

# Superficial Siderosis Due to Spinal Myxopapillary Ependymoma Mimicking Idiopathic Intracranial Hypertension

Dear Sir,

Superficial siderosis (SS) of central nervous system is a rare and slowly progressive disease. It results from hemosiderin deposition in the subpial layers of the brain and spinal cord. The typical clinical presentation of SS includes progressive sensorineural hearing impairment, cerebellar ataxia, pyramidal dysfunction and dementia.<sup>[1]</sup> We present an unusual case of SS caused by a spinal myxopapillary ependymoma presenting with visual impairment mimicking idiopathic intracranial hypertension (IIH).

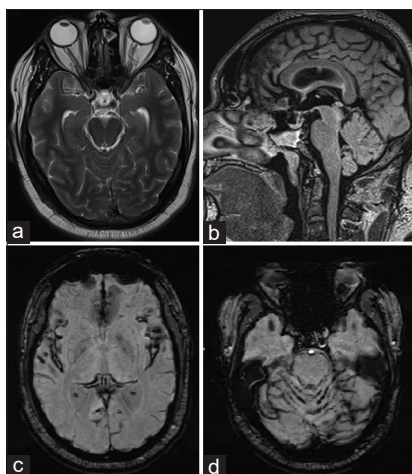
A 34-year-old man presented with subacute onset bilateral visual blurring of 3 weeks duration. He had no headache or eye pain. He was diagnosed as IIH elsewhere after brain imaging and initiated on acetazolamide. At presentation, he had bilateral papilledema with peripapillary haemorrhages. Central visual acuity was normal while visual field testing showed constriction of peripheral fields in both eyes. Visual evoked potentials and pure tone audiometry were normal.

Magnetic resonance imaging (MRI) of brain showed features of raised intracranial pressure, specifically, prominent peri-optic subarachnoid space, vertical buckling of the optic nerve sheath complex, flattening of the posterior sclera and partial empty sella [Figure 1a, b]. Susceptibility weighted images (SWI) showed extensive leptomeningeal hemosiderin staining along bilateral cerebral sulci, cerebellar folia and pial surfaces of brainstem suggesting SS [Figure 1c, d]. Right transverse and sigmoid sinuses were not visualised in MR venography and

intracranial time-of-flight MR angiography (TOF MRA) was normal. Digital subtraction angiography ruled out vascular malformations except for hypoplastic right transverse and sigmoid sinuses. Lumbar puncture showed xanthochromic cerebrospinal fluid (CSF) with an opening pressure of 18 cm of water. CSF protein was elevated (848 mg/dl) with normal cytology and sugar. Normal CSF opening pressure and elevated CSF protein in the context of raised intracranial tension were suggestive of Froin's syndrome.

For etiological workup, the patient underwent an MRI spine which revealed a large expansile intradural lesion in the lumbosacral spinal canal with multi-focal enhancing lesions along the conus and filum terminale. The largest lesion at the level of L4-S1 showed 'hemosiderin cap sign' [Figure 2a, b]. Following surgical resection, histopathological examination revealed a glial neoplasm composed of elongated, glial fibrillary acidic protein (GFAP)-positive cells with monomorphic nuclei dispersed in abundant myxoid stroma [Figure 2c, d]. Fresh haemorrhage, hemosiderin pigment and Gamma-Gandy bodies were also noted in the stroma. The tumour was diagnosed as myxopapillary ependymoma, WHO Grade I. The patient was asymptomatic on follow-up after 2 months.

Ependymomas are the most common intramedullary spinal cord tumours in adults. Myxopapillary ependymomas occur almost exclusively in the conus medullaris and filum terminale. They are believed to arise from the ependymal glia of the filum terminale.<sup>[2]</sup> The most commonly reported symptoms



**Figure 1:** T2 axial (a) and sagittal fluid attenuation inversion recovery (FLAIR) (b) images show prominent peri-optic subarachnoid space, flattening of the posterior sclera and partial empty sella suggestive of raised intracranial pressure. Susceptibility weighted images (SWI) (c, d) show extensive leptomeningeal hemosiderin staining along bilateral cerebral sulci, cerebellar folia and pial surface of the brainstem suggestive of superficial siderosis



**Figure 2:** T2 sagittal (a) and post-contrast T1 sagittal (b) spine images show large expansile intradural lesion widening and scalloping the lumbosacral spinal canal at L5-S1 level with multifocal enhancement along the conus and filum terminale. Glial neoplasm with elongated tumour cells dispersed in myxoid stroma (c) and expressing glial fibrillary acidic protein (d) [c: haematoxylin and eosin, d: immunoperoxidase, original magnification: 100 $\times$ , scale bar: 100  $\mu$ m]

are numbness or paresthesias (67%), back pain (58%) and weakness (51%). Because they are slow growing, the diagnosis is often delayed for over 2 years from symptom onset.<sup>[3]</sup> They are soft, encapsulated, 'sausage-like' vascular masses with a propensity for recurrent haemorrhage. Blood products may produce a characteristic intratumoural 'cap sign' or spill within the subarachnoid compartment leading to SS.<sup>[2]</sup>

Detection of SS warrants evaluation of the cause. In a review of 63 SS cases, an underlying cause could be identified in one-half. A dural pathology (CSF cavity lesions or root pathology) was the most common aetiology followed by tumours and vascular abnormalities. Cauda equina ependymoma was the aetiology in 12% of cases.<sup>[1]</sup> The clinical signs and symptoms of SS are dictated by the anatomical distribution of hemosiderin deposits within the brain. Gait and stance ataxia result from dysfunction of cerebellar vermis. The vestibulocochlear nerve is commonly involved at the pontocerebellar angle due to its long glial course and tendency to take up iron.<sup>[4,5]</sup> Chronic-increased intracranial pressure can occur in about a third of patients with SS secondary to obstruction of CSF absorption into the arachnoid granulations by the hemosiderin deposits.<sup>[1,4]</sup>

Our case presented with visual complaints alone, mimicking IHH which is rare both for spinal myxopapillary ependymoma and SS. MRI of brain showed features of raised intracranial pressure and extensive (supra and infratentorial) SS. The presence of SS with CSF picture suggestive of Froin's syndrome raised the suspicion of a spinal tumour in this case which was confirmed with MRI of the spine.

SS is a rare neurological condition which can complicate spinal tumours and can be missed without specific imaging. SS can present with visual complaints alone secondary to raised intracranial pressure and SWI should be included in the standard MRI sequences with additional spinal imaging in patients with unexplained papilloedema.

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### Declaration of patient consent

The authors certify that they have obtained all appropriate

patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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