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## Further In-Depth Look at Superficial Siderosis (and Intracranial Hypotension)

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## Further In-Depth Look at Superficial Siderosis (and Intracranial Hypotension)

We greatly appreciated the excellent review of superficial siderosis (SS) by Kumar.<sup>1</sup> Regarding a few details, however, we do not share the same view.

One is the presence of “calcifications at sites of hemosiderin deposition around the brain stem,” responsible for a rim of hyperattenuation sometimes seen on CT. Kumar<sup>1</sup> attributes this interpretation (calcium for high densities) to our previous article published in the *American Journal of Neuroradiology* in 1993.<sup>2</sup> Actually, in that article, we considered this hyperattenuation “compatible with . . . hemosiderin” on the basis of several accurate pathologic studies carried out by Koeppen and Dentinger,<sup>3</sup> also quoted by Kumar. As in hemochromatosis, iron is sufficient to justify the hyperattenuation, without necessarily invoking calcium that, as far as we know, has never been described in SS.

The second point deals with the involvement of cranial nerves. Because the active mechanism of transforming blood by-products into hemosiderin and ferritin described by Koeppen and Dentinger<sup>3</sup> requires the presence of central myelin (ie, of glial cells), the nerves involved should be the first, second, and eighth: the first and second because they are part of the central nervous system, the eighth because the transition from central to peripheral myelin (produced by Schwann cells) is known to occur in the acoustic canal, more than 10 mm from the entrance into the brain stem. For the other cranial nerves (and spinal roots), the transition is within 0.5–1 mm from the brain stem (and spinal cord). See Fig 1 for the first cranial nerve and illustrations from previous articles for the other cranial nerves and spinal roots.<sup>2,4,5</sup> In a case of SS we recently observed, however, the loss of signal intensity along the trigeminal root extending for a few millimeters along the cisternal segment; it has been demonstrated that in this root, the central myelin–peripheral myelin transitional zone is not always so close to the brain stem as is usually considered, but it may be found up to 6–7 mm from the brain stem. In good-quality studies, the seventh nerve, in our experience, is always normal, in contrast to the T2 or T2\* hypointensity of the eighth nerve.<sup>4</sup> (See also Fig 6 in the article by Koeppen and Dentinger.<sup>3</sup>) In summary, the T2 hypointensity on the brain and spinal cord surface and along the short initial segment of the nerves and roots where central myelin is present perfectly corresponds to the histologic and immunocytochemical observations made by Koeppen and Dentinger.<sup>3</sup>

An interesting feature of SS of the cranial nerves is that there is a

clinical discrepancy between olfactory, visual, and acoustic involvement. The explanation is simple: Deafness severely impairs the patient’s daily life. Loss of smell and impairment of peripheral vision (macular fibers are centrally located in the optic nerve and therefore not affected) are much better tolerated and sometimes not even tested by the neurologist but are, indeed, present.<sup>2</sup>

Finally, in mentioning the association of SS with dural defects, spinal epidural collections, and dural enhancement, Kumar also refers to spontaneous intracranial hypotension (SIH) using the term “CSF hypovolemia.” We think that this term is a misnomer.<sup>6</sup> As we already pointed out, “hypovolemia” means decreased volume of blood, whereas in SIH, in agreement with the Monro-Kellie doctrine, the venous blood is increased to compensate for the loss of CSF, as manifested by dilation of venous sinuses, engorgement of the pituitary gland, and dilation of spinal epidural venous plexus.<sup>6</sup> The correct term (according to *Stedman’s Medical Dictionary*, Lippincott, Williams & Wilkins, 2006) should be “CSF hypovolemia,” but “hypovolemia” has never been used and may be a potential source of confusion. If we want to emphasize the leakage of CSF rather than its decreased pressure, we prefer using the expression “CSF loss of volume,” which certainly is correct, clear, and unambiguous.

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**Fig 1.** T2\*-weighted gradient recalled-echo axial (A) and T2-weighted turbo spin-echo coronal images 8 mm apart (B and C) show remarkable hypointensity consistent with iron incrustations on the olfactory bulb (B) and tract (A and C) on both sides (arrows) in a 63-year-old patient with SS of the central nervous system.