



IDIOPATHIC DIFFUSE SUPERFICIAL SIDEROSIS IN CNS: A CASE REPORT WITH APPLICATION OF RATIONAL INVESTIGATION

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Case report

A 60-year-old man with 4-year history of slowly progressive **algoparesthesias** in the lower limbs, bilateral **hearing loss** and **gait instability**.

No evidence of spontaneous or traumatic bleedings in the CNS, no systemic diseases.

Medical history: previous **traumas** (diving head concussion during childhood, severe back trauma skiing at age 20, car accident with back trauma two years before admission). Familiar history: unremarkable for neurological diseases.

Physical exam: right 6th cranial nerve palsy, bilateral hypoacusis, paretic hypertonus and diffuse brisk reflexes, bilateral Babinski's sign, slight dysmetria, dysesthesias in lower limbs, hypopallestesia, ataxic gait, positive Romberg and pull test.

Lab exams: **iron-deficient anaemia**.

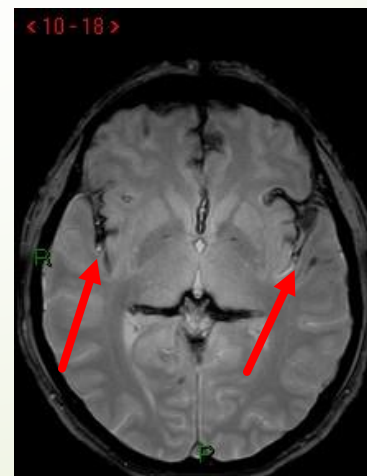
Audiogram: **sensorineural hearing loss**.

MEPs and SSEPS both delayed.

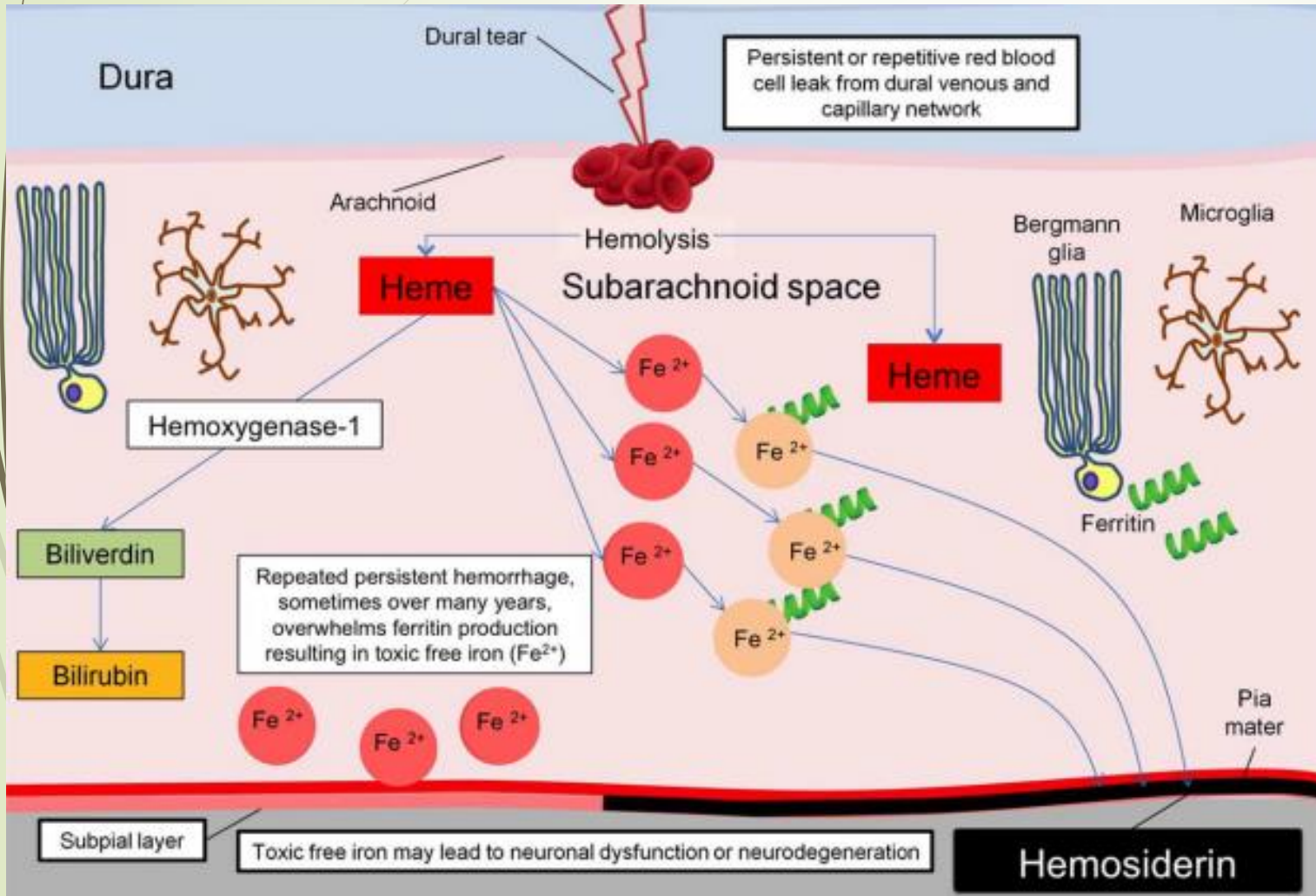
Lumbar puncture: raising of albumin, tau and p-tau levels, 22 white cells/mmc; **spectrophotometry:** raised levels of **bilirubin**.

MR imaging:

linear **hemosiderin deposit** on the brainstem, in paravermian liquoral spaces, lateral sulci, the mesial temporal-frontal lobes and the anterior cervical and dorsal regions of the medulla. Gadolinium-enhanced suspected vascular abnormalities at D9 level.



Superficial siderosis (SS)of the central nervous system



- first described in 1908
- rare neurodegenerative condition resulting from hemosiderin deposition in the subpial layers of CNS.
- hypothesized to result from a low-volume protracted "leak" of red blood cells into the subarachnoid space.
- neurotoxic action of heme leads to Bergmann glia and microglia release of hemoxygenase-1 and ferritin (scavengers).

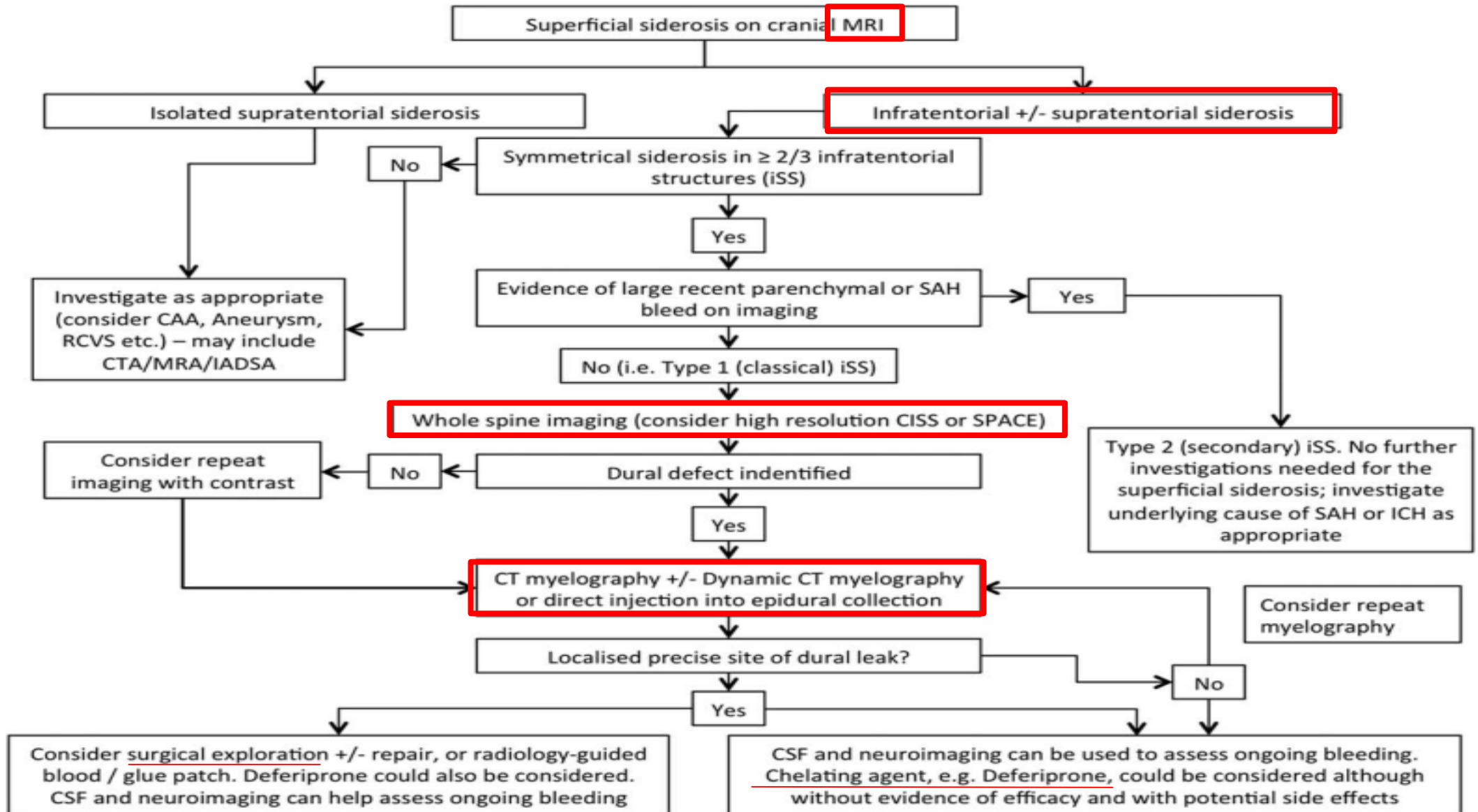
Two anatomical patterns

1. Cortical superficial siderosis (**cSS**):
 - Supratentorial
 - often associated with cerebral amyloid angiopathy (CAA – older individuals)
 - transient focal neurological episodes (e.g. TIA)
2. Infratentorial superficial siderosis (**iSS**):
 - with or without supratentorial involvement
 - slowly progressive bilateral hearing loss, ataxia, and myelopathy
 - **symmetrical** enhancement in at least 2 of the following regions: (1)**brainstem** (2) **cerebellum** (3) **spinal cord or craniocervical junction**.
 - A wide range of suggested causes

	cSS (n=54)	iSS not fulfilling diagnostic criteria (n=23)	Idiopathic iSS (n=48)	Secondary iSS (n=17)
Intracerebral hemorrhage ± convexity SAH	28 (52%)	6 (26%)	0	9 (53%)
Traumatic contusions	16 (30%)	3 (13%)	0	0
Aneurysmal SAH	5 (9%)	3 (13%)	1 (2%) atypical aneurysm with slow rather than acute SAH	6 (35%)
Ruptured AVM	2 (4%)	1 (4%)	0	0
Cerebral venous sinus thrombosis	1 (2%)	1 (4%)	0	0
Nonaneurysmal/perimesencephalic SAH	0	1 (4%)	0	0
Surgical trauma directly causing hemorrhage and related siderosis	2 (4%)	4 (17%)	0	1 (6%)
Subdural hemorrhage	0	1 (4%)	0	1 (6%)
<u>Dural abnormality^b</u>	0	0	40 (83%)	0
Tumor	0	0	4 (9%)	0
Spinocerebellar ataxia with associated brain iron accumulation	0	1 (4%)	0	0
Hemorrhagic transformation of an infarct	0	1 (4%)	0	0
<u>Cause identified</u>	54 (100%)	22 (96%)	45 (94%)	17 (100%)
No cause identified	0	1 (4%)	3 (6%); none had adequate spinal imaging	0

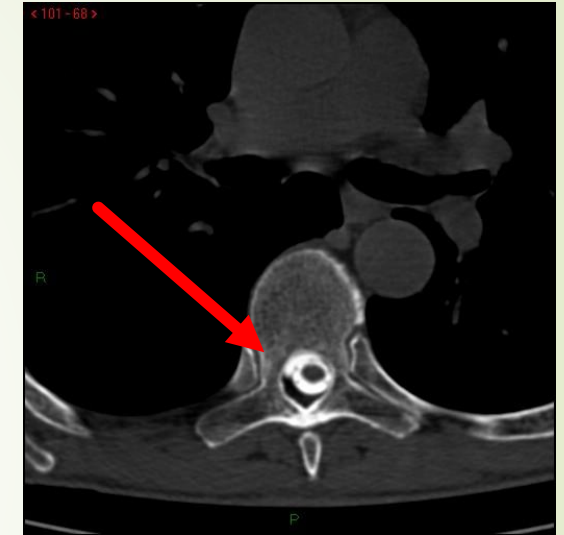
IS IT REALLY IDIOPATHIC ???

Proposed diagnostic algorithm for SS



myelography CT (Iopamidol, 300 mg/ml)

diffuse contrast opacification of a ventral pseudomeningocele in D2-D7 segment with tear at D7 level, and diffuse epidural abnormalities.



Thanks for the attention

