Superficial siderosis of the central nervous system

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DESCRIPTION

A 58-year-old man presented with insidious onset of sensorineural bilateral deafness, cerebellar ataxia, dysarthria, dizziness and mild cognitive impairment for 2 years. Five years prior, he had suffered an accidental fall. The diagnosis of superficial siderosis was made by brain and spine MRI, which revealed a hallmark T2 hypointensity surrounding cerebellar folia (figure 1), brain stem and spinal cord surface (figure 2), corresponding to hemosiderin deposition. There was also a superior pattern of cerebellar atrophy and subcortical gliosis (figure 1A), frequently associated with this condition. An intraspinal fluid-filled collection (figure 2A), and clumping and peripheralisation of cauda equina

nerve roots (figure 2B) suggested a dural defect and arachnoiditis, respectively. The patient was symptomatically treated with betahistine, with mild relieve of dizziness.

Superficial siderosis of the central nervous system is an underestimated entity that results from chronic/recurrent haemorrhage (namely, arteriovenous malformations, tumours or trauma), which leads to the accumulation of cytotoxic hemosiderin in the subpial layers of the brain and spinal cord.^{1 2} In this case, the traumatic event was the possible aetiology, since other possible causes were excluded by MRI.

The typical presentation is slowly progressive adult onset of hearing loss, cerebellar dysfunction

Figure 1 Brain MRI. (A) Coronal T2 showing hypointensity surrounding cerebellar folia (arrows), marked superior cerebellar atrophy (arrowheads) and subcortical gliosis (stars). (B) Sagittal T2 showing hypointensity delineating brain stem (arrows) and superior vermician atrophy (arrowheads).

Figure 2 Spine MRI. (A) Sagittal T2 depicting fluid-filled intraspinal collection. (B) Axial T2 showing peripheral and clumped cauda equine roots.

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with dysarthria and ataxia, cognitive impairment and, eventually, myelopathy. The preferential VIII nerve impairment is

Learning points

- ▶ MRI of the brain and spine is the investigation of choice for diagnosis of superficial siderosis.
- ▶ Superficial siderosis is due to toxic hemosiderin deposition in the subpial layers of brain and spinal cord because of chronic/recurrent haemorrhages.

probable due to the greater length of hemosiderin deposition, since it has a long glial segment. MRI of the brain and spine is the investigation of choice and, whenever possible, removing the bleeding source is the indicated treatment.^{1 2}

Competing interests None declared.

Patient consent Obtained.

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