CASE REPORT

"Superficial Siderosis", a rare concomitant finding in Spontaneous Intracranial Hypotension

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ABSTRACT

Superficial siderosis as a concomitant finding in SIH has not been quite frequent. Superficial siderosis, especially in the posterior fossa, when seen in patients with headaches, is vital to evaluate for spontaneous intracranial hypotension. Subpial hemosoderin deposition in the brain and spinal cord usually occurs as a result of recurrent or persistent haemorrhage into the subarachnoid space, the source of which is not discernible in all cases, in spite of extensive neuroimaging. We report a patient who presented with headache and vomiting, whose neuroimaging findings were indicative of features pointing towards superficial siderosis and SIH. Early identification of superficial siderosis in headaches not only helps us diagnose SIH but also guides clinicians to provide proper treatment, which could avoid complications.

KEY WORDS: Superficial Siderosis, Spontanous Intracranial Hypotension, Dural Leak.

Introduction

Intracranial hypotension has been a topic of interest in the field of neuroscience for more than a century. One of the initial recognitions of it as a syndrome is credited to Lerische, who in 1920 described the symptomatology consisting of headache, nausea, vomiting, hyperthermia, and coma in patients with closed skull fractures, secondary to CSF leaks through fracture defects[1]. The term spontaneous intracranial hypotension caught attention when people with no history of trauma, lumbar punctures, or intradural surgeries presented with symptoms of orthostatic headache, nausea, photophobia, or neck pain. The root cause of SIH is attributed to CSF leaks from osteodiscogenic microspurs, rupture of the spinal nerve root sleeve, or rarely, a CSF venous fistula[2,3]. Triggering factors like sneezing, coughing, bouts of laughter, and straining during bowel movements have been identified in a subset of patients, especially in those who have an underlying connective tissue disorder like Marfan syndrome and Ehler Danlos syndrome[4].

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Superficial siderosis as a concomitant finding in SIH has not been quite frequent. Subpial hemosoderin deposition in the brain and spinal cord[5–8] usually occurs as a result of recurrent or persistent haemorrhage into the subarachnoid space, the source of which is not discernible in all cases, in spite of extensive neuroimaging. In symptomatic patients, the classical features of progressive gait ataxia, dysarthria, SNHL, and cognitive impairment run a chronic course. We report a patient who presented with headache and vomiting, neuroimaging findings of whom was indicative of features pointing towards Superficial siderosis and SIH.

Case report

A 42-year-old female patient presented with a history of acute onset of holocranial headaches of moderate to severe intensity without any postural variation,

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which was associated with multiple episodes of vomiting. There was no history of fever, recent head trauma, LOC, or seizures. There was no past history of significant headaches. The neurological examination was unremarkable. A non-contrast CT scan of the brain was normal. She was given analgesics and supportive care. As she continued to have a disturbing headache, she was admitted, and an MRI study of the brain was done, which revealed T2 hypointensities with blooming within the subpial layers of the left parieto-temporal, bilateral occipital, and tentorial surfaces of the cerebellum (Figure 1 (A) and (D)). TOF MRAngiogram did not reveal any aneurysms in major intracranial arteries. Further detailed evaluation of MRI images showed mild dropping of the splenium of the corpus callosum, reduced pontomescencephalic angle, enlarged pituitary (Figure 1 A), and engorged venous sinuses, which raised the suspicion of intracranial hypotension. Subsequently, the patient underwent an MRI study of the whole spine, and MR venography showed engorged venous sinuses(Figure 1 B). MRI of the spine revealed the presence of a longitudinally extending spinal epidural fluid collection from C7 to T12 spinal level (Figure 2 A) and a focal spinal longitudinal extradural CSF collection (SLEC) at the L4-L5 spinal level (Figure 2 (A-C)). All these imaging findings led to the diagnosis of superficial siderosis secondary to SIH. It was confirmed from past history that there was no past history of trauma or SAH. The patient was managed conservatively with IV fluids, amitriptyline, theophylline tablets, and bed rest. During the course at the hospital, the patient had a persistent headache for the initial two days, but later it became better, and he was discharged on the 7th day at the hospital. She was advised regarding the need for a CT/MRI myelogram to localise the site of the dural tear and epidural blood patch treatment in case of any recurrence of similar symptoms in the future. On her review visit after four weeks, she was completely asymptomatic.

Discussion

Superficial siderosis is a chronic condition consisting of hemosiderin deposition in the subpial layers of the brain due to chronic or intermittent low-grade extravasation of blood into the subarachnoid space. Its classified into cortical and infratentorial, cortical classically secondary to amyloid angiopathy, which presents with focal cortical signs, classically seen in older patients. Infratentorial is seen typically around the cerebellum, which is seen as a sequalae to extravasation of blood secondary to traumatic brain injury. It presents with progressive sensory neural hearing loss and ataxia. SIH can have serious complications if not identified early and treated, including dural venous sinus thrombosis, subdural hematoma, and subarachnoid haemorrhage.

Spontaneous intracranial hypotension (SIH) is an important etiology of infratentorial superficial siderosis of the central nervous system. The mechanism of SIH is usually secondary to dural leak; it can be associated with collagen disorder. The mechanism of superficial siderosis in SIH is due to a loss of CSF volume leading to venous engorgement[9] (according to the Monro-Kellie doctrine), and the associated traction of superior vermian veins due to brainstem sagging might have contributed to the gradual oozing of blood into the subarachnoid space[10] ultimately leading to hemosiderin deposition over the subpial layers of the brain. In over 30% of patients, the underlying source of subarachnoid haemorrhage may not be identifiable[5]. A dynamic CT myelogram is key to localising the site of the CSF leak. In our case, although a CT myelogram was planned, the patient had considerable clinical improvement with conservative management; hence, it was not done. Spontaneous sealing off or slowing down of leaks due to unclear reasons might be the cause of clinical remission. The majority of cases of SIH can be treated conservatively; only those that are not responding to medical management or those that have complicated SIH may be considered for an epidural blood patch or surgical intervention.

The presence of MRI evidence of superficial siderosis, especially in the posterior fossa in patients with headaches, makes it vital to evaluate for spontaneous intracranial hypotension. In our case, even though the patient did not have any classical symptoms of SIH, evidence of posterior cranial fossa superficial siderosis and MRI features of SIH led to an early diagnosis. Further evaluation with a spine MRI demonstrated spinal longitudinal extradural CSF collection (SLEC). Unlike in this case, the requirement to identify the location of the dural defect and treat it with an epidural patch in cases of smaller defects or surgical dural repair for extensive defects should be a serious concern for the treating clinician in headaches refractory to conservative management. This might be important in SIH patients with MRI evidence of superficial siderosis, as the interventional therapy would prevent oxidative brain damage induced by hemosiderin deposition, which might otherwise

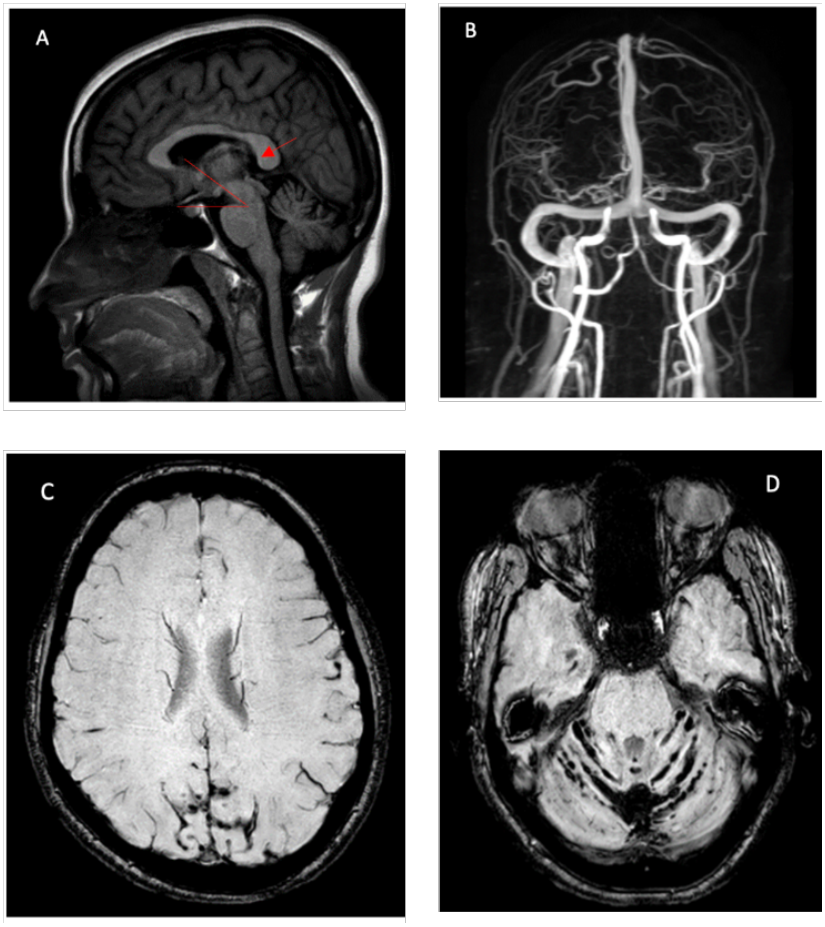

Figure 1: (A) and (B): MRI showing pontomescencephalic angle <50 and sagging of corpus callosum, (C and D) SWI image showing superficial siderosis

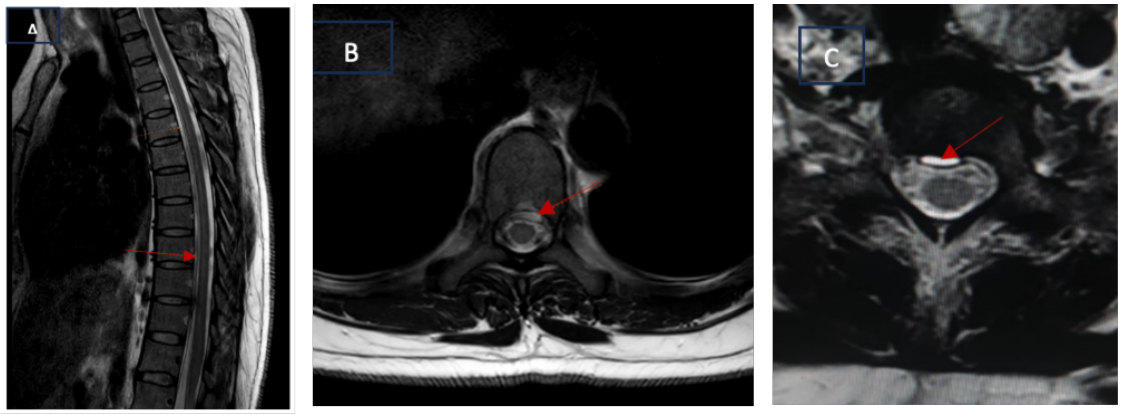

Figure 2: (A): Longitudinally extending spinal longitudinal extradural CSF collection (SLEC) from C7 to T12 spinal (B) and (C): Focal epidural fluid collection at the L4-L5 spinal level

end up clinically manifesting superficial siderosis. So early identification of superficial siderosis in headaches not only helps us diagnose SIH but also guides clinicians to provide prompt treatment, which could avoid complications.

Conclusion

Patients with head aches with MRI evidence of superficial siderosis, particularly posterior cranial fossa, must be re-evaluated for any evidence of SIH. Even though dynamic CT myelography is recommended, a non-invasive MRI spine screening to look for any dural leak and SLEC may help in the localization of dural tears. This can help in managing recurrent or complicated SIH with further interventions. Treating physicians must be vigilant to pick up superficial siderosis in the MRI of a headache patient, as it can be a radiological clue for SIH, which is a treatable disease.

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