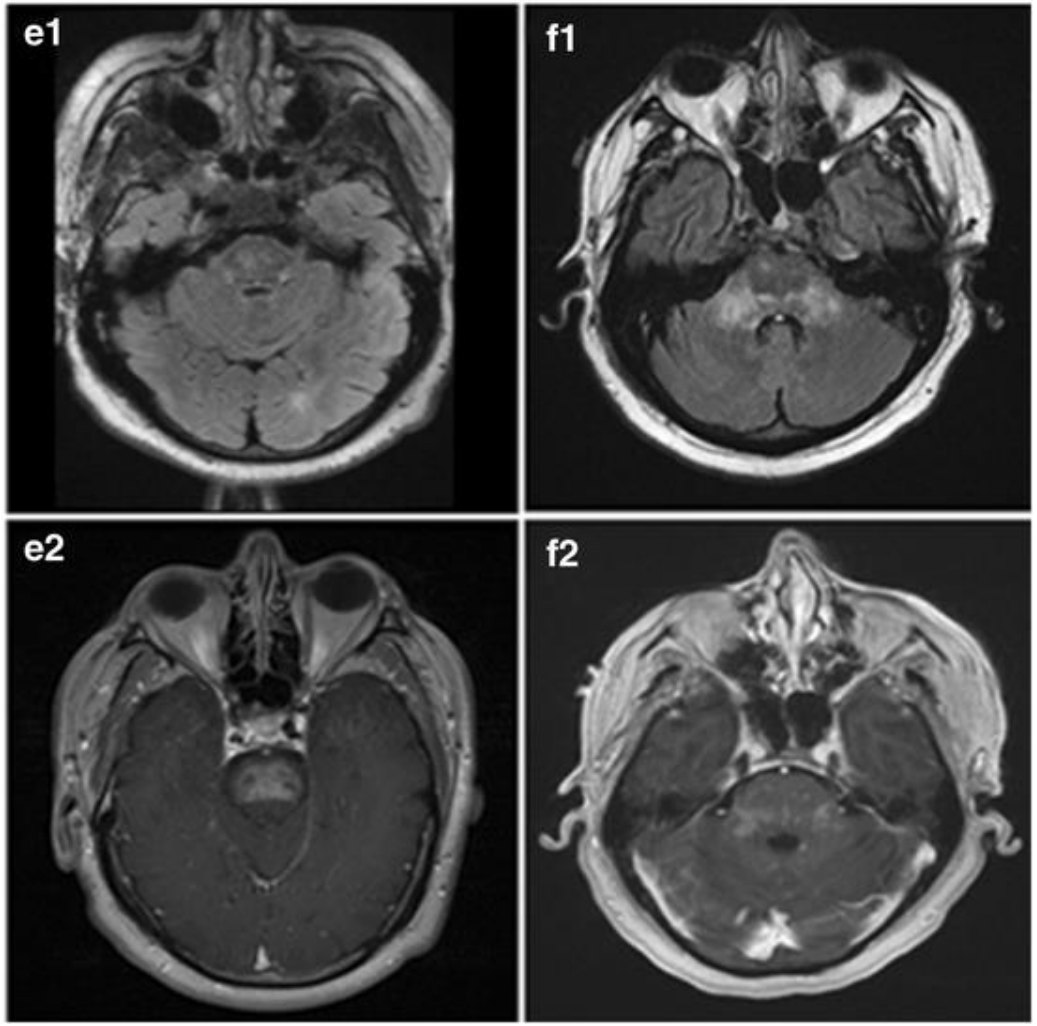
Imaging

-Lesioni sfumate, irregolari, puntiformi a livello del PONTE iperintense in T2 e T2-FLAIR.

-Potenziamento lineare o puntiformi.

-Ipointensità puntiformi in T2*WI (vene prominenti!!)

-DWI restrizione in fase acuta.



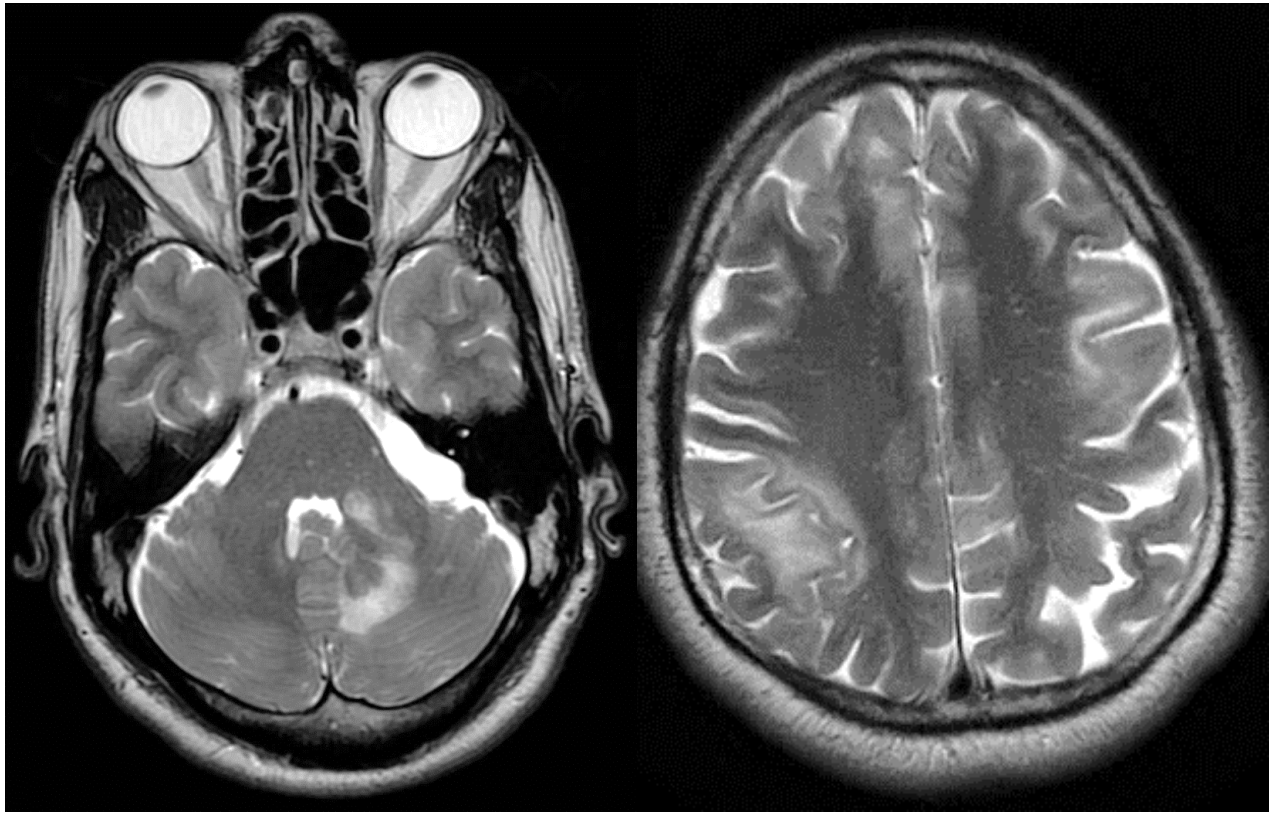
Pattern RM abbastanza univoco

Malattie infettive

Leucoencefalopatia multifocale progressiva

- Disordine progressivo demielinizzante
- Causa: **riattivazione JC virus**
- **Deficienza** immunitaria
- Secondaria a terapie **farmacologiche**

- CD4 < 100 (solitamente)



Imaging

-Multiple e piccole aree di ipersegnale T2, asimmetriche, nella sostanza bianca sottocorticale e profonda

-caratteristico coinvolgimento delle fibre sottocorticali a U

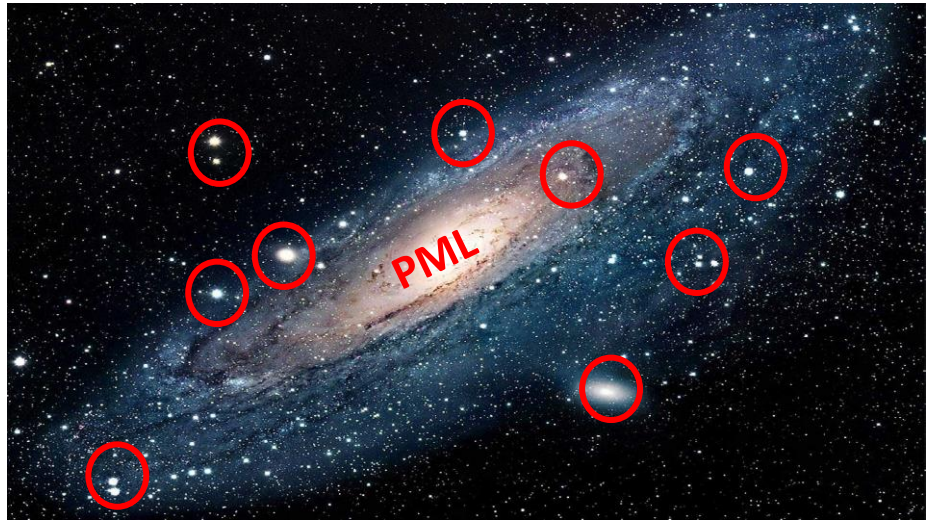
-solitamente non effetto massa né potenziamento (potenziamento nella fase inziale infiammatoria)

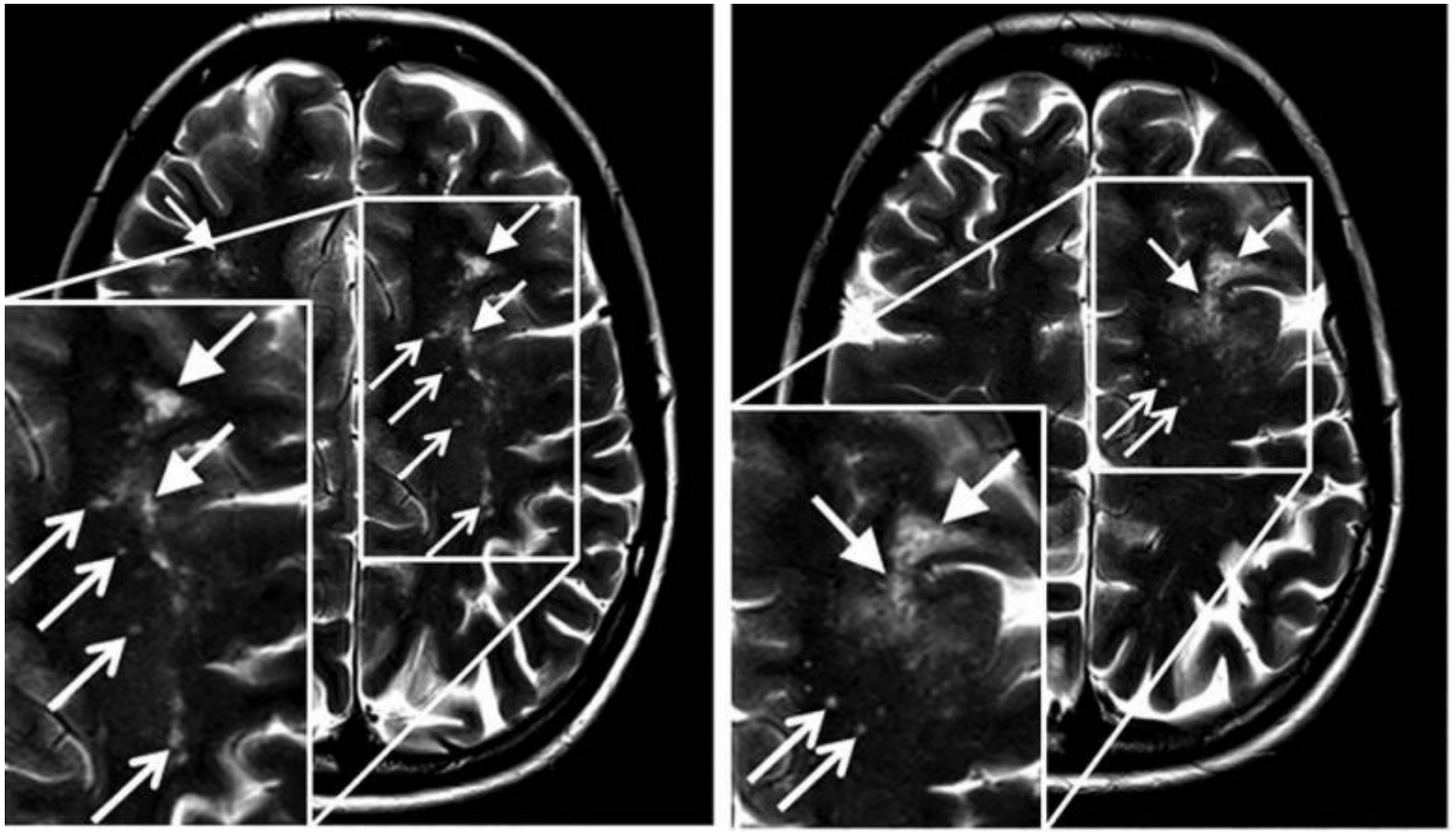
Leucoencefalopatia multifocale progressiva

The Milky way sign

- Pattern puntato
- Utile nella diagnosi differenziale con SM

- ***Multiple alterazioni T2 che circondano il core di una nuova lesione di PML***

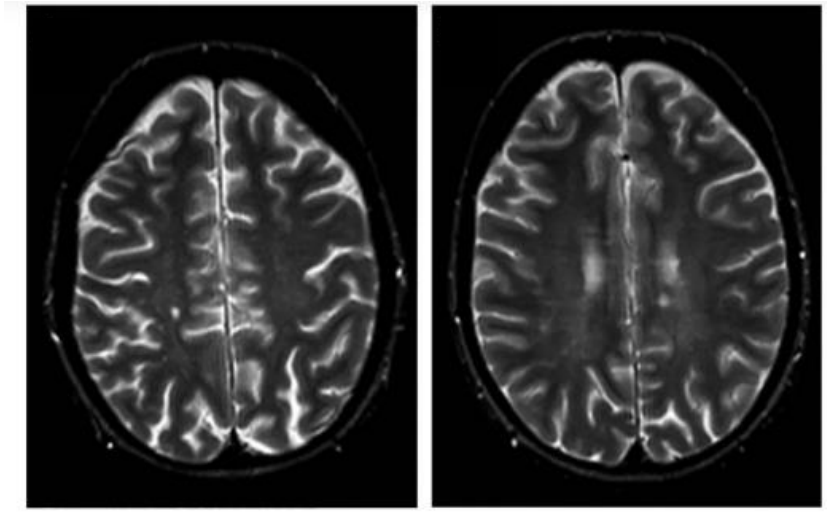




Frecce chiuse: PML

Frecce aperte: alterazioni T2 contigue

Leucoencefalopatia multifocale progressiva

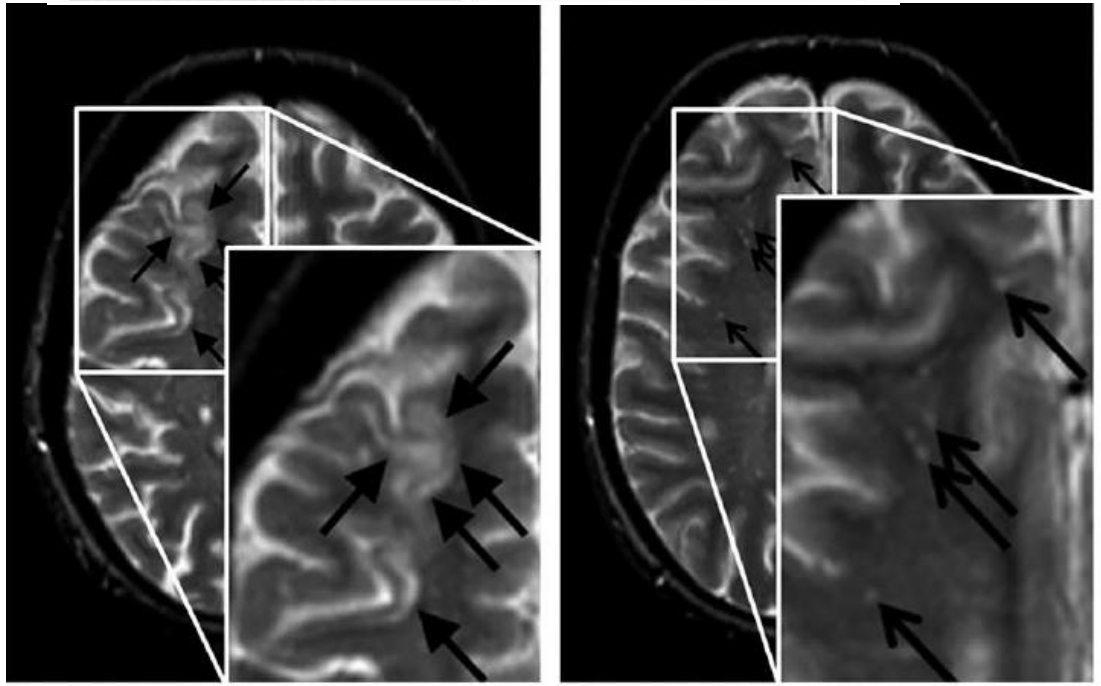


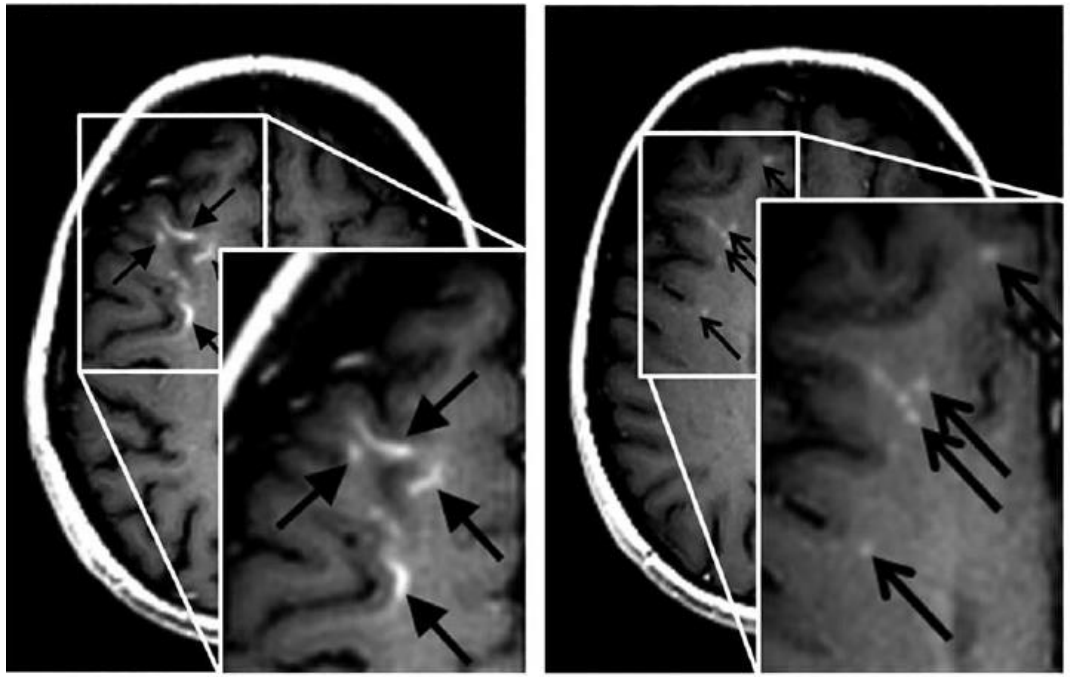
Multiple sclerosis

RESEARCH PAPER

MRI criteria differentiating asymptomatic PML from new MS lesions during natalizumab pharmacovigilance

Wijburg et al. 2016





PML - IRIS

- IRIS: sindrome da ricostituzione immunitaria
- Pazienti immuno-modulati (sclerosi multipla) e con HIV/AIDS
- JC positivi
- Trattamenti prolungati con Natalizumab

- Due tipi di PML-IRIS sono riconosciuti:

-**PML-s-IRIS**: simultaneo sviluppo di IRIS e PML per ricostituzione immunitaria

-**PML-d-IRIS:** ricostituzione immunitaria con peggioramento di preesistente PML

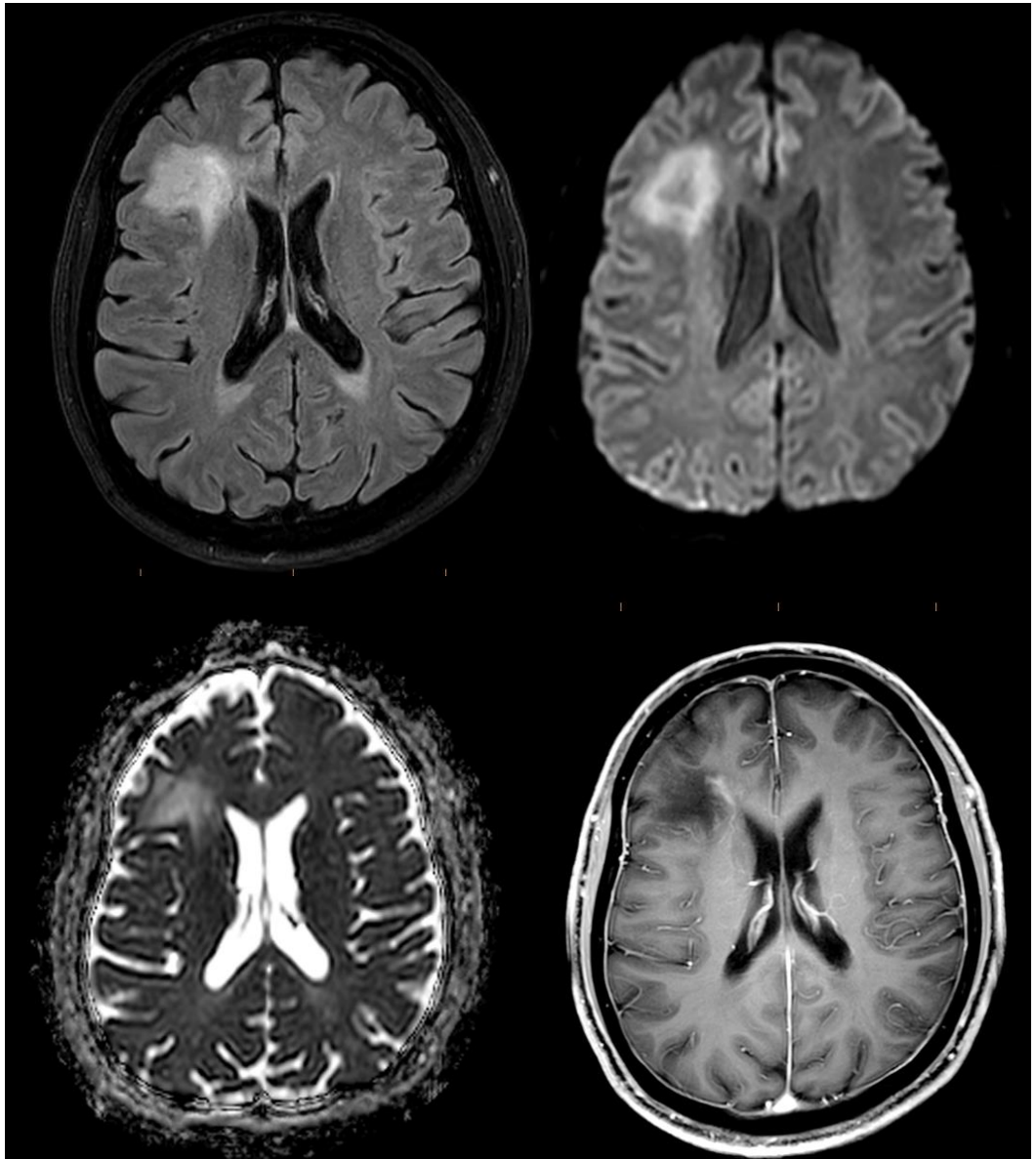
Imaging

-incremento del carico lesionale T2

-rigonfiamento cerebrale

-potenziamento prevalentemente periferico

-effetto massa



Leucoencefalopatia HIV relata

- Sindrome caratterizzata da alterazioni cognitive, comportamentali e motorie secondarie ad effetto diretto di HIV sull'encefalo
- 15-20% dei pazienti con AIDS

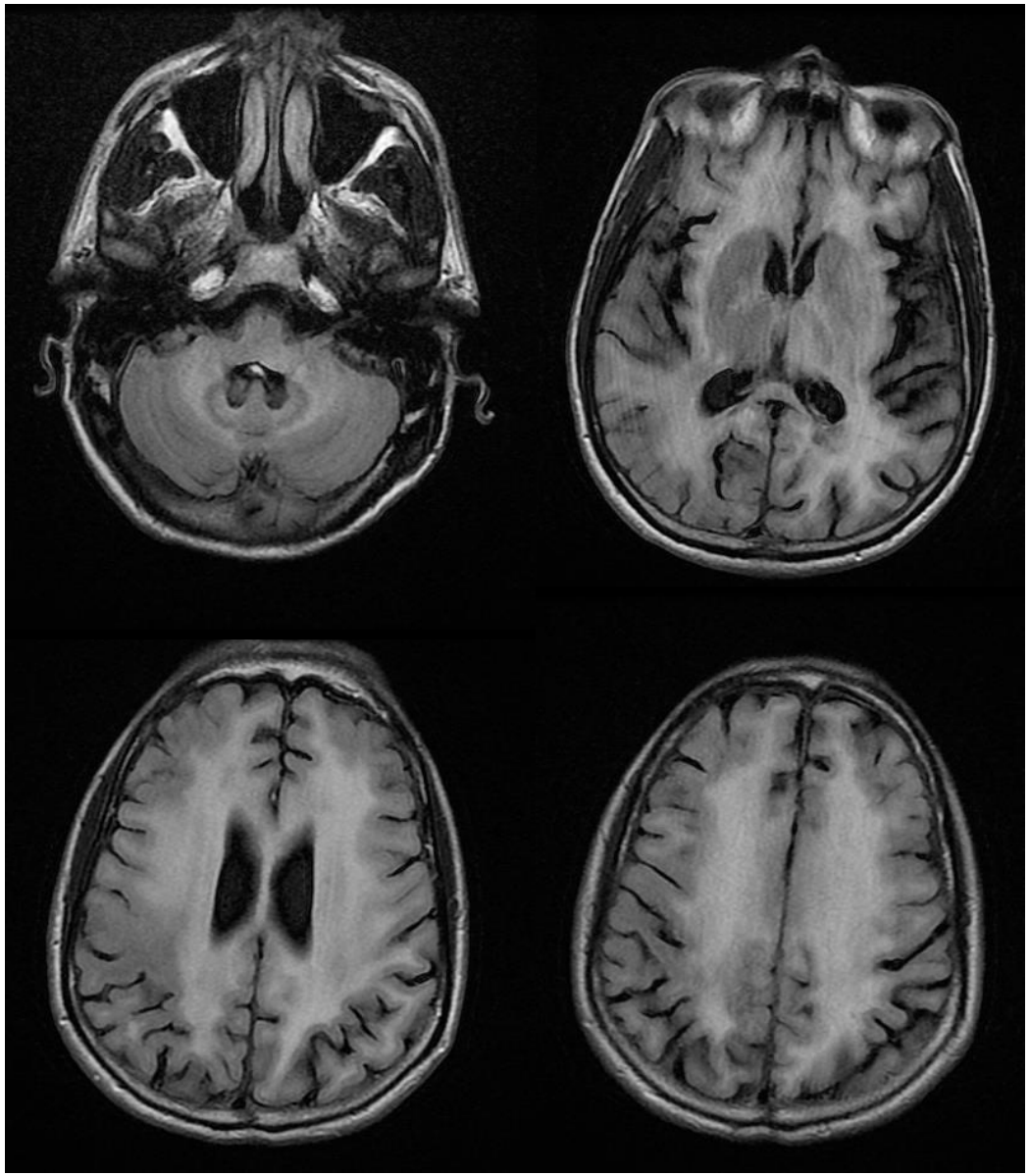
Imaging

-Atrofia diffusa

-Diffusa e simmetrica iperintensità del segnale T2 a livello della sostanza bianca profondo e periventricolare

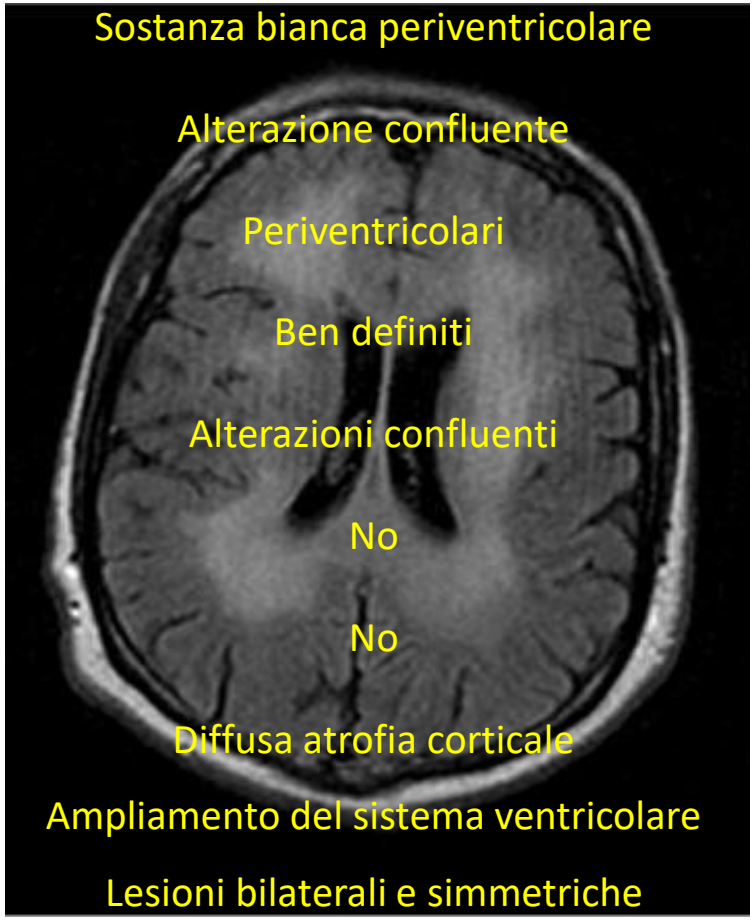
-No effetto massa

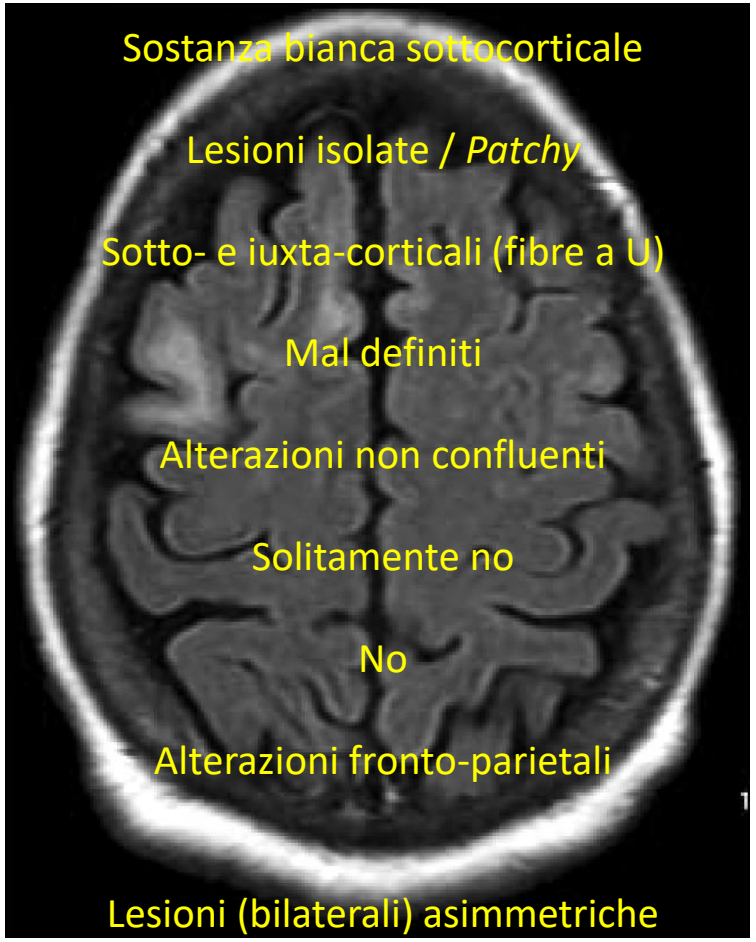
-No potenziamento



Leucoencefalopatia HIV vs PML

Leucoencefalopatia HIV relata		Leucoencefalopatia multifocale progressiva
Sostanza bianca periventricolare	SEDE	Sostanza bianca sottocorticale
Alterazione confluente	PATTERN	Lesioni isolate / *Patchy*
Periventricolari	SEDE LESIONI	Sotto- e iuxta-corticali (fibre a U)
Ben definiti	ESTENSIONI	Mal definiti
Alterazioni confluenti	ESTENSIONE	Alterazioni non confluenti
No	POTENZIAMENTO	Solitamente no
No	EFFETTO MASSA	No
Diffusa atrofia corticale Ampliamento del sistema ventricolare	HALLMARK	Alterazioni fronto-parietali
Lesioni bilaterali e simmetriche		Lesioni (bilaterali) asimmetriche

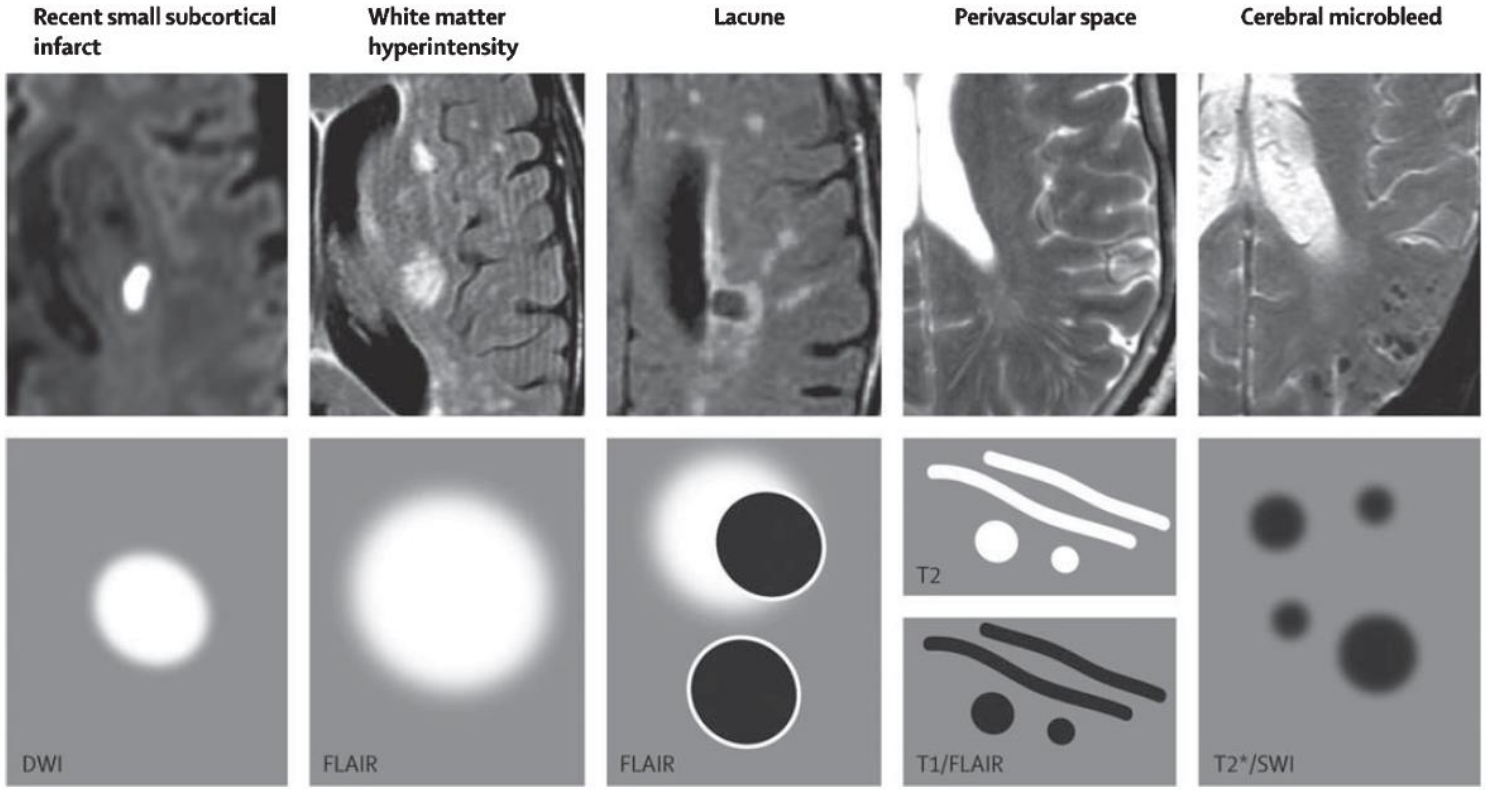




Malattie vascolari

Malattie dei piccoli vasi

Arteriolosclerosi



	Recent small subcortical infarct	White matter hyperintensity	Lacune	Perivascular space	Cerebral microbleed
Usual diameter	≤20 mm	Variable	3–15 mm	≤2 mm	≤10 mm
Comment	Best identified on DWI	Located in white matter	Usually have hyperintense rim	Most linear without hyperintense rim	Detected on GRE seq., round or ovoid, blooming
DWI	↑	↔	↔/(↓)	↔	↔
FLAIR	↑	↑	↓	↓	↔
T2	↑	↑	↑	↑	↔
T1	↓	↔/(↓)	↓	↓	↔
T2*-weighted GRE	↔	↑	↔ (↓ if haemorrhage)	↔	↓↓

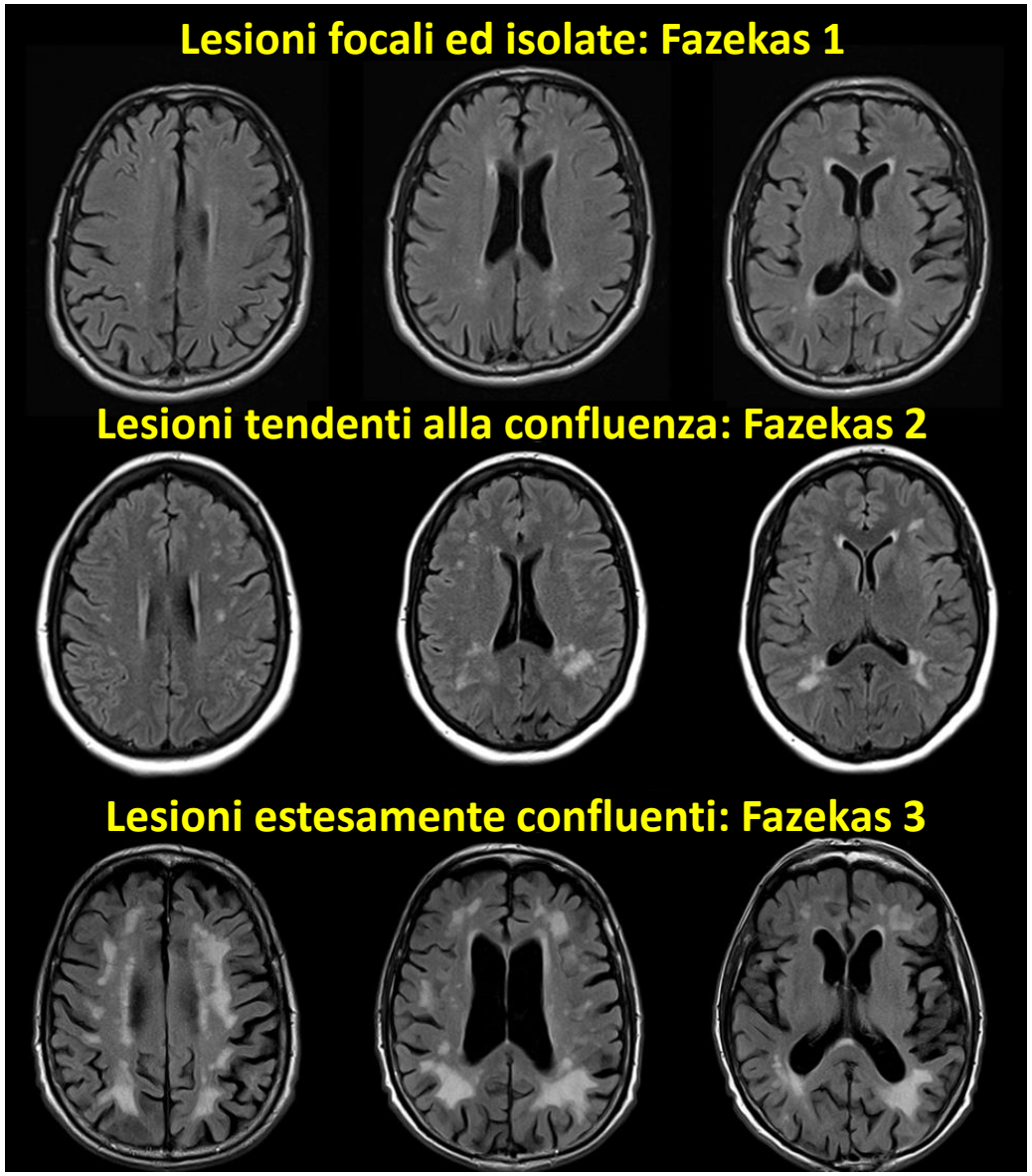
Journal of Alzheimer's Disease 62 (2018) 1417–1441
DOI 10.3233/JAD-170803
IOS Press

Review

Update on Vascular Cognitive Impairment Associated with Subcortical Small-Vessel Disease

Anders Wallin[a,1,*], Gustavo C. Román[b,c,1], Margaret Esiri[d], Petronella Kettunen[a,e], Johan Svensson[f], George P. Paraskevas[g] and Elisabeth Kapaki[g]

Arteriolosclerosi



Pathologic correlates of incidental MRI white matter signal hyperintensities

F. Fazekas, MD; R. Kleinert, MD; H. Offenbacher, MD; R. Schmidt, MD; G. Kleinert, MD; F. Payer, MD; H. Radner, MD; and H. Lechner, MD

NEUROLOGY 1993;43:1683-1689

	Sostanza bianca periventricolare	Sostanza bianca profonda
0	Assenti	Assenti
1	Esile cappuccio periventricolare	Isolati e millimetrici foci
2	Alone periventricolare	Aree tendenti alla confluenza
3	Alterazione periventricolare estesa alla sostanza bianca profonda	Aree ampiamente confluenti

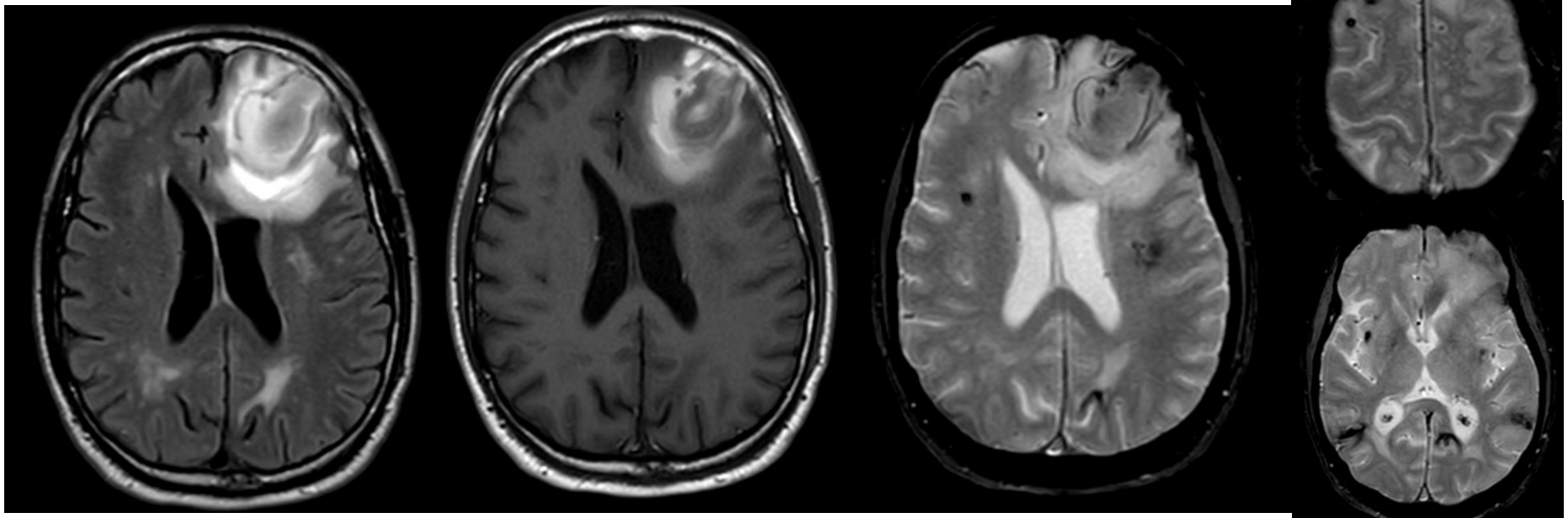
Combinazione di demielinizzazione, ependimite granulomatosa, gliosi e patologia microcircolatoria ischemica cronica

Patologia microcircolatoria ischemica cronica (possibile applicazione nei Pz con demenze)

Angiopatia amiloide cerebrale

- Malattia dei piccoli vasi **sporadica** o **ereditaria**
- Istologia:
  - -depositi di **β-amiloide** nelle pareti delle a. leptomeningee e corticali

- Imaging:
  - -Macroemorragie lobari (frontali e occipitali) >5mm
  - -Microemorragie sottocorticali (pattern posteriore) <5mm
  - -Leucoaraiosi sovratentoriale
  - -Siderosi superficiale e/o ESA



Topical Review

Section Editors: Jean-Marc Olivot, MD, PhD, and Chelsea Kidwell, MD

Stroke 2018

Diagnosis of Cerebral Amyloid Angiopathy
Evolution of the Boston Criteria

Steven M. Greenberg, MD, PhD; Andreas Charidimou, MD, PhD

Table 1. Modified Boston Criteria for CAA

Definite CAA
Full postmortem examination demonstrating:
Lobar, cortical, or cortical–subcortical hemorrhage
Severe CAA with vasculopathy
Absence of other diagnostic lesion
Probable CAA with supporting pathology
Clinical data and pathological tissue (evacuated hematoma or cortical biopsy) demonstrating:
Lobar, cortical, or cortical–subcortical hemorrhage (including ICH, CMB, or cSS)
Some degree of CAA in specimen
Absence of other diagnostic lesion
Probable CAA
Clinical data and MRI or CT demonstrating:
Multiple hemorrhages (ICH, CMB) restricted to lobar, cortical, or cortical–subcortical regions (cerebellar hemorrhage allowed), or single lobar, cortical, or cortical–subcortical hemorrhage and cSS (focal or disseminated)
Age ≥55 y
Absence of other cause of hemorrhage*
Possible CAA
Clinical data and MRI or CT demonstrating:
Single lobar, cortical, or cortical–subcortical ICH, CMB, or cSS (focal or disseminated)
Age ≥55 y
Absence of other cause of hemorrhage*

CADASIL

Cerebral Autosomal-dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy

- Malattia dei piccoli vasi **ereditaria**: ***NOTCH3*** (Cr 19)
- Esordio tipico 3ª-4ª decade, progressione "a gradini"
- Istologia:

-depositi di **granular osmiophilic material** (GOM) a. perforanti lunghe e leptomeningee, perdita delle cellule muscolari lisce

-aree di demielinizzazione

-infarti sottocorticali

REGIONI TEMPORALI ANTERIORI

CAPSULE ESTERNE

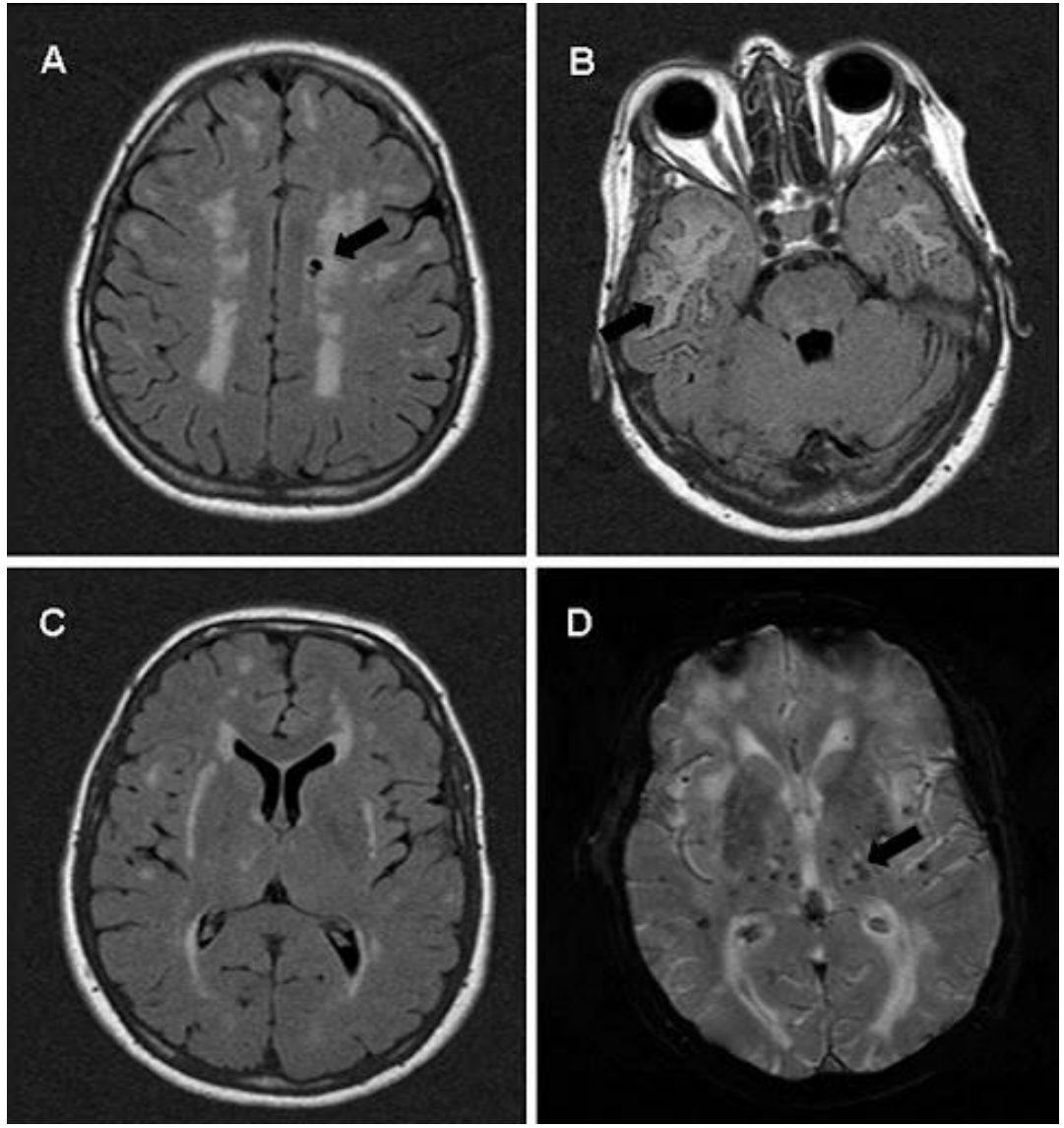
- Imaging:

-vaste aree di demielinizzazione **simmetriche** e **confluenti**

-da **piccoli infarti SB sottocorticale** ad infarti lacunari profondi

(talamo, nuclei della base, capsula interna)

-possibile coinvolgimento delle fibre ad U



van den Boom et al, AJNR 2006

Sindrome di Susac

- Malattia dei piccoli vasi **rara**, patogenesi **autoimmune** (**F>M**)
- Sordità neurosensoriale, perdita della vista, encefalopatia
- Istologia: **Anticorpi antiendotelio**

  -danno endoteliale

  -piccoli trombi

  -occlusione del lume

COCLEA
RETINA
ENCEFALO

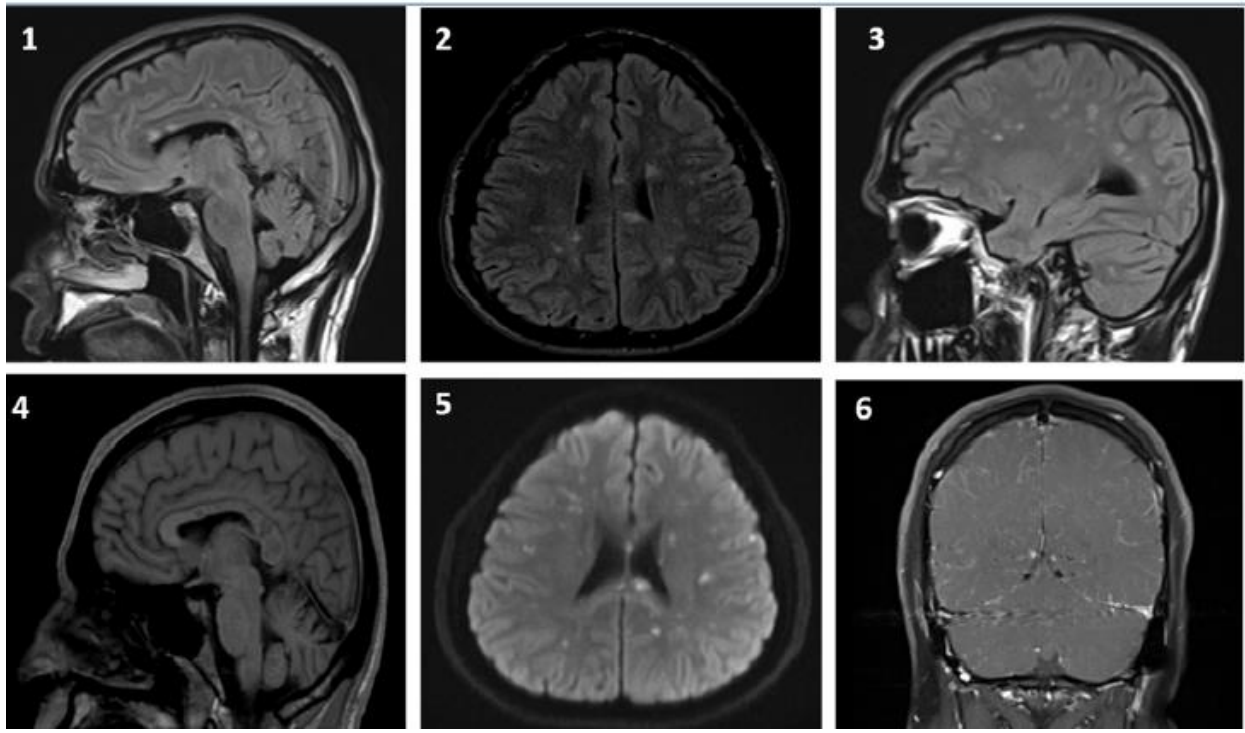
Wilf-Yarkoni *et al. BMC Neurology* (2020) 20:332
https://doi.org/10.1186/s12883-020-01892-0

BMC Neurology

RESEARCH ARTICLE Open Access

Increased incidence of Susac syndrome: a case series study

A. Wilf-Yarkoni[1*], O. Elkayam[2,3], O. Aizenstein[4], Y. Oron[5], V. Furer[2,3], D. Zur[3,6], M. Goldstein[3,6], D. Barequet[3,6], H. Hallevi[7], A. Karni[3,7,8], Z. Habot-Wilner[3,6] and K. Regev[7]



- Imaging:

-***Snowball lesions*** piccole e multiple lesioni SB e **corpo calloso**

-**diffusione ristretta** (fasi acute)

-Iposegnale T1 corpo calloso: aspetto ***punched out***

-T1postGD: potenziamento leptomeningeo e parenchimale

Malattie Tossico Metaboliche

Malattie tossico-metaboliche

- CLINICA: stato confusionale acuto e delirio
- PATTERN NEURORADIOLOGICI

-distribuzione **bilaterale** e **simmetrica**

-**diffusione ristretta**

-non effetto massa, né potenziamento contrastografico (solitamente)

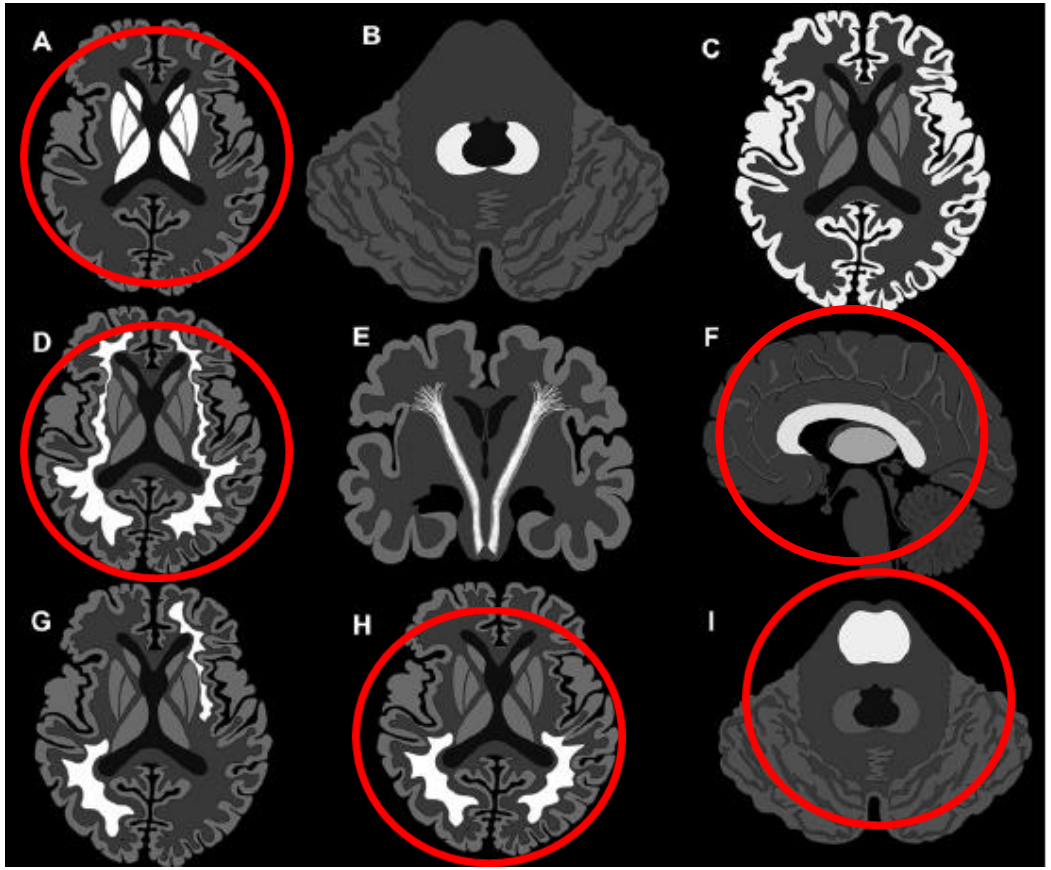
- SEDI: sostanza grigia corticale e profonda (nuclei della base), talami, **sostanza bianca periventricolare**, **corpo calloso**, **ponte** sensibili al **DANNO ECCITOTOSSICO**
- PROGNOSI: peggiore se interessata la sostanza grigia

PATTERN → PIÙ MALATTIE METABOLICHE

MALATTIA METABOLICA → PIÙ PATTERN

Imaging Patterns of Toxic and Metabolic Brain Disorders

De Oliveira et al 2019 - Radiographics



Sindromi da demielinizzazione osmotica

- Ha sostituito il termine **mielinolisi pontina** centrale perché nella patologia possono essere colpite strutture extra-pontine (**mielinolisi extra-pontina**)

- Sindrome da demielinizzazione osmotica acuta (rapidi cambiamenti dell'osmosi tipicamente successivi a rapida correzione dell'iponatriemia)

- **Oligodendrociti** sensibili alla **rapida correzione** dell' **iponatriemia** (alcolismo cronico, malnutrizione)

- Decorso bifasico

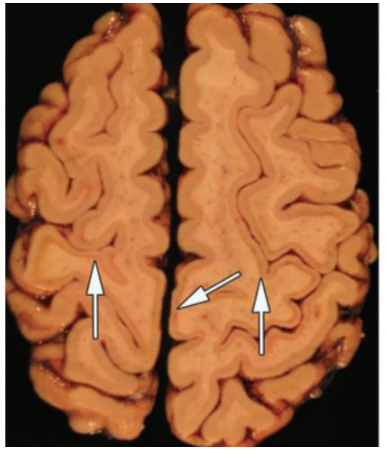
- Istologia:

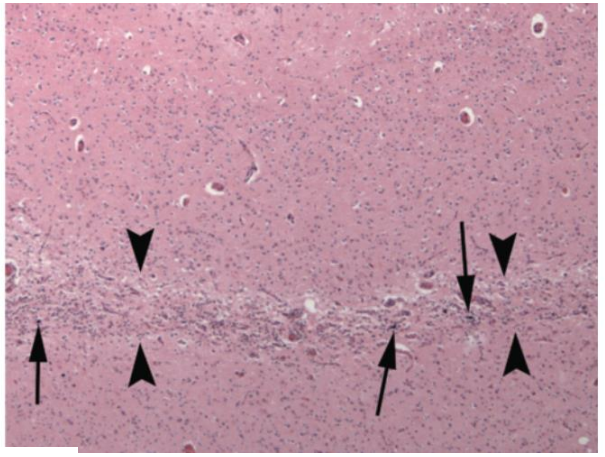
  -demielinizzazione 'pura' (non infiammatoria)

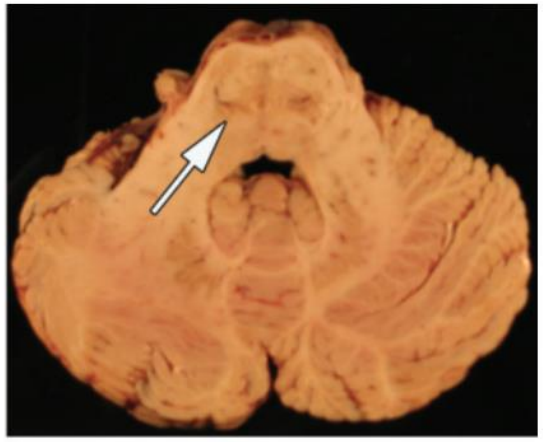
RadioGraphics 2009; 29:933–938 • DOI: 10.1148/rg.293085151 •

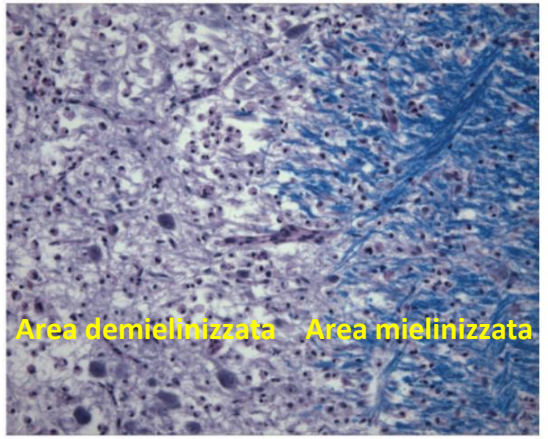
Osmotic Demyelination Syndrome[1]

Stephanie A. Howard, MD • Justine A. Barletta, MD • Roman A. Klufas, MD • Ali Saad, MD • Umberto De Girolami, MD









Sindromi da demielinizzazione osmotica

- Imaging:

-**pontina** (50%): coinvolgimento **simmetrico** regioni **centrali** del **ponte**;

-risparmio regioni periferiche e tratti cortico-spinali (pattern a **tridente**)

-**extrapontina**: coinvolgimento simmetrico nuclei della base, talami, peduncoli cerebrali, SB sottocorticale, necrosi laminare

AJNR Am J Neuroradiol 25:210–213, February 2004

Case Report

Early Diagnosis of Central Pontine Myelinolysis with Diffusion-Weighted Imaging

Kimberly A. Ruzek, Norbert G. Campeau, and Gary M. Miller

DWI + entro 24 h

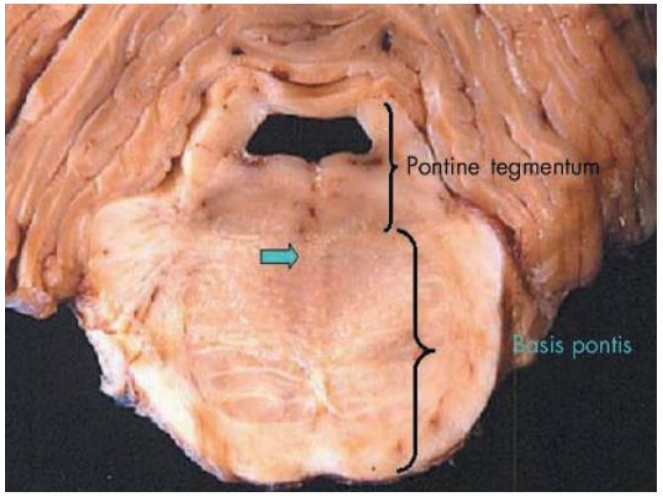
ELSEVIER

Seminars in Ultrasound, CT and MRI

Volume 35, Issue 2, April 2014, Pages 153-159

Osmotic Demyelination Syndrome: Central Pontine Myelinolysis and Extrapontine Myelinolysis

Anthony M. Alleman MD, MPH

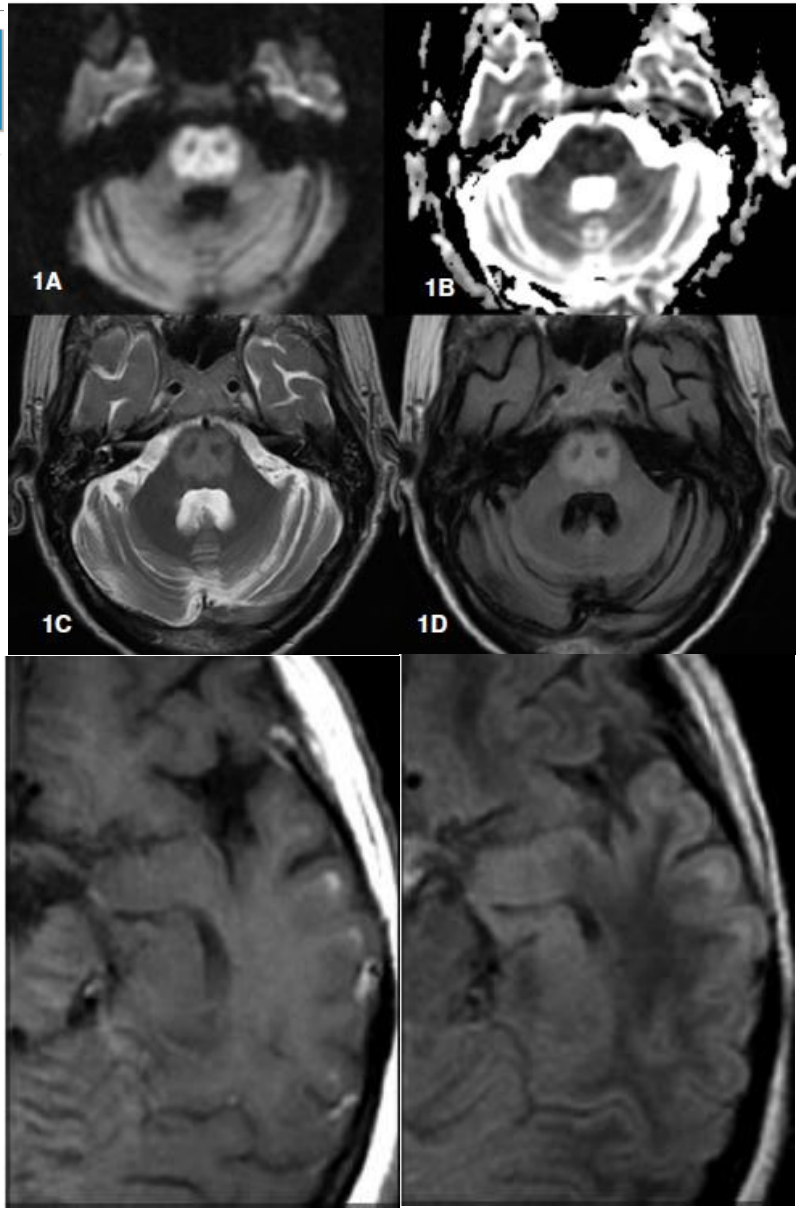


The British Journal of Radiology, 85 (2012), e87–e90

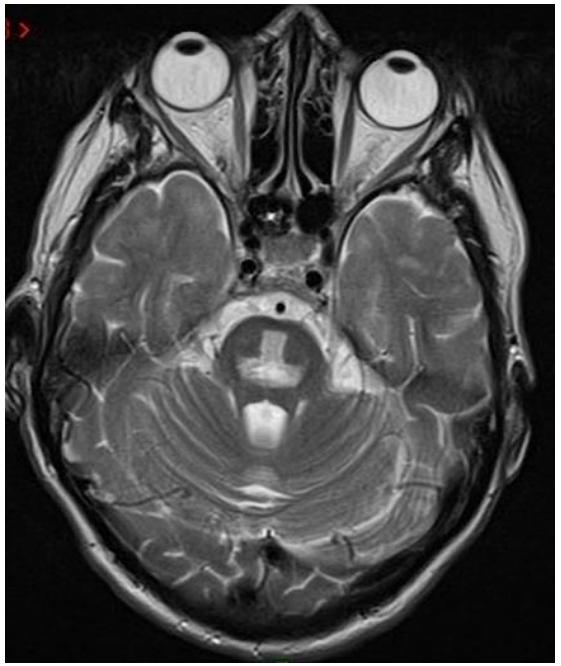
CASE REPORT

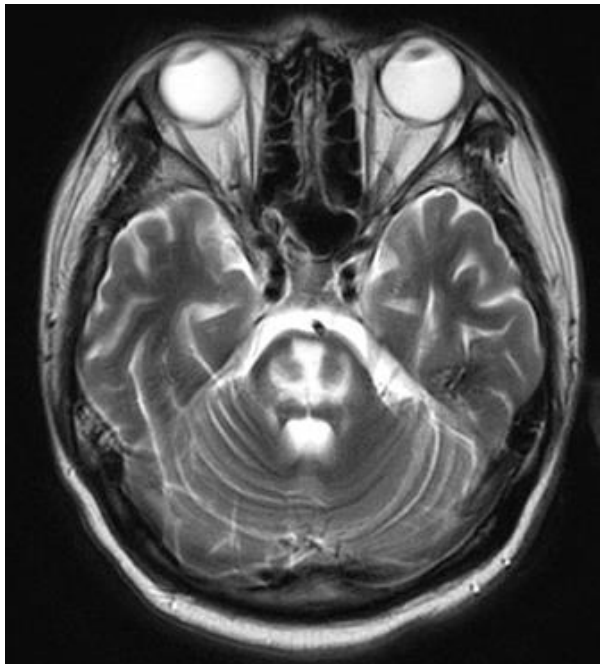
MRI findings of corticosubcortical lesions in osmotic myelinolysis: report of two cases

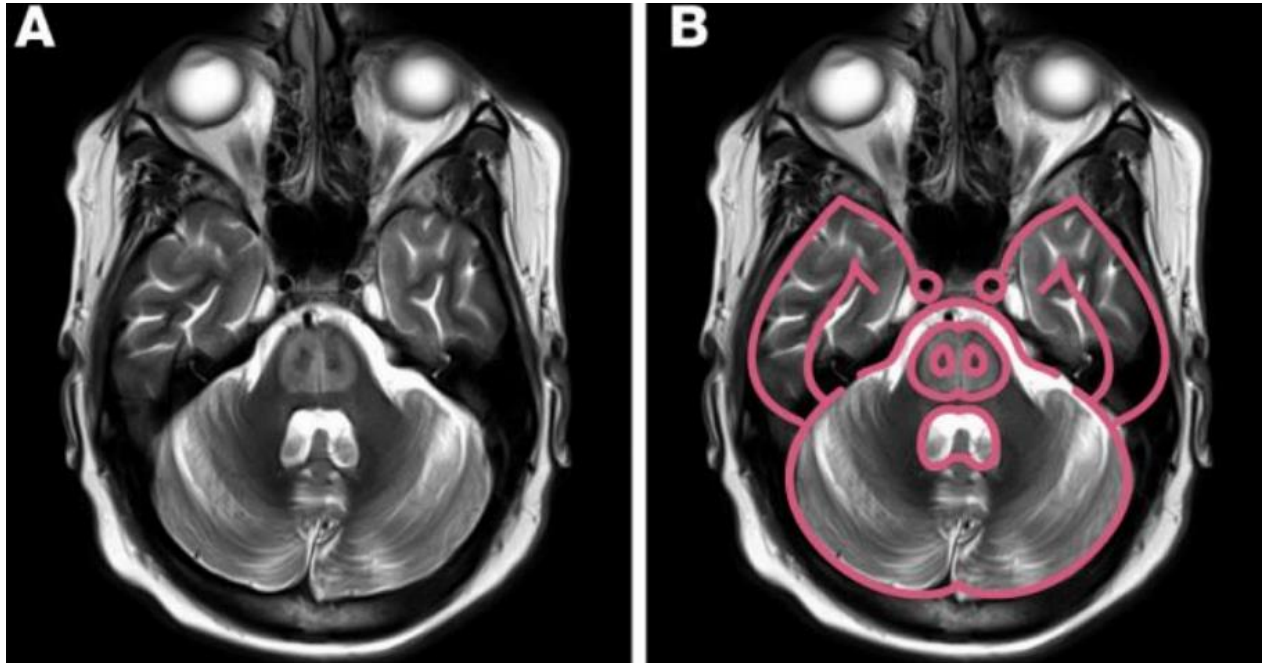
[1]Y TATEWAKI, MD, [2]K KATO, MD, [3]Y TANABE, MD, PhD and [1]S TAKAHASHI, MD, PhD



Mielinolisi pontina







Mexican hat sign

Trident or Maserati sign

Piglet sign

CLINICAL SHORT COMMUNICATION | VOLUME 373, P268-273,
FEBRUARY 15, 2017

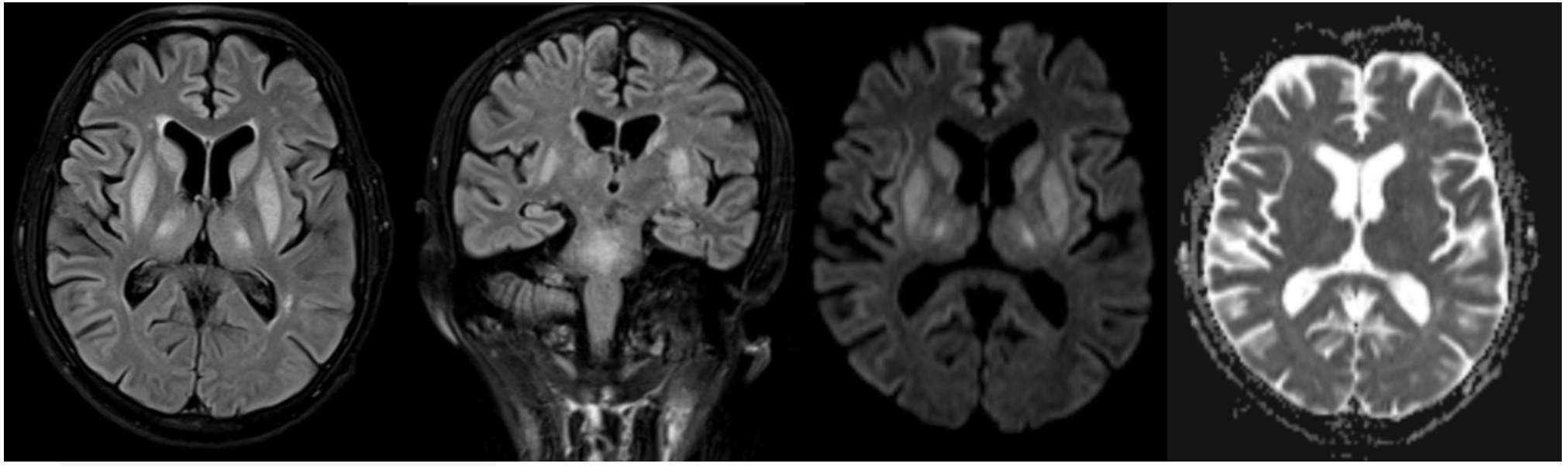
Purchase

Temporal evolution of the trident and piglet signs of osmotic demyelination syndrome

Shin C. Beh

Published: January 07, 2017 • DOI: https://doi.org/10.1016/j.jns.2017.01.024 • Check for updates

Mielinolisi extra-pontina



Central Pontine and Extra Pontine Myelinolysis

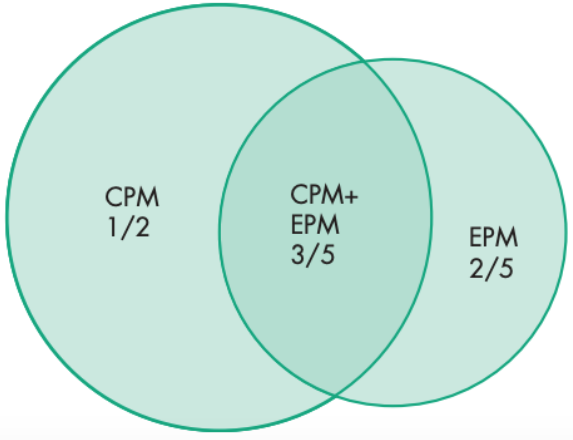
In Eurorad 2017
Priyank S Chatra
10.1594/EURORAD/CASE.14449

Table 1 Lesions of central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) (in descending order of frequency)[3]

- Pons
- Cerebellum
- Lateral geniculate body
- External capsule
- Extreme capsule
- Hippocampus
- Putamen
- Cerebral cortex/subcortex
- Thalamus
- Caudate nucleus

The following 10% or less:

- Claustrum
- Internal capsule
- Midbrain
- Internal medullary lamella
- Mamillary body
- Medulla oblongata



CENTRAL PONTINE AND EXTRA-PONTINE MYELINOLYSIS: THE OSMOTIC DEMYELINATION SYNDROMES

R J Martin

J Neurol Neurosurg Psychiatry 2004;**75**(Suppl III):iii22–iii28. doi: 10.1136/jnnp.2004.045906

PRES

Sindrome dell' encefalopatia posteriore reversibile

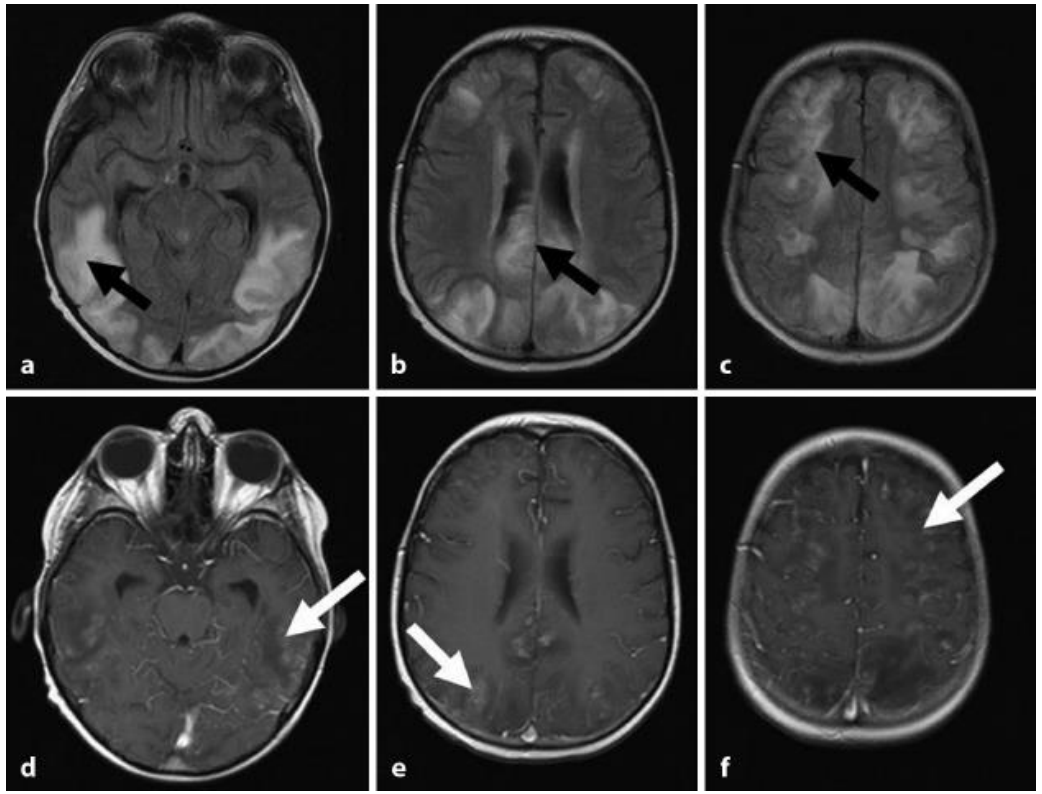
➢Imaging:

-edema cortico-sottocorticale **bilaterale** ed **simmetrico** delle regioni **parieto-occipitali** e **frontali posteriori**

-pattern: **oloemisferico**, del **solco frontale superiore**, **parieto-occipitale**, coinvolgimento di altre regioni encefaliche (lobi temporali, talami, cervelletto, tronco encefalico, nuclei della base). Unilaterale

-T2* emorragie ed ESA (15%);

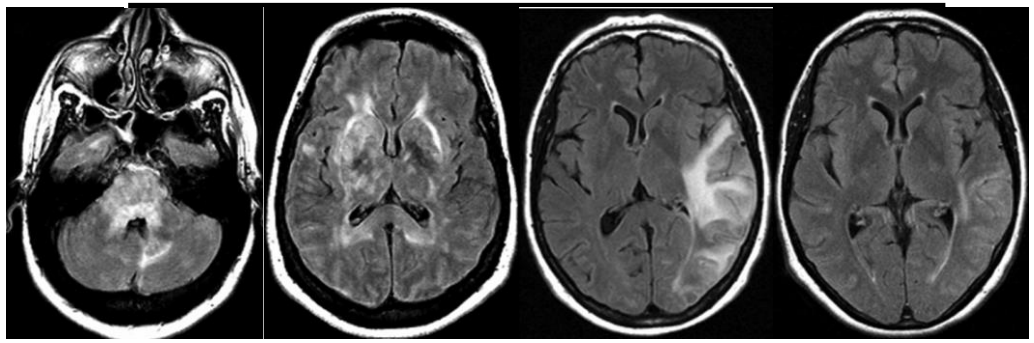
-potenziamento disomogeneo



Neuroradiology • Original Research

Alexander M. McKinney[1,2]
James Short[1]
Charles L. Truwit[1]
Zeke J. McKinney[1]
Osman S. Kozak[1]
Karen S. SantaCruz[1]
Mehmet Teksam[1]

Posterior Reversible Encephalopathy Syndrome: Incidence of Atypical Regions of Involvement and Imaging Findings



Clin Neuroradiol (2015) 25:161–171
DOI 10.1007/s00062-014-0293-7

ORIGINAL ARTICLE

Posterior Reversible Encephalopathy Syndrome: The Spectrum of MR Imaging Patterns

O. Kastrup · M. Schlamann · C. Moenninghoff · M. Forsting · S. Goericke

NON SEMPRE POSTERIORE!

NON SEMPRE REVERSIBILE!

Leucoencefalopatia da methotrexate

- Esordio **acuto**, da 2 a 14 giorni dopo la somministrazione
- Pz **pediatrici**, somministrazione **intratecale**, **alte dosi**
- Pattern:

-encefalopatia tossica

-encefalopatia necrotizzante disseminata

- Imaging:

-**diffusione ristretta** ed ipersegnale T2/FLAIR **centri semiovali** e SB periventricolare (risparmiate le fibre ad U)

contrast enhancement **nodulare** o **periferico**; effetto massa.

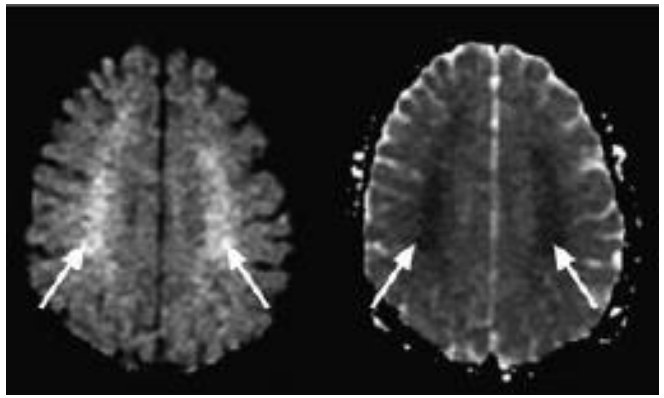
TUMORLIKE PATTERN
ANAMNESI
CLINICA, SOSPETTO CLINICO

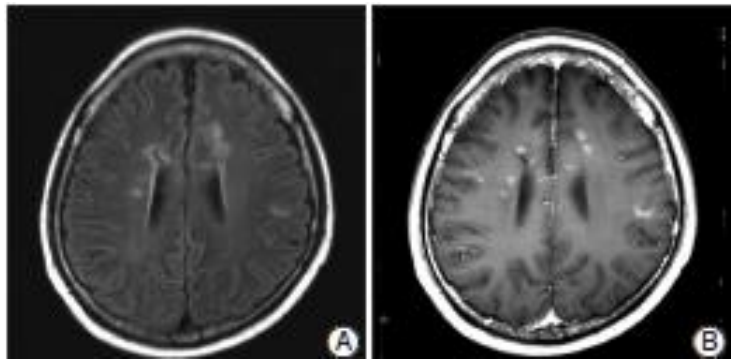
original article

Annals of Oncology
doi:10.1093/annonc/mdm466

Clinical and radiological characteristics of methotrexate-induced acute encephalopathy in pediatric patients with cancer

H. Inaba[1,2], R. B. Khan[3,4], F. H. Laningham[3,5], K. R. Crews[6,7], C.-H. Pui[1,2] & N. C. Daw[1,2*]





J Korean Neurosurg Soc 50 : 304-310, 2011

Clinical Article

Leukoencephalopathy and Disseminated Necrotizing Leukoencephalopathy Following Intrathecal Methotrexate Chemotherapy and Radiation Therapy for Central Nerve System Lymphoma or Leukemia

Ji Yeon Kim, M.D.,[1] Sung Tae Kim, M.D., Ph.D.,[2*] Do-Hyun Nam, Ph.D.,[1] Jung-Il Lee, Ph.D.,[1] Kwan Park, Ph.D.,[1] Doo-Sik Kong, M.D., Ph.D.[1*]
Departments of Neurosurgery,[1] Radiology,[2] Center for Imaging Science, Samsung Medical Center, Samsung Biomedical Research Institute, Sungkyunkwan University School of Medicine, Seoul, Korea

Patologie alcol-relate

Malattia di Marchiafava-Bignami

- Progressiva demielinizzazione del corpo calloso
- Istologia:

-fase acuta demielinizzazione, edema citotossico, infiltrati macrofagici

-fase cronica necrosi cistica del corpo calloso

- CLINICA

-**tipo A**: esordio improvviso, prognosi severa

-**tipo B**: esordio graduale

- Imaging:

-Iperintensità T2 e FLAIR **CORPO CALLOSO** e della SB periventricolare con potenziamento post-Gd (fase acuta)

-Assottigliamento del corpo calloso (fase cronica)

FLAIR 3D

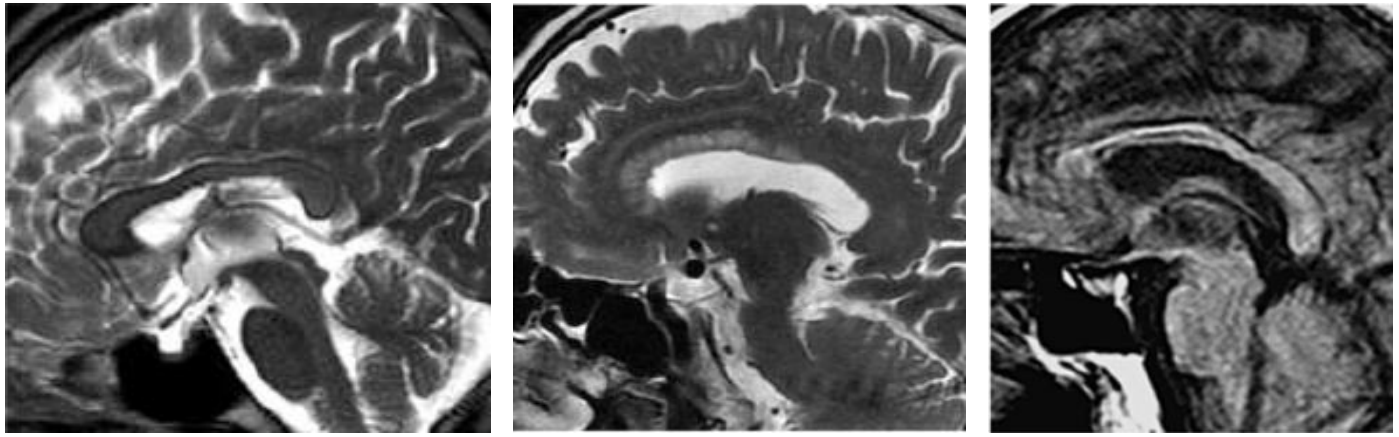
Original Article

Diverse MRI findings and clinical outcomes of acute Marchiafava-Bignami disease

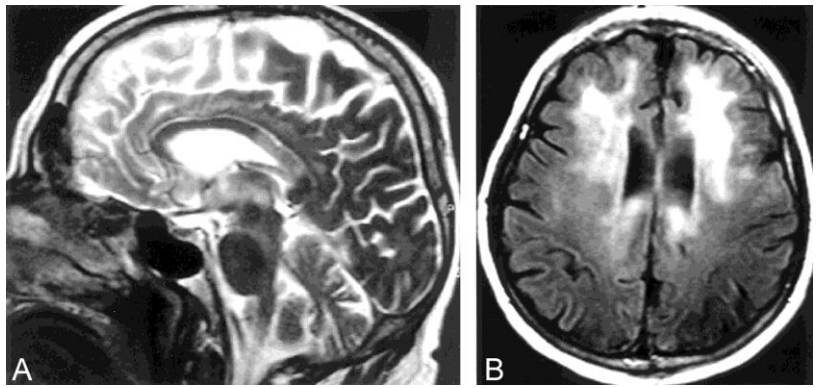
Acta Radiologica
0(0) 1–5

DOI: 10.1177/0284185120943040
journals.sagepub.com/home/acr
SAGE

Wei Li[1,*], Chao Ran[2,*] and Jun Ma[1]



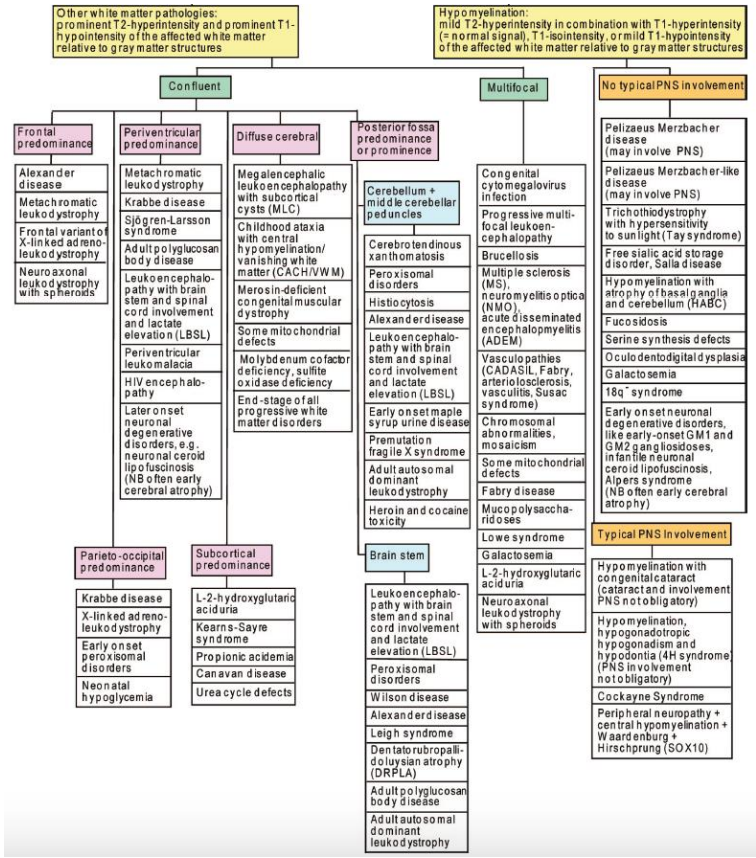
Sandwich sign



Inquadramento neuroradiologico

Criteri RM

- **Sede**: frontale, temporale, occipitale, parietale, tronco encefalico, cervelletto
- **Pattern**: isolato, multifocale, confluente
- **Localizzazione**: sostanza bianca iuxta-corticale, sottocorticale, profonda
- **Identificazione segnale** RM: T1, T2, DWI
- **Pattern di potenziamento**: girale, nodulare, ad anello, ad anello incompeto, periventricolare
- **Imaging avanzato**: spettroscopia RM, Diffusione e Perfusione RM

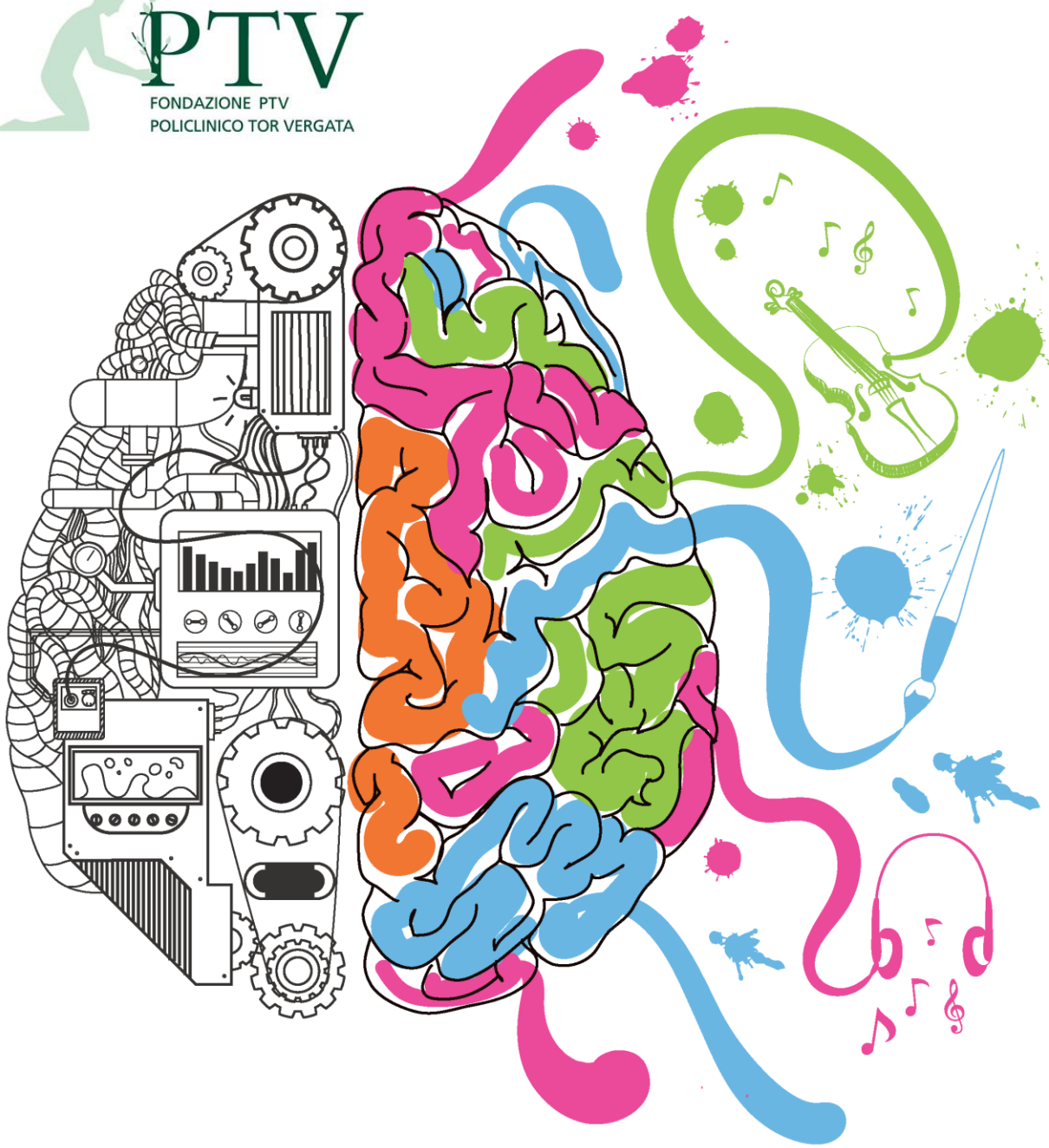


Invited Article:
An MRI-based approach to the diagnosis of white matter disorders

Raphael Schiffmann, MD
Marjo S. van der Knaap, MD, PhD

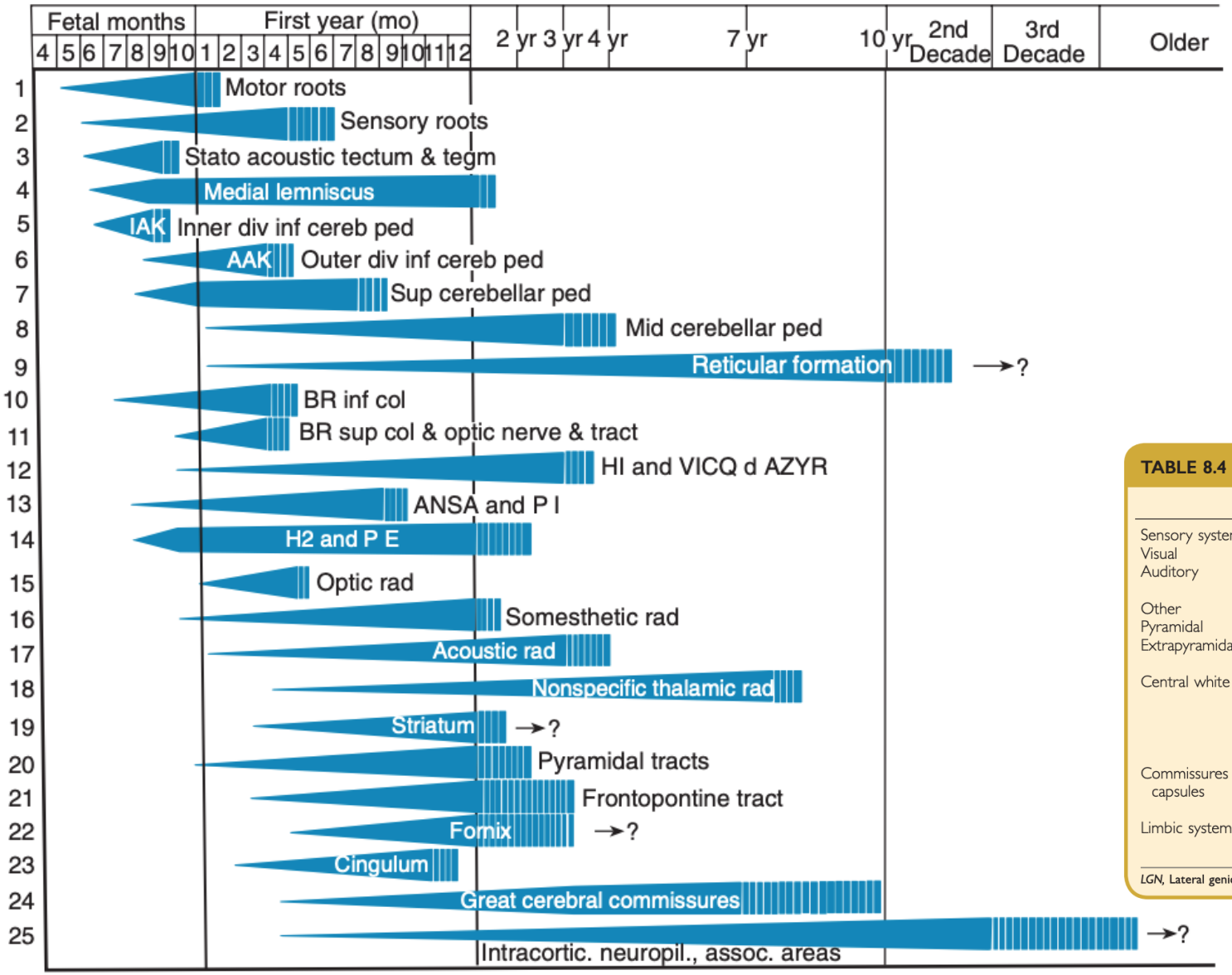
PTV
FONDAZIONE PTV
POLICLINICO TOR VERGATA

Università di Roma
Tor Vergata

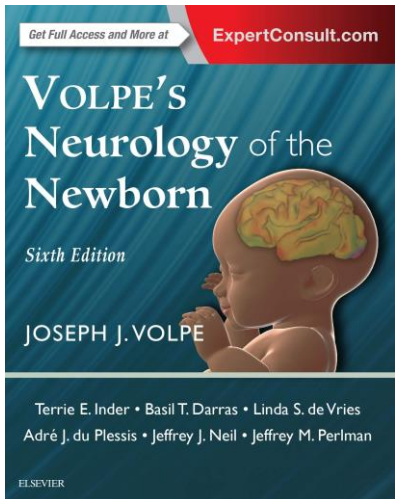


Grazie per l'attenzione

Mielinizzazione



Yakovlev 1967



Chapter 8
Myelination Events

TABLE 8.4 Sequences of Myelination: Sites That Begin to Myelinate *After* Birth

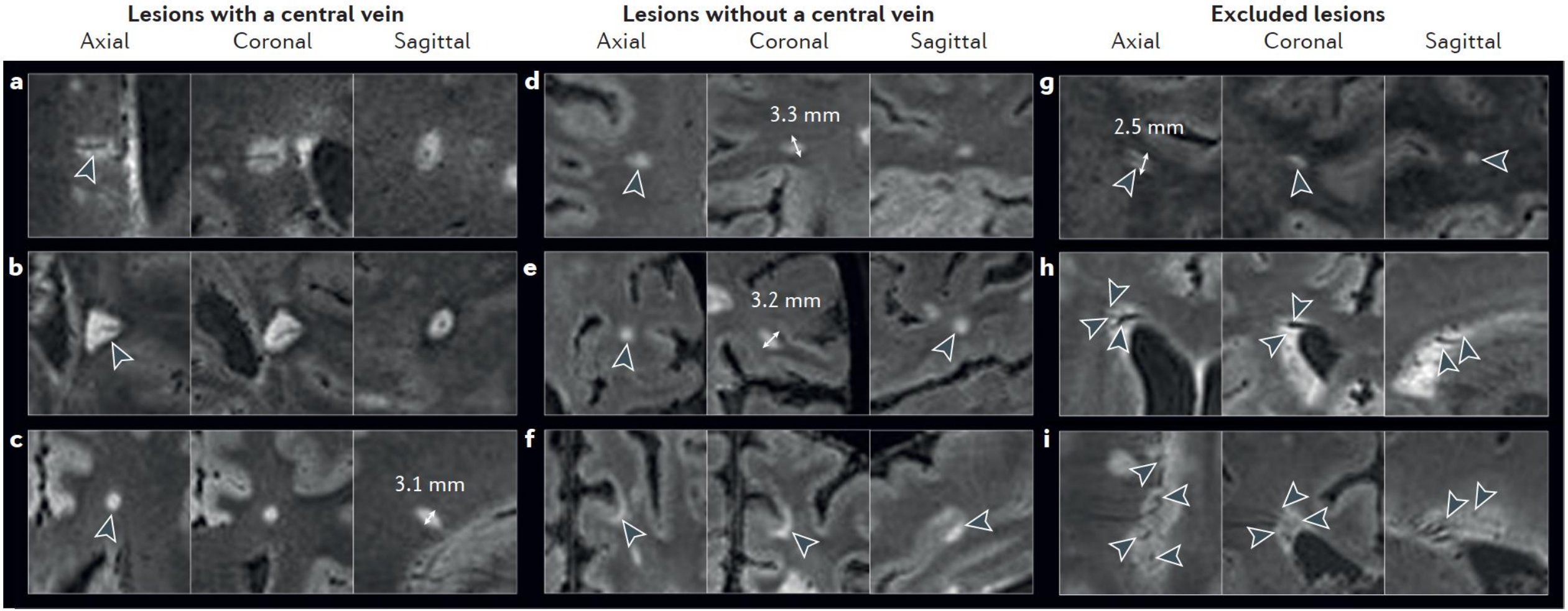
	≤68 WEEKS	70–109 WEEKS	119–142 WEEKS	>144 WEEKS
Sensory system	Optic radiation proximal	SAF calcarine cortex		Stripe of Gennari
Visual	Optic radiation distal			
Auditory	Auditory radiation proximal	Heschl's gyrus		
Other			Lateral olfactory stria	
Pyramidal				
Extrapyramidal system		Lateral crus pedunculi	Medial crus pedunculi	
			Putaminal pencils	
Central white matter		Distal radiation to precentral gyrus	Temporal lobe at LGN	
			Temporal pole	
		Posterior frontal	Frontal pole	
		Posterior parietal	SAF all sites	
		Occipital pole		
Commissures and capsules	Corpus callosum body	Rostrum	Anterior commissure	Extreme capsule
	Splenium	Anterior limb	Inner	
		External capsule		
Limbic system		Cingulum	Mammillothalamic tract	Medial fornix
			Alveus, fimbria	Lateral fornix

LGN, Lateral geniculate nucleus; *SAF*, subcortical association fibers.

FONDAZIONE PTV
POLICLINICO TOR VERGATA

Sclerosi Multipla

Università di Roma Tor Vergata

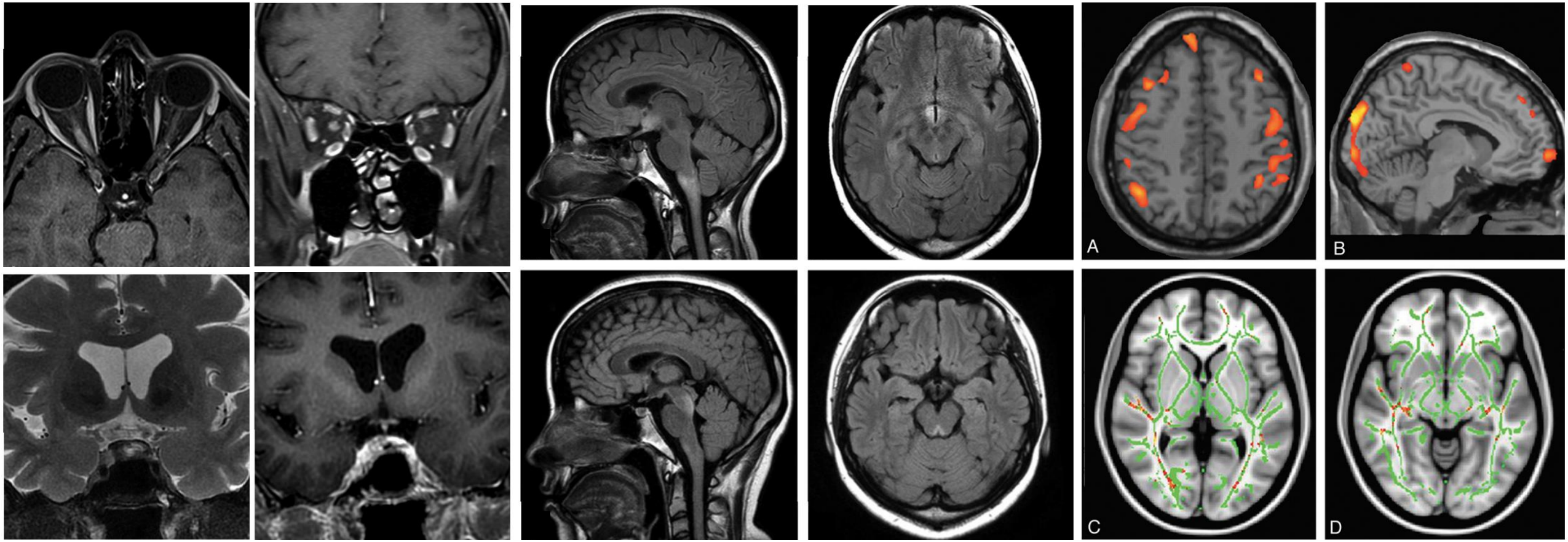


Neuromielite ottica – m. di Devic

Conventional and Advanced Imaging in Neuromyelitis Optica

AJNR 2014

Y. Barnett, I.J. Sutton, M. Ghadiri, L. Masters, R. Zivadinov, and M.H. Barnett

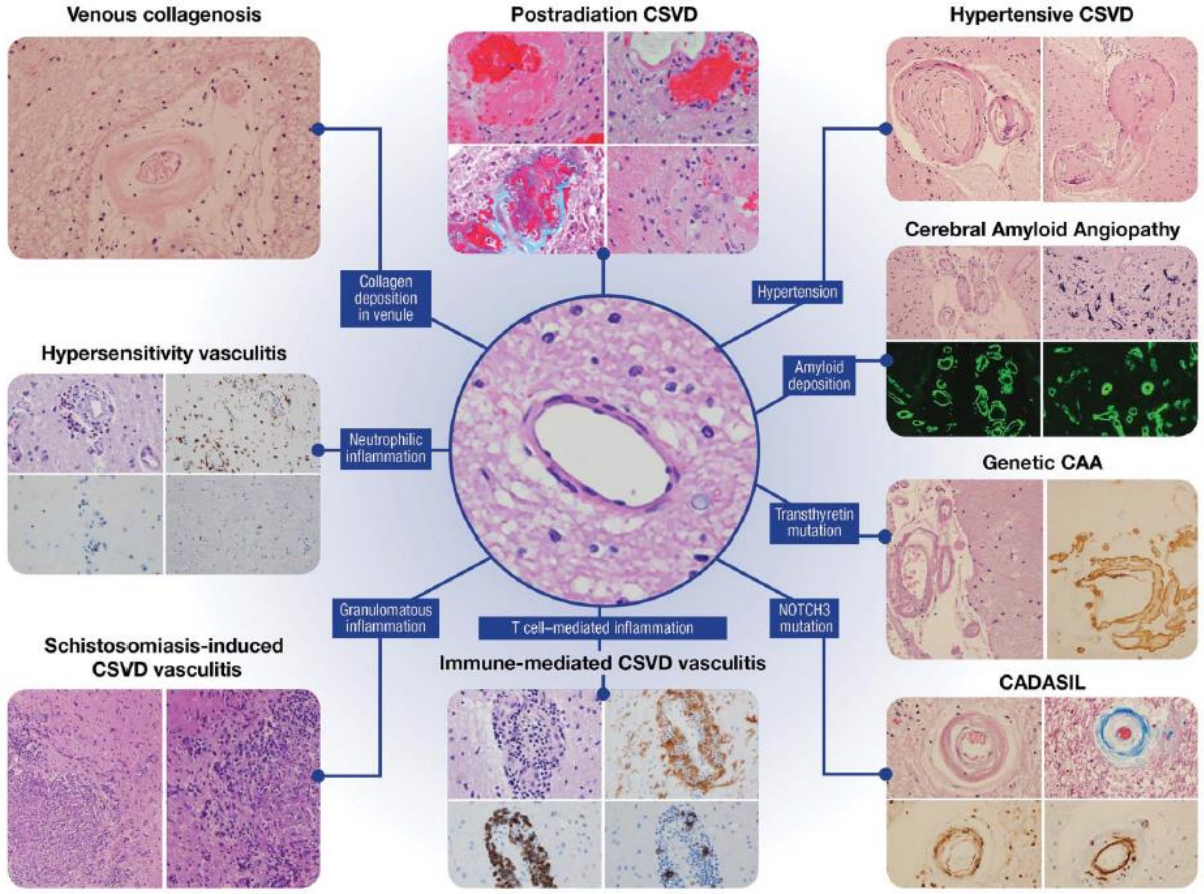


CNS small vessel disease

A clinical review

Rocco J. Cannistraro, MD, Mohammed Badi, MD, Benjamin H. Eidelman, MD, Dennis W. Dickson, MD, Erik H. Middlebrooks, MD, and James F. Meschia, MD

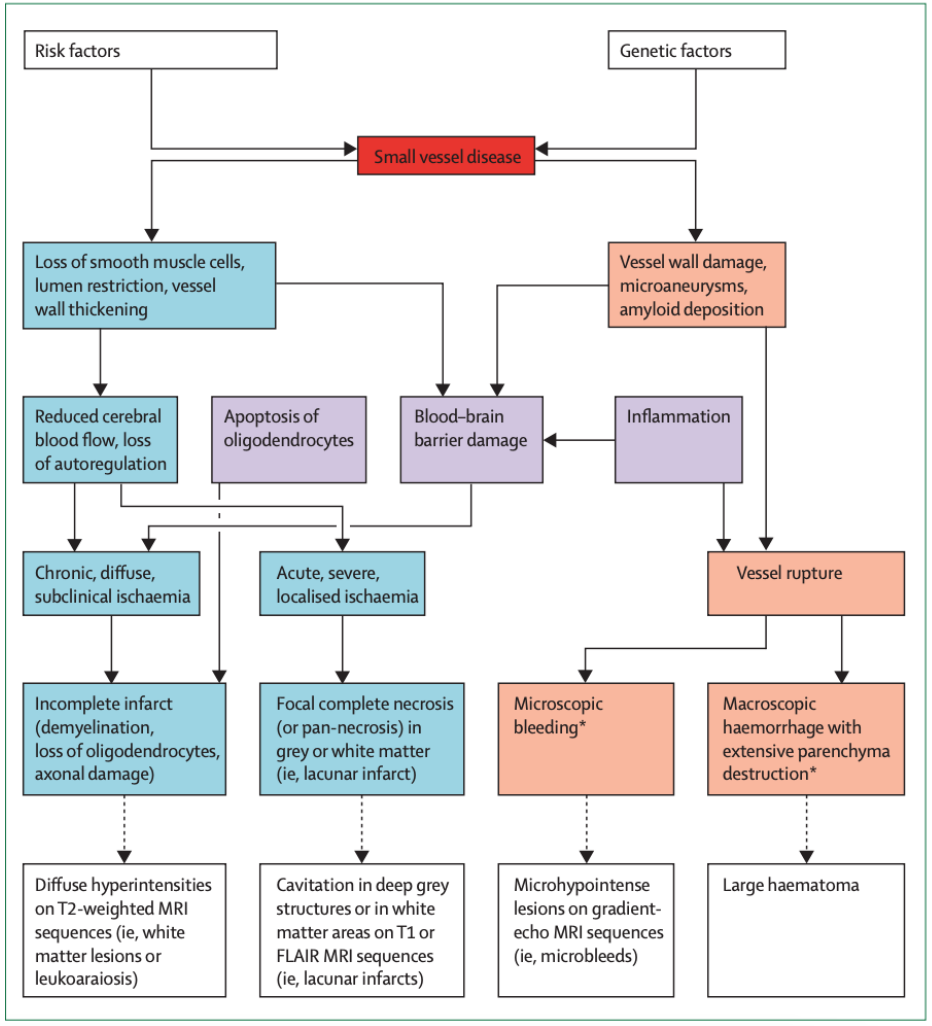
Neurology® 2019;92:1146-1156. doi:10.1212/WNL.0000000000007654



Cerebral small vessel disease: from pathogenesis and clinical characteristics to therapeutic challenges

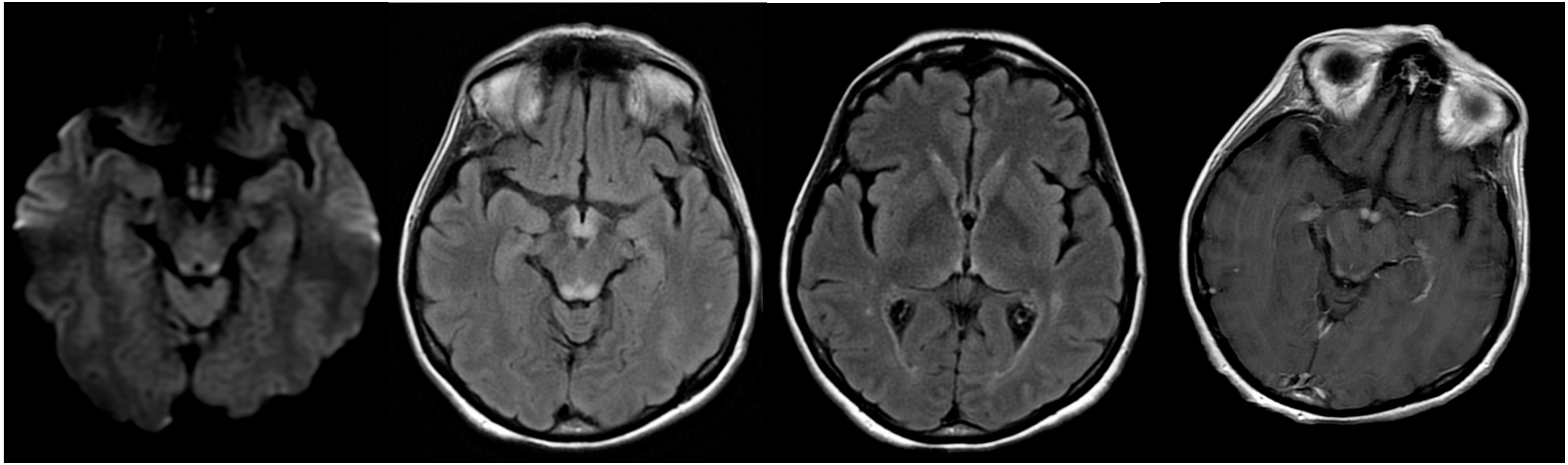
Leonardo Pantoni

Lancet Neurol 2010; 9: 689–701



Deficit di tiamina

Encefalopatia di Wernicke



Corpi mammillari, sostanza **grigia periacqueduttale**, porzioni dorso-mesiali dei **talami**, **collicoli superiori**.

Diffusione ristretta in fase acuta

Potenziamento post-mdc dei corpi mammillari

Deficit di tiamina

Encefalopatia di Wernicke

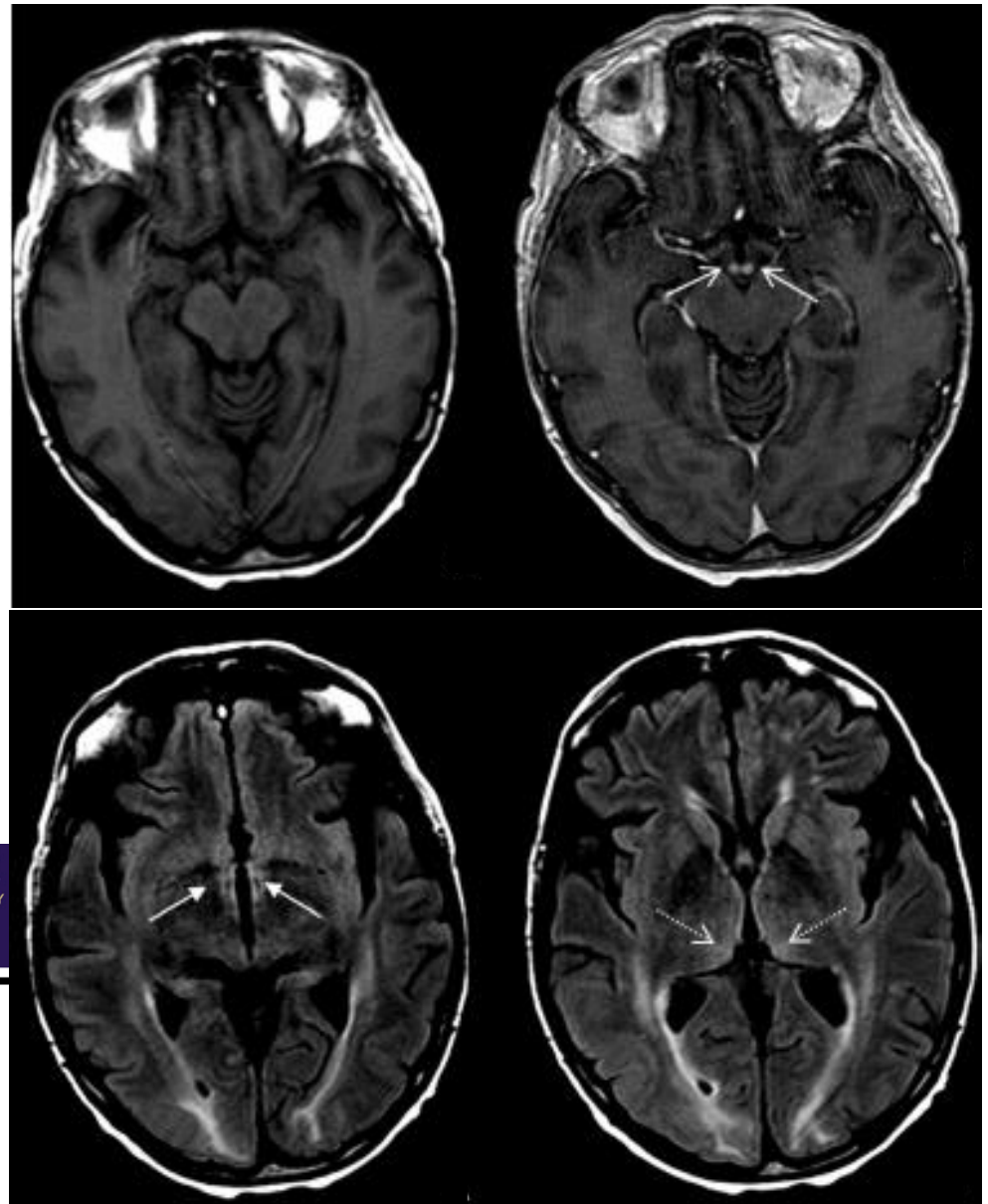
- Alcolica VS Non alcolica
- Patogenesi: perdita omeostasi osmotica delle membrane
- Clinica:
  - -nistagmo/oftalmoplegia
  - -atassia
  - -alterazione dello stato di coscienza

  } 30% dei pz
- Imaging:

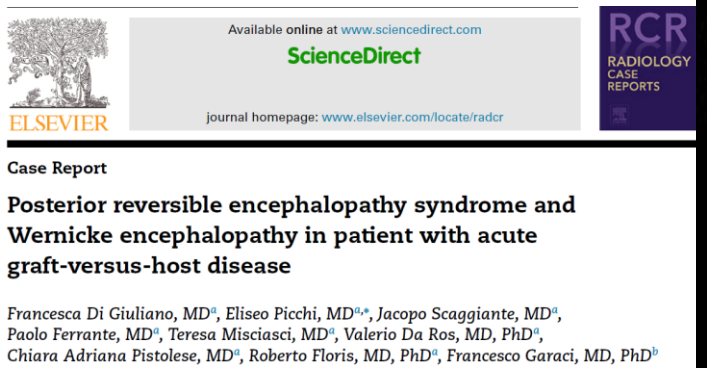
-ipersegnale T2/FLAIR bilaterale e simmetrico nei **corpi mammillari**, SG periacqueduttale, porzioni dorsomesiali dei **talami**, **collicoli superiori**.

-Diffusione ristretta fase acuta

-sedi "atipiche" ipersegnale T2/FLAIR cervelletto, nuclei tronco encefalici, porzioni dorsali del bulbo



T1postGd: enhancement corpi mammillari



Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

RCR RADIOLOGY CASE REPORTS

Case Report

Posterior reversible encephalopathy syndrome and Wernicke encephalopathy in patient with acute graft-versus-host disease

Francesca Di Giuliano, MD[a], Eliseo Picchi, MD[a,], Jacopo Scaggiante, MD[a], Paolo Ferrante, MD[a], Teresa Misciasci, MD[a], Valerio Da Ros, MD, PhD[a], Chiara Adriana Pistolese, MD[a], Roberto Floris, MD, PhD[a], Francesco Garaci, MD, PhD[b]*

FONDAZIONE PTV
POLICLINICO TOR VERGATA

CADASIL

Cerebral Autosomal-dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy

Università di Roma Tor Vergata

The Spatial Distribution of MR Imaging Abnormalities in Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy and Their Relationship to Age and Clinical Features

AJNR Am J Neuroradiol 26:2481–2487, November/December 2005

Sumeet Singhal, Philip Rich, and Hugh S. Markus

BACKGROUND AND PURPOSE: **Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a condition causing recurrent subcortical strokes. MR imaging, which shows focal lacunar infarcts and leukoaraiosis, plays a central role in the diagnosis and evaluation. We studied MR imaging abnormalities in a large prospectively recruited cohort of CADASIL patients to describe the spatial distribution of abnormalities, determine how this distribution alters with age, and identify any correlations with the clinical features of the disease.**

METHODS: **In this study, 112 CADASIL subjects from 64 families were prospectively recruited. MR imaging scans were graded by a single neuroradiologist, by using the modified Scheltens scale, to quantify the severity of high-signal-intensity changes in different brain regions.**

RESULTS: **Lesion load increased progressively with age. Scores were maximal in the frontal, parietal, and anterior temporal cortex, and the external capsule; intermediate in the pons; and relatively low in the corpus callosum, caudate, globus pallidus, cerebellum, midbrain, and medulla. Anterior temporal pole involvement was common at all ages and, when present, usually confluent, but this was absent in 33% of patients 20–29 years of age. A history of stroke correlated with total Scheltens score and internal capsule and pontine scores. Dementia correlated with total Scheltens score and subcortical white matter score, whereas depression correlated with subcortical white matter score but not total Scheltens score.**

CONCLUSIONS: **There is a characteristic pattern of MR imaging abnormalities in CADASIL that aids in differential diagnosis; however, some characteristic features, such as anterior temporal pole involvement, can be absent. MR imaging lesion load correlated with some clinical features including stroke and dementia, whereas depression is more common in individuals with deep white matter changes.**

ORIGINAL RESEARCH

R. van den Boom
S.A.J. Lesnick Oberstein
A.A. van den Berg-Huysmans
M.D. Ferrari
M.A. van Buchem
J. Haan

Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy: Structural MR Imaging Changes and Apolipoprotein E Genotype

BACKGROUND AND PURPOSE: Apolipoprotein E (apoE) genotype plays an important role in the development, maintenance, and response to injury of the central nervous system. It has been suggested that apoE ϵ4 genotype is a risk factor for several neurologic disorders. We investigated the correlation between the apoE genotype and radiologic data in patients with cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL).

METHODS: T1-weighted, dual fast spin-echo, T2*-weighted gradient echo, and fluid-attenuated inversion recovery MR imaging scans were obtained from 36 CADASIL patients (21–59 years of age). The number of lacunar infarcts and microbleeds and the presence of subcortical lacunar lesions were determined. The amount of white matter hyperintensities was assessed by using semiautomated segmentation software. The relation between the radiologic endophenotype of CADASIL and the apoE genotype was assessed by using a Student *t* test for unpaired data and Fisher exact test.

RESULTS: White matter hyperintensities, lacunar infarcts, microbleeds, and subcortical lacunar lesions were not found to be associated with the presence of an ϵ4 allele.

CONCLUSION: The variability of structural MR imaging lesions in CADASIL is independent of apoE genotype and other processes must underlie the variable natural history of the disease.

CADASIL indipendente dal profilo genotipico delle apoE

Le alterazioni temporali anteriori possono non essere presenti

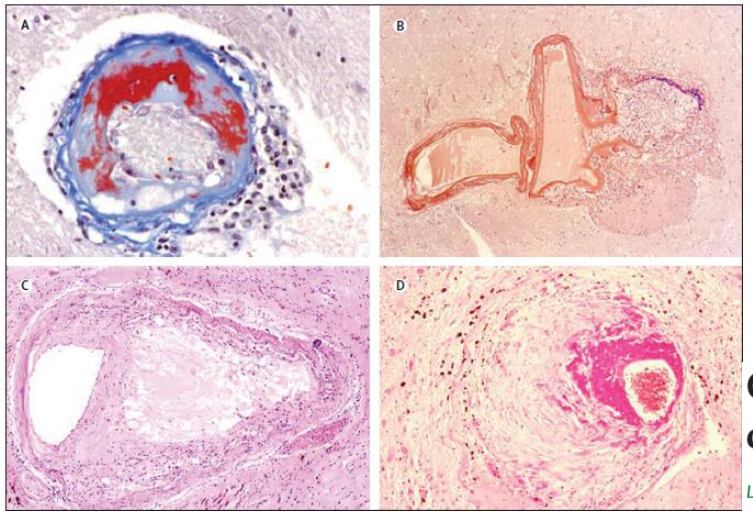
GENETICA: NOTCH 3

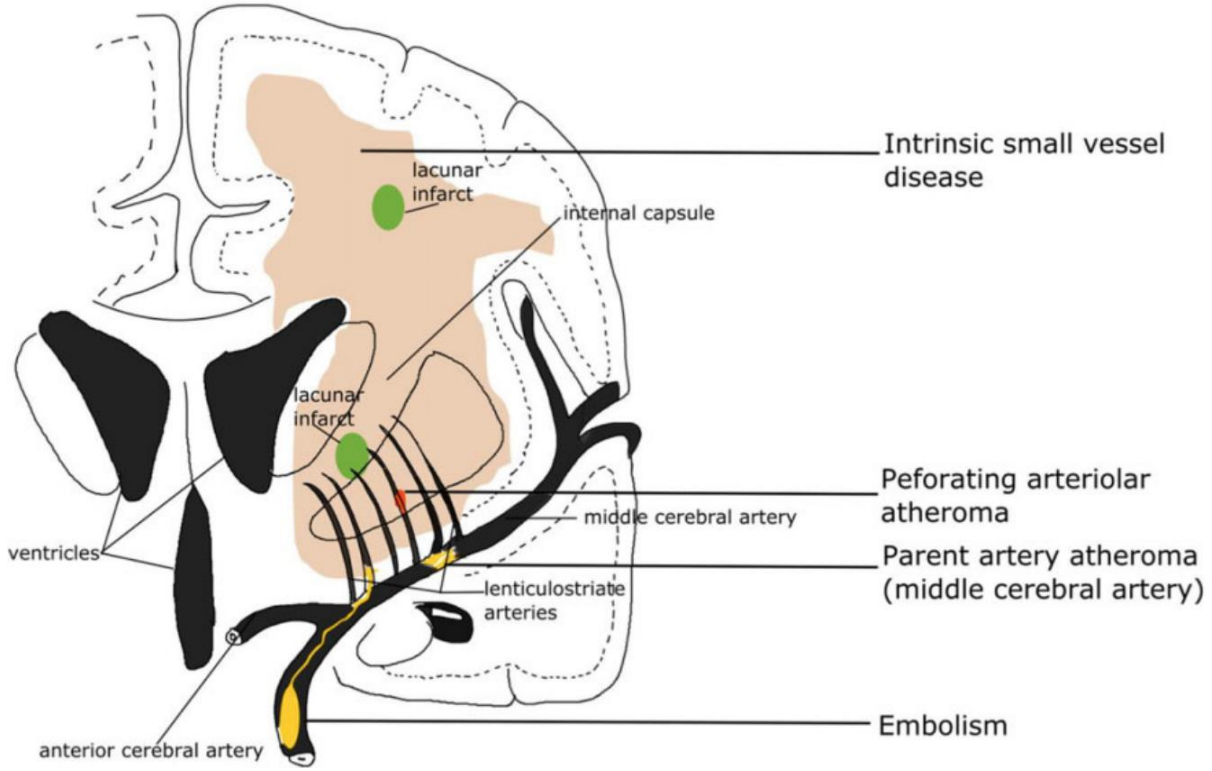
ISTOLOGIA: depositi di materiale granulare osmiofilico vicino i vasi

Arteriolosclerosi

- Malattia dei piccoli vasi
- Correlata fattori di rischio cardiovascolari ed età
- Istologia:
  -demielinizzazione; gliosi
  -arterie perforanti lunghe
    -lipoialinosi
    -microateromatosi
    -necrosi fibrinoide
    -ispessimento delle pareti dei vasi
    -restringimento del lume

PATOLOGIA SISTEMICA:
RENI
RETINA





Cerebral Small Vessel Disease

Cell Transplantation
2018, Vol. 27(12) 1711–1722

DOI: 10.1177/0963689718795148
journals.sagepub.com/home/cll
SAGE

Qian Li[1,*], Yang Yang[2,*], Cesar Reis[3], Tao Tao[2], Wanwei Li[1], Xiaogang Li[2], and John H. Zhang[3,4]

Cerebral small vessel disease: from pathogenesis and clinical characteristics to therapeutic challenges

Lancet Neurol 2010; 9: 689–701

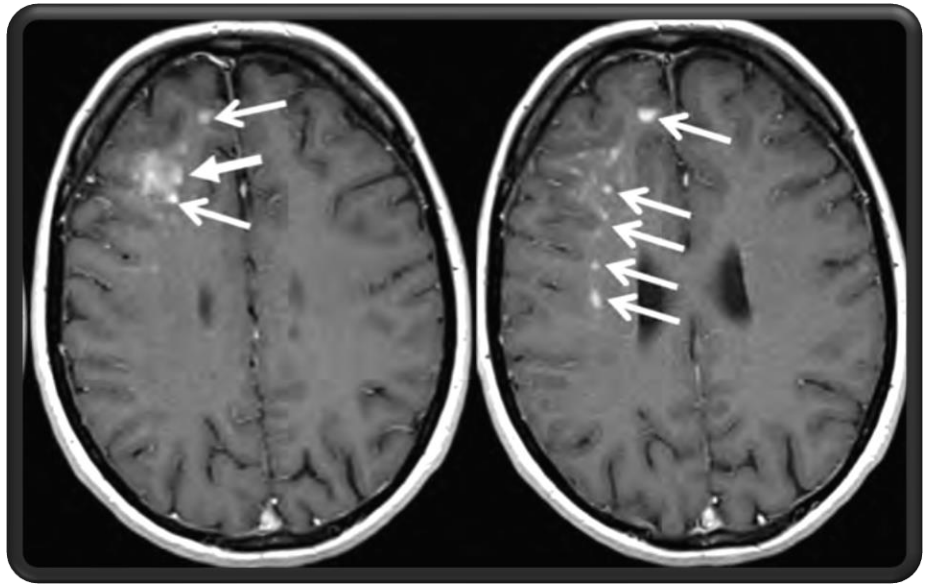
Leonardo Pantoni

IRIS – Immune Reconstitution Inflammatory Syndrome

Paradosso peggioramento dei sintomi neurologici in pazienti sottoposti a plasmaferesi(PLEX) o Immunoadsorbimento (IA) per rimuovere il natalizumab dal plasma

Flogosi da improvviso ripristino delle difese immunitarie

Aspetto tipico (anche HIV-PML): infiltrato di cellule T-CD8 e macrofagi anche nella sostanza bianca mielinizzata e nella sostanza grigia



- Quadro clinico-radiologico
- Fisiopatologia:

-stato ipertensivo severo → perdita delle capacità di autoregolazione del flusso intracranico

-tossicità sistemica/tempesta citochinica →

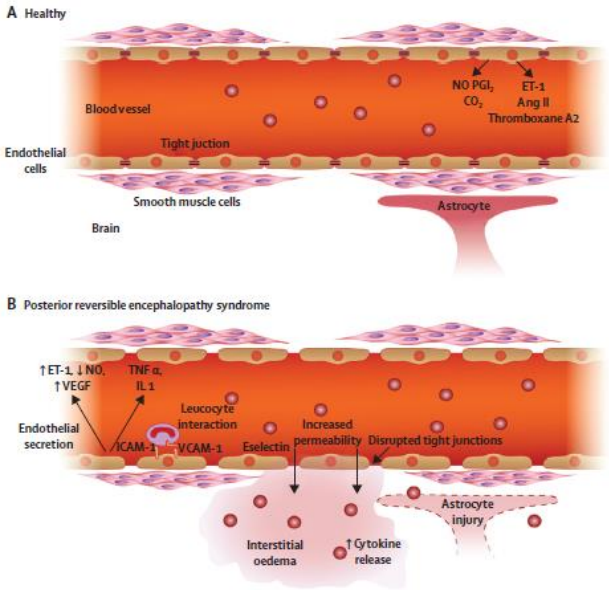
danno endoteliale →vasocostrizione ed ipoperfusione

ECLAMPSIA
GVHD
MAL AUTOIMMUNI

- Istologia:

-**edema vasogenico subcorticale parieto-occipitale** (più frequente)

-demielinizzazione e danno assonale???????



Posterior reversible encephalopathy syndrome: clinical and radiological manifestations, pathophysiology, and outstanding questions

Jennifer E Fugate, Alejandro A Rabinstein

Lancet Neurol 2015; 14: 914-25

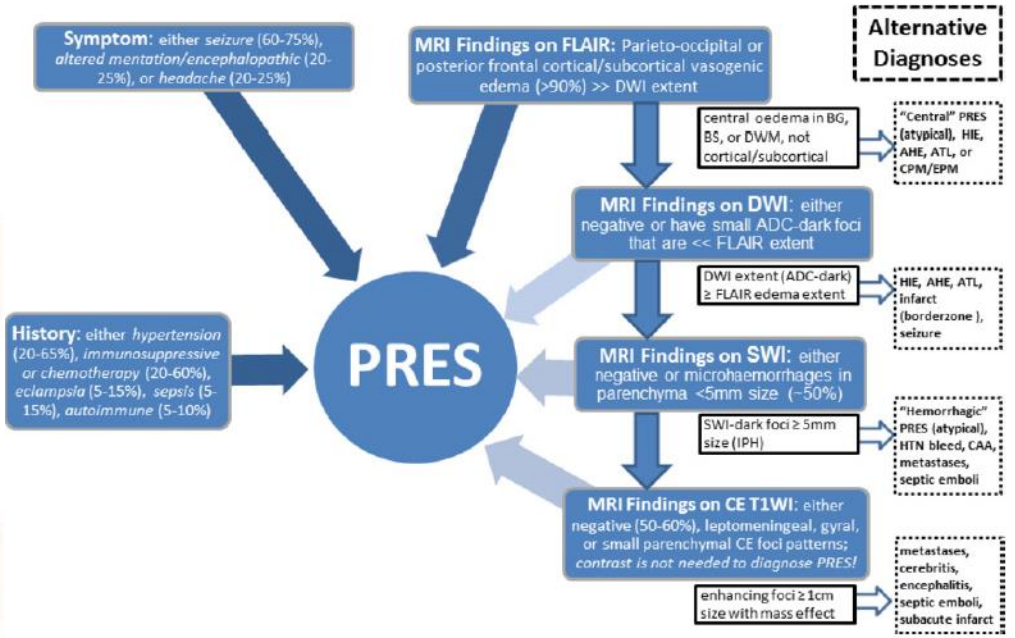
Cerebrovascular disease

REVIEW

Controversy of posterior reversible encephalopathy syndrome: what have we learnt in the last 20 years?

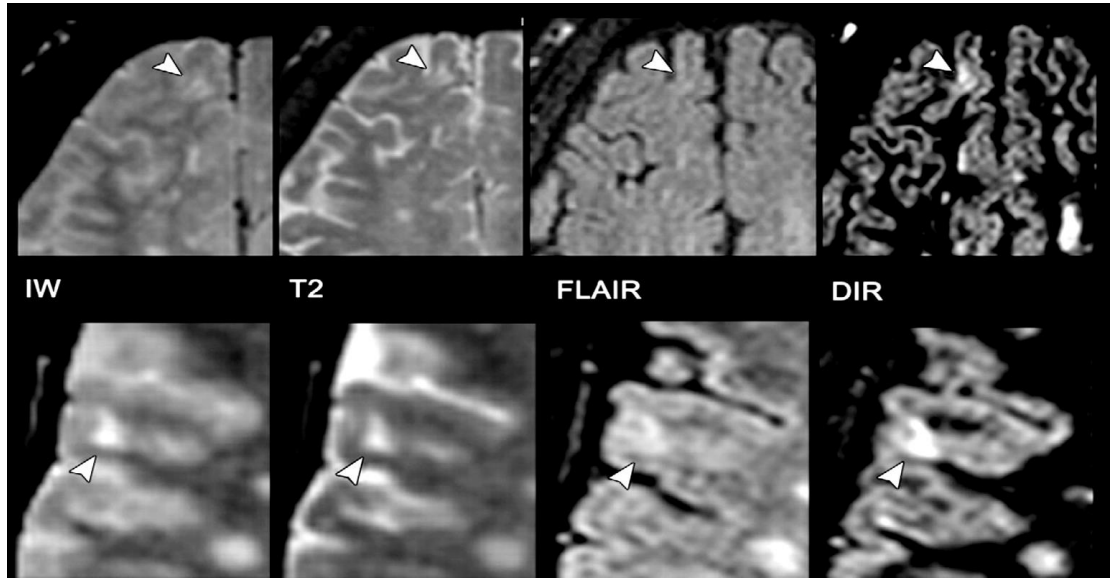
Bo Gao,[1] Cui Lyu,[2] Alexander Lerner,[3] Alexander M McKinney[4]

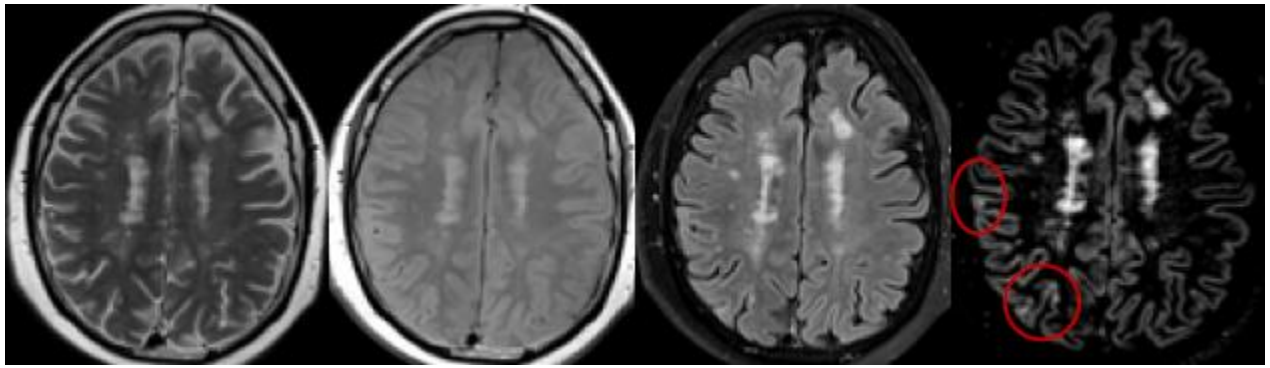
Converging on the Diagnosis of "Typical" PRES in >90% of Patients



Sclerosi Multipla

RM sedi meno frequenti /atipiche: sostanza grigia

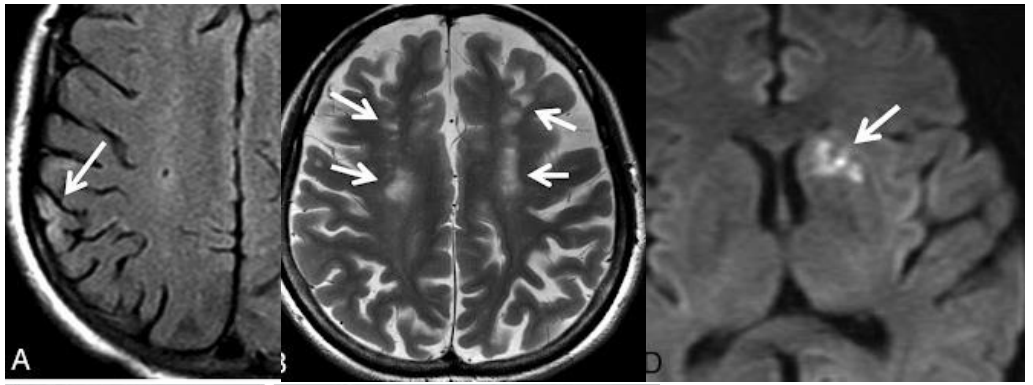


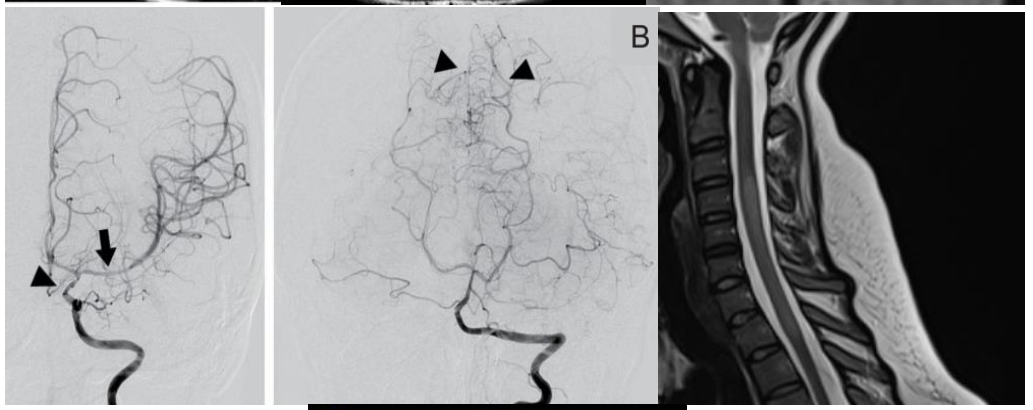


Neurolupus

Malattie cerebro-vascolari

1) Malattie dei piccoli vasi ed atrofia cerebrale (25% dei casi)
    Alterazioni T2 sostanza bianca sottocorticale e periventricolare
2) Lesioni ischemiche
3) Emorragie intra-assiali
4) Trombosi seni venosi durali
5) Vasculiti cerebrali
    Quadro RM non specifico; angiografia: focali riduzioni di calibro



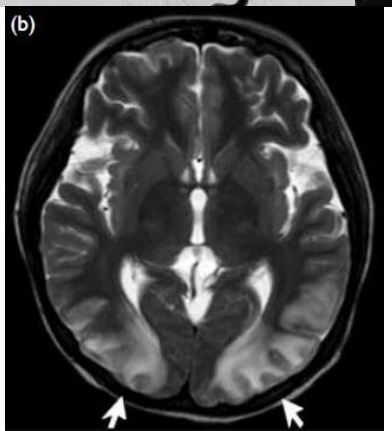


Mielopatie

1) Mieliti trasverse
2) Neuriti ottiche

Ulteriori manifestazioni

1) Meningiti asettiche (leptomeningiti focali o diffuse)
2) Ascessi cerebrali (secondari a emboli micotici valvolari)
3) PRES (secondaria ad ipertensione)
4) Encefalite anti recettori NMDA (legati a patologie e sintomi psichici)



Neurolupus

- Malattia sistemica autoimmune, 30-40% neurolupus
- Patogenesi:

  -vasculiti

  -coagulopatie LAC-relate

- Imaging:

-**multiple** lesioni SB sottocorticale e profonda (pz asintomatici)

-aree di demielinizzazione **estese** e **confluenti** (pz sintomatici)

-**diffusione ristretta**

-T1postGd: **potenziamento**

} FASE ACUTA

Quadro RM non specifico; angiografia: focali riduzioni di calibro