Superficial siderosis presenting as chronic migraine: Rare presentation of a rare disease

Sir,

Superficial siderosis (SS) is a rare degenerative condition characterized by progressive cerebellar ataxia and sensorineural hearing loss.[1] It results from pathological deposition of iron in superficial parts of the brain and structures in contact of cerebrospinal fluid (CSF), predominantly in the hindbrain. The sources of hemorrhage are recent or previously resected central nervous system (CNS) tumors, CSF cavitary lesions, vascular malformations, CNS trauma, subarachnoid hemorrhage, amyloid angiopathy, and post hemispherotomy. In one-third, the cause of bleeding remains undetermined and hence categorized as idiopathic SS.[2,3] Although acute thunderclap headaches, acute severe headaches, and episodic migraine-like headaches have been described in SS, chronic migraine-like headaches as presenting feature have not been previously described in SS. The purpose of this letter is to highlight such a presentation.

A 75-year-old hypertensive, nonsmoker, nonalcoholic man presented with complaints of insidious onset throbbing, holocranial headache for 2 years. Headache was moderate in intensity (visual analog scale scoring of 6/10), associated with phonophobia and occasional photophobia, vomiting, and dizziness. Headaches used to exacerbate by head movements, coughing, and bending forward. Autonomic features were absent. He would mostly retire to bed during the attacks. On an average, he would have 20 days of such headaches in a month of which about half of the days will have significant migrainous features. The patient categorically denied any migraine headache during his youth although the possibility cannot be excluded completely due to recall problems. There was also no history of thunderclap headaches. There was no history of head injury. On examination, his vitals were normal. Temporal arteries were nontender. His mini mental state examination score was 19/30 and frontal assessment battery score was 11/18 with deficits in visual pattern completion test, mental flexibility, alternating motor pattern test, and positive prehension behavior. Bilateral sensorineural hearing loss (right > left) was present. Subtle impairment in finger nose, dysdiadochokinesia and subtle tilt toward the left side on walking were noted. Pure tone audiometry and brainstem evoked response confirmed sensorineural hearing loss. Thus, although apparently our patient fulfilled the current diagnostic criteria of chronic migraine as per International Classification of Headache Disorders 3 beta in terms of frequency, duration, and character, the presence of other subtle neurological signs clearly stamped it as a secondary headache. He was thoroughly investigated for secondary causes. His routine blood studies such as hemogram, liver function test, kidney function test, and sugar and lipid profile were normal. His CSF examination was normal. Magnetic resonance imaging (MRI) of the brain showed hemosiderin deposition in cerebellum and around brain stem suggestive of SS [Figure 1]. Microbleeds suggestive of amyloid angiopathy were not seen. Intracranial MR angiography, MRI of the spine, and MR myelogram were normal thereby suggesting idiopathic SS.

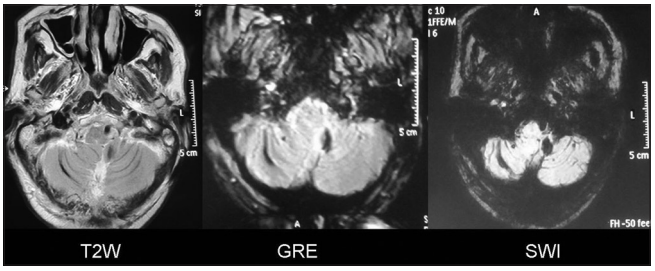

Figure 1: T2-weighted, gradient echo, and susceptibility echo (susceptibility-weighted imaging) sequences showing deposition of hemosiderin in the crests of bilateral cerebellar folia, vermis, and brainstem

We searched for case reports and review articles documenting headache as past or present symptom in SS and found that in 14% of patients, headaches were described that were thunderclap type, acute severe type, or episodic migrainous type.[1-4] Our report suggests that chronic migraine-like headaches can also be a presenting feature in SS. Classic features of gait ataxia and deafness in SS may be subtle and can be overlooked. SS is a potentially treatable condition.[5] Hence, all chronic daily headache patients must be thoroughly worked up by history, examination, and investigations to rule out a potentially treatable secondary cause. Mechanisms of chronic headache in SS remain speculative. Chronic hemosiderin deposition in the superficial parts of the brain (especially subpial layers of hindbrain) may stimulate the dural afferents resulting in chronic migraine-like headaches.

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Conflicts of interest

There are no conflicts of interest.

Debashish Chowdhury, Amit Shankar Singh, Geeta Anjum Khwaja

Department of Neurology, Govind Ballabh Pant Institute of Postgraduate Medical Education and Research, New Delhi, India

For correspondence:
Prof. Debashish Chowdhury, Room Number 508, Academic Block, Govind Ballabh Pant Institute of Postgraduate Medical Education and Research, Jawaharlal Nehru Marg, New Delhi - 110 002, India.
E-mail: debashishchowdhury@hotmail.com

References

1. Kumar N. Superficial siderosis: Associations and therapeutic implications. Arch Neurol 2007;64:491-6.
2. Levy M, Turtzo C, Llinas RH. Superficial siderosis: A case report and review of the literature. Nat Clin Pract Neurol 2007;3:54-8.
3. Fearnley JM, Stevens JM, Rudge P. Superficial siderosis of the central nervous system. Brain 1995;118(Pt 4):1051-66.
4. Lummel N, Wollenweber FA, Demaerel P, Bochmann K, Malik R, Opherk C, *et al.* Clinical spectrum, underlying etiologies and radiological characteristics of cortical superficial siderosis. J Neurol 2015;262:1455-62.
5. Levy M, Llinas R. Pilot safety trial of deferiprone in 10 subjects with superficial siderosis. Stroke 2012;43:120-4.

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