Spinal ependymoma complicated by superficial siderosis

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DESCRIPTION

A 64-year-old woman presented with progressive hearing loss. A pure-tone audiography confirmed bilateral high-frequency sensorineural hearing loss. Her medical and surgical history was unremarkable except for long-standing back pain. Neurological examination revealed mild dysmetria and gait ataxia. There was no leg weakness or sphincteric dysfunction.

MRI revealed superficial siderosis lining the cerebellum and brainstem, which prompted further imaging of the entire neuraxis in order to exclude an underlying cause. A myxopapillary ependymoma seen at the level of the conus medullaris was felt to be responsible for recurrent haemorrhage within the subarachnoid space.

Excision of the spinal ependymoma was performed and the patient regained near-normal hearing following a cochlear implant.

DISCUSSION

Superficial siderosis presents with slowly progressive sensorineural hearing loss, gait ataxia and cerebellar dysarthria. Involvement of other cranial nerves has also been described. Clinical history and examination often suggest a neurodegenerative disorder.¹

Superficial siderosis results from haemosiderin deposition in the subpial layers of the neuraxis, and complicates chronic haemorrhage within the subarachnoid space. The posterior fossa is preferentially affected, which may in part be explained by the presence of Bergmann glia in the cerebellum that display increased ferritin synthesis.² The long glial segment of the vestibulocochlear nerve makes it more vulnerable to axonal damage by iron deposition.¹ Bilateral sensorineural hearing loss is present in 95% of affected individuals.³

Figure 1 Linear hypointensity is seen along the cerebellar folia and superior vermis on the axial gradient echo images (A) compatible with extensive haemosiderin deposition. There is also hypointense lining of the midbrain tectum and pons. The sagittal T1-weighted image (B) reveals low signal intensity of the upper cerebellum and superior vermis. The linear hypointensities are also appreciated on the axial T2 sequence (C) where they clearly outline the cerebellar folia and ventral pons and reflect the characteristic subpial haemosiderin deposition. Mild underlying cerebellar atrophy is also appreciated. The radiological findings are pathognomonic of superficial siderosis.

Figure 2 Sagittal T2-weighted image shows an oval-shaped heterogeneous mass at the level of L2 vertebral body. The mass is intradural and arises from the filum terminale. A subtle hypointense rim along the caudal aspect of the lesion and coarse hypointense foci at the inferior tumoral pole (arrows, A) represent blood products and are frequent findings in spinal ependymomas. The lesion enhances homogeneously after contrast administration (B, precontrast T1; C, postcontrast T1) and reflects the hypervascular nature of the tumour. (D) Clearly shows the intradural location of the tumour. The diagnosis of myxopapillary ependymoma was confirmed pathologically after resection.

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MRI is the investigation of choice. T2-weighted imaging reveals linear hypointensity outlining the brainstem, cerebellar folia (figure 1) and the pial surface of the cord. Less commonly haemosiderin deposition is seen lining the Sylvian fissures, the cerebral sulci and cauda equina roots. Gradient echo imaging is exquisitely sensitive as the haemosiderin produces blooming artefacts. Imaging of the entire neuraxis should be performed to rule out underlying neoplasms, arteriovenous malformations/fistulas and dural defects.

Superficial siderosis complicating spinal ependymoma has been reported in the literature.³ Ependymomas present as soft, encapsulated 'sausage-like' masses with a propensity to haemorrhage (figure 2). Blood products may produce a characteristic intratumoral 'cap sign' or spill within the subarachnoid compartment.

Learning points

- ▶ Superficial siderosis presents with slowly progressive sensorineural hearing loss, gait ataxia and cerebellar dysarthria.
- ▶ Superficial siderosis complicates chronic haemosiderin deposition in the subpial layers of the neuraxis and the imaging findings are characteristic.
- ▶ Imaging of the entire neuraxis should be carried out to exclude underlying neoplasia.

Competing interests None.

Patient consent Obtained.

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