Images in Otolarinology

Neurosensorial deafness: Superficial siderosis of the central nervous system

Hipoacusia neurosensorial: hemosiderosis superficial del sistema nervioso central

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We present the case of a 63 year-old man with a clinical history of lymphoma in complete remission and cervical hernia surgery 18 years previously. Following two transitory ischemic cerebrovascular accidents, he was admitted for study.

Cerebral MR showed striking hypointense subarachnoid lineal deposits, especially in the magnetic susceptibility sequence. These are compatible with haemosiderin deposits (the blue arrows) in the cerebral folia, quadrigeminal plate and protuberance (Fig. 1), and in the cisternal portion of both vestibulo-ocular cranial pairs (Fig. 2).

In the craniocervical junction (Fig. 3, sagittal sequence amplified in T2), the deposits of haemosiderin (blue arrows) are visible in the cerebral folia, the quadrigeminal plate, the protuberance and frontal surface of the spinal cord. C5 and C6 laminectomies are visible in this sequence together with an associated postoperative pseudomeningocele (red arrow).

Given these radiological findings the clinical history of the patient was revised. He reported neurosensorial deafness and instability developing over more than 15 years, while the
superficial haemosidosis of the central nervous system was considered to be secondary to the postoperative cervical pseudomeningocele. This rare disease is due to deposition of haemosiderin in the pia mater, causing progressive neurological dysfunction: bilateral progressive neurosensory deafness (95%) and cerebral ataxia (88%). Other symptoms are: dementia, cephalgia, urinary incontinence and anosmia.

The origin of this disease lies in recurrent bleeding into the subarachnoid and sub pia mater spaces, in connection with arteriovenous malformations, traumatisms, previous surgery and tumours. Intracellular captation of iron occurs, giving rise to cerebral parenchymatous damage in the glial tissue.