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Spinal ependymoma with central nervous system diffuse siderosis – a rare case

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Abstract
Superficial siderosis of the central nervous system is a syndrome caused by deposition of hemosiderin in the subpial layers of the central nervous system, occurring as a result of recurrent asymptomatic or symptomatic bleeding into the subarachnoid space. We report a rare case of superficial diffuse siderosis in a 37-year-old man who presented with Bilateral sensorineural hearing loss Right more than left along with bilateral lower limb weakness with decrease sensation below groin region. The detection of a spinal ependymoma at L3-S2, Intramedullary leads to further evaluation of MRI Brain with contrast showing diffuse superficial siderosis. Near total resection of the tumor arrested the progression of the neurological deterioration. Our report underlies the importance of early diagnosis and surgical management, with imaging examination of the full neuroaxis to identify the source of bleeding, to halt disease progression and improve prognosis.

Keywords: Central nervous system; Spinal ependymoma; superficial siderosis

Introduction
Superficial siderosis of the central nervous system (SSCNS) is a rare disease resulting from the accumulation of hemosiderin in the meninges, brain surface, spinal cord, and cranial nerves. SSCNS presents as slowly progressive sensorineural hearing loss due to involvement of the 8th cranial nerve, gait ataxia, pyramidal signs, and cerebellar dysfunction[1]. Despite extensive laboratory, imaging, and clinical evaluations, the bleeding site remains unidentified in approximately half of the cases [1, 2]. In this report, we describe a patient with hearing impairment, ataxia, and cognitive decline owing to SSCNS caused by spinal ependymoma-induced subarachnoid bleeding. Repeated subarachnoid hemorrhage results in deposition of hemosiderin on layers of the CNS in close proximity to the cerebrospinal fluid. Hemosiderin is toxic to neural tissue and may cause parenchymal damage. SSCNS affects males more often than females, we report a patient with SSCNS caused by clinically asymptomatic subarachnoid hemorrhage from a myxopapillary spinal ependymoma.

Case Report
A 37-year-old previously healthy man experienced gradual onset of decreased hearing in bilateral ear more in right ear than left ear for last ten years. An otorhinolaryngology examination was diagnostic for sensorineural hearing loss. Patients also complained about low back pain for last two years with radiation to bilateral lower limbs. Patient also complained about decreased sensation bilateral lower limbs more on left than right side for one year. Patient also had history of forgetfulness for last one year. For all these symptoms spinal MRI was performed which showed an intramedullary space occupying lesion at L3 to S2 level [Fig 1, 2, 3]. For all these symptoms patient was referred to neurosurgery OPD and patient admitted in neurosurgery department for clinico-radiological complete assessment. MRI brain with contrast showed evidence of diffuse sulcul hypo intensity on T2 FLAIR [Fig 4] images and evidence of blooming on GRE [Fig 5] sequence involving bilateral cerebral hemisphere, basal ganglia, cerebellum, brain stem suggestive of superficial siderosis. MRI Brain also shows EX-Vacuo dilatation of ventricular system with atrophic changes. MRS Brain was not conclusive. HRCT Temporal bone showed no obvious abnormality. His physical and neurological examination was significant only for bilateral ear hearing loss and positive straight leg raising test bilaterally. Pure tone audiometry showed right ear profound SNHL and left ear moderate-severe SNHL. Owing to the patient's bilateral deafness, it was difficult to conduct a detailed cognitive assessment. We excluded various coagulation disorders as a cause of the chronic subarachnoid hemorrhage through further laboratory

evaluation. Brain MR Angiography and venography was performed to determine the cause of the subarachnoid hemorrhage.

The patient underwent Bilateral L3, L4, L5, and S1 Laminectomy after lumber transpedicular screw rod fixation with complete resection of the spinal tumor. The postoperative course was uneventful and he was discharged 5 days after surgery. Histopathology examination of the resected tumor was diagnostic for a World Health Organization (WHO) grade I myxopapillary ependymoma. Following surgery, he had complete resolution of his back pain. At 6month follow-up the patient remained stable neurologically. Follow-up evaluation showed arrest of the back pain and lower limb weakness progression.

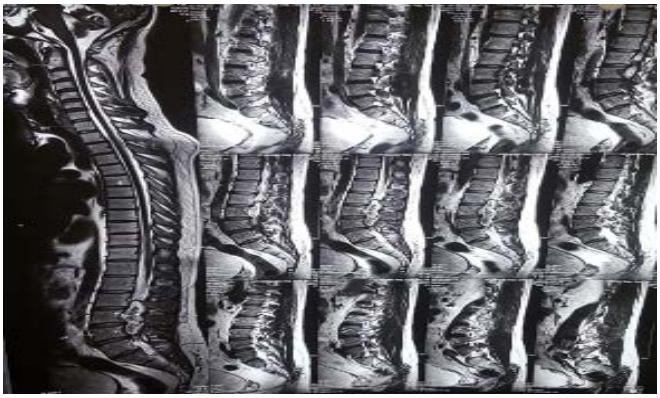

Fig 1: Sagital T1 MRI Lumbosacral showing SOL at L3-S2 Level

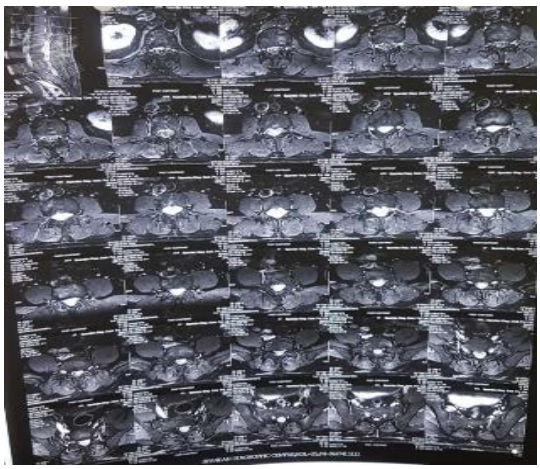

Fig 2: Axial T1 contrast Lumbosacral showing SOL at L3-S2

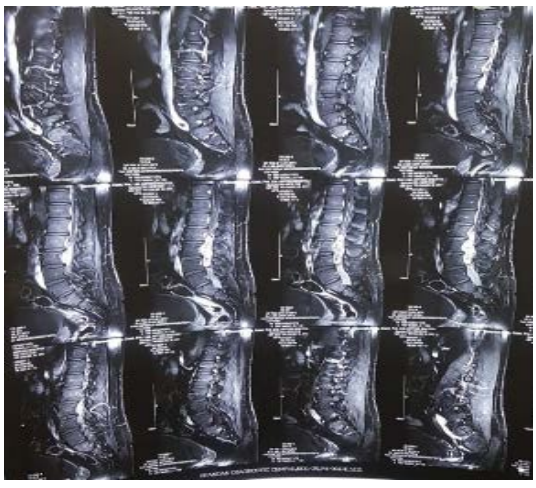

Fig 3: Sagital T1 MRI lumbosacral showing SOL at L3-S2

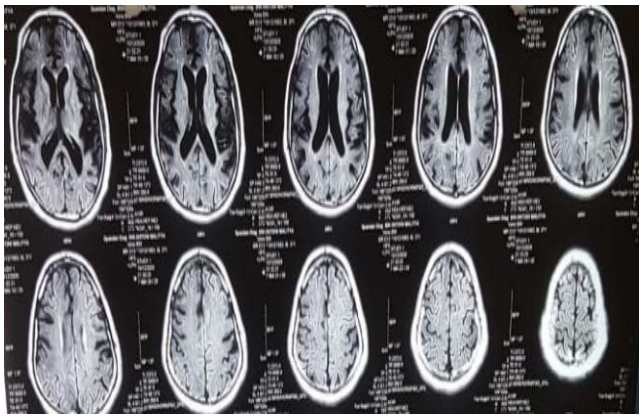

Fig 4: T2 FLAIR showing diffuse sulcul hypo intensity suggestive of Superficial siderosis

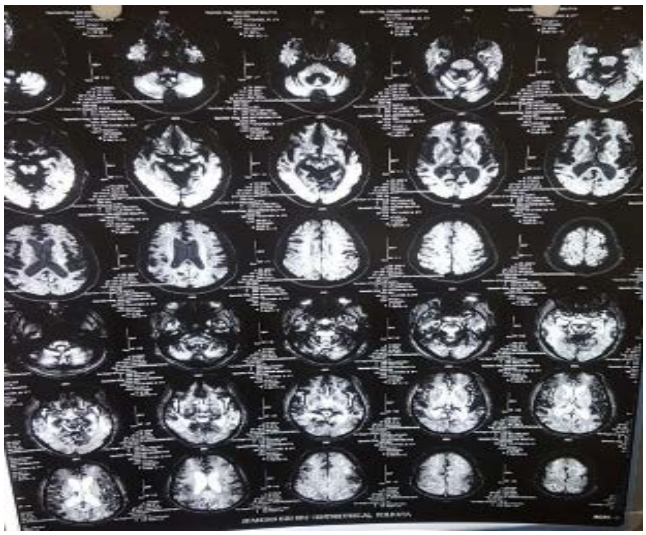

Fig 5: GRE sequence showing involving bilateral cerebral hemisphere and basal ganglia and brain stem suggestive of siderosis

Discussion

The most common causes of SSCNS include ruptured aneurysms, arteriovenous malformation, intradural cranial or spinal surgery, traumatic injury, and bleeding of unknown etiology.[1] However, more unusual causes of chronic bleeding include dural pathologies and tumors [3]. Tumors, including ependymomas, meningiomas, oligodendrogliomas, and pineocytomas, are found in 35% of cases [1]. In our patient, the cause of SSCNS was ependymoma of the spinal cord identified as the bleeding source. If our patient had not complained of severe back pain, the cause of her SSCNS might not have been found. Therefore, to find the cause of SSCNS, careful history taking and neurological examination are essential. Hearing loss, cerebellar ataxia, and pyramidal signs are the most frequent clinical findings, occurring in 95%, 88%, and 76% of cases, respectively [5]. The long glial segment of the vestibulocochlear nerve makes it vulnerable to axonal damage by iron deposition.

SSCNS is a rare, slowly progressing, and potentially devastating syndrome occurring as a result of continuous or recurrent bleeding into the subarachnoid space. The most common causes include ruptured aneurysms, bleeding of unknown etiology, arteriovenous malformation and traumatic injury; however, more unusual sources of chronic

bleeding include dural pathologies and tumors. Resulting deposition of hemosiderin in the subpial layers of the CNS may cause parenchymal damage.

SSCNS is typically characterized by sensorineural hearing loss, ataxia [1, 2], and pyramidal signs[3] mimicking spinocerebellar degeneration. Other SSCNS manifestations may include sensory and cognitive disturbances, seizures, headache, urinary symptoms, visual and olfactory complaints[2], anisocoria, lumbar backache, bilateral sciatica (10%) and neck pain, extra-oculomotor nerve palsy, and lower motor neuron signs with muscle wasting [3].

In their 1995 review, Fearnley *et al.* [4] described SSCNS secondary to a variety of dural pathologies, CNS tumors, and vascular anomalies. Kumar *et al.* [2] identified a history of trauma as a possible cause of SSCN in 17 of 22 patients in their series. Myxopapillary ependymoma is a rare, WHO grade I ependymoma occurring predominantly in the lumbosacral region, particularly the filum terminale. MRI evaluation of the entire neuraxis to diagnose SSCNS and to identify a potential source of bleeding is recommended[5]. On MRI, ependymomas without capsular rupture appear as well-circumscribed masses, which are hypo intense on T1-weighted and hyper intense on T2-weighted MRI. Homogenous enhancement following gadolinium administration distinguishes ependymomas from neurinomas of the cauda equina, which enhance heterogeneously. Optimal management of SS remains to be determined. Successful surgical management depends on early diagnosis of SSCNS with identification and ablation of the bleeding source before permanent CNS damage has occurred. In accordance with our report, arrest of SS symptoms following surgical excision of a spinal ependymoma has been described [6].

Conclusion

SS is a rare, slowly progressive condition with potentially severe clinical sequelae. SSCNS manifestations vary and presentation may mimic a variety of neurological and otorhinolaryngological diseases; thus, clinicians should be aware of and include SSCNS in the differential diagnosis of cerebellar-pyramidal syndromes. Optimal management remains to be determined. However, early diagnosis with identification and surgical ablation of the bleeding source appears to halt disease progression, thus improving the prognosis of the patient.

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