Neuroradiology

Superficial siderosis: Chronic sequelae following brain hemorrhage

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ABSTRACT

Superficial siderosis is a rare disease of the central nervous system. It is caused by hemosiderin deposition usually following subarachnoid hemorrhage. We report a 67-year-old man with history of motor vehicle accident in 1974 who presents with tremors, worsening ataxia, and impaired auditory, olfactory, and gustatory sensation. The patient was evaluated with magnetic resonance imaging of the brain that showed areas of superficial low T2 signal throughout the posterior fossa, ventricles, sulci, and cisterns, most conspicuous on the gradient-recalled echo T2* susceptibility-weighted sequence. These findings are compatible with old blood products (hemosiderin) and the diagnosis of superficial siderosis.

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Introduction

Superficial siderosis (SS) is a rare disease of the central nervous system (CNS). It is a result of hemosiderin deposition beneath the pia on the brain and CNS (1). It is most commonly believed to be a sequela of hemorrhage in the subarachnoid space [1–3]. It was initially diagnosed in biopsy postmortem in 1908 [3–6]. Recent advances in imaging technology, most notably magnetic resonance, have made these diagnoses possible in premortem. The most common initial presentation is sensorineural hearing loss followed by gait ataxia [2,3].

Here we report a 67-year-old man with tremors, worsening ataxia, and impaired auditory, olfactory, and gustatory sensation 43 years after a motor vehicle accident (MVA) and a cerebral hemorrhage. Magnetic resonance imaging (MRI) of the brain revealed SS.

Case report

History

A 67-year-old man with history of an MVA in 1974 presented to our hospital for progressively worsening tremors that were coarse, resting, intentional, and postural in nature. He noted 3 years of progressively decreasing auditory, olfactory, and gustatory sensation. On physical examination, there was decreased vibratory sensation of the lower extremities asymmetrically prominent on the left. The patient had no previous imaging available. An MRI of the brain was obtained.
Clinical imaging

A MRI examination of the brain without contrast material on a 1.5-Tesla magnet was used as part of the workup.

Radiological description

MRI of the brain showed superficial hypointensity on T2 signal throughout the surface of the posterior fossa, the ventricles, and the spine, most conspicuous in gradient-recalled echo (GRE) T2* susceptibility-weighted images.

Focused differential

The differential diagnosis for low T2 signal diffused throughout the surface of the brain and spinal cord includes SS, neurocutaneous melanosis, and meningoangiomatosis [7]. SS may result from repetitive subarachnoid bleeds [8]. The gradually progressive loss of auditory, olfactory, and gustatory sensation and ataxia were highly suggestive of SS as a sequela of the MVA and the brain injury 43 years earlier.

Final diagnosis

The clinical findings combined with the imaging confirmed the final diagnosis of SS of the brain and the spine. Treatment is supportive.

Discussion

SS is a rare and commonly incidental finding previously found postmortem during autopsy [3,6] before the advances of recent imaging, in particular MRI. SS is typically a sequela of recurrent hemorrhage in the subarachnoid space [1–3]. Hemosiderin, a blood product, deposits along the leptomeninges. Over time, methemoglobin breakdown products are accumulated within the macrophages as hemosiderin and ferritin. This deposition of this “foreign” material can occur all along the CNS, including the brain and the spine. This deposition process manifests with classical clinical syndromes [4] as seen in our case. Our patient presented with tremors, worsening of ataxia, and impaired auditory, gustatory, and olfactory sensation. Hemosiderin deposition causes a local inflammatory process that includes reactive gliosis, neuronal loss, and demyelination [3–5]. The hemosiderin precipitate commonly settles within gravity-dependent areas, especially the posterior cerebellum. When the cerebellum is affected, the deposition of the hemosiderin is usually at the peripheral folia, often associated with atrophy of the vermis. Involvement of the cerebellum manifests with ataxia and tremors. Cranial nerves I and VIII are most commonly involved because of their long extents within the subarachnoid spaces. This explains the auditory and the olfactory losses.

Hemosiderin iron cores and ferritin contain approximately 2000 iron molecules ferromagnetically coupled to produce a “superparamagnetic” product that is highly susceptible on MRI. T2-weighted MRI and GRE are the most sensitive to susceptibility effect of paramagnetic substances. T2-weighted images manifest the T2 shortening with hypointensity especially on GRE. This produces the “bloom” effect because of strong susceptibility effect [9,10]. Spin-echo and fast spin-echo T2-weighted sequences demonstrate progressively decreased sensitivity to the susceptibility effects of hemosiderin and therefore are less useful. This results in a hypointense rim at the margin of the lesion along the leptomeninges of the brain and the spinal cord. The pons and cerebellum are the most affected. The extent of the distribution does not have to correlate with the severity of the symptoms [3,9]. The etiology of SS may involve any chronic or recurrent bleeding etiology within the subarachnoid space such as vascular anomalies (eg, aneurysms, AVM, and cavernous hemangioma), trauma, and neoplasm. About 80%-90% of nontraumatic subarachnoid hemorrhages are a result of intracranial aneurysm rupture. The bleeding source may not always be identified intracranially and might originate from the spine, (eg, myxopapillary ependymoma), therefore in absence of identification of an intracranial source of SS, a total spine MRI or a cerebral-spinal...
Angiography may help to identify subtle vascular abnormalities. A bleeding source has been found in 54%-75% [3,6,9] of cases. An interesting differential diagnosis includes leptomeningeal melanin, meningioangiomatosis, and neurocutaneous melanosis. Leptomeningeal melanin is often associated with the amount of skin pigmentation, and appears as a thick hypointense rim on the ventral surface of the medulla oblongata; therefore, correlation with the skin pigmentation is needed if there is absence of other possible etiologies [10].

SS is caused by deposition of hemosiderin within the subarachnoid space especially along the surface of cerebellum and cranial nerves. This process is not benign and involves local inflammation at the surface of the leptomeninges. This can result in ataxia and impairment of auditory and olfactory senses; however, the extent of the imaging findings does not always correlate with the severity of the clinical symptoms. Identification of SS indicates a search for the source of bleeding in order to prevent progression. It should be emphasized that if no source of bleeding is identified, a total spine MRI and even cerebral-spinal angiography should be considered. Therefore, MRI plays a role both in the diagnosis of the disease and in the evaluation for the cause (Figs. 1–3).

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Figure 2 – A and B—Coronal gradient echo T2* susceptibility-weighted slices showing hemosiderin deposition over the cerebellar folia (black arrows) demonstrating the bloom effect.

Figure 3 – Sagittal short-T1 inversion recovery slice showing a hypointense rim at the margin leptomeninges of the spinal cord (white arrows).