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Tinnitus as the presenting symptom in a case of Lhermitte-Duclos disease

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Abstract
Dysplastic gangliocytoma or Lhermitte-Duclos disease (LDD) is a hamartomatous malformation of the cerebellar hemisphere that usually presents with signs of increased intracranial pressure or symptoms of cerebellar dysfunction. In this paper, we report a case of LDD presenting with tinnitus, and postulate a probable mechanism for this unusual presentation.

Key words: Cerebellar diseases; Tinnitis

Introduction

Dysplastic gangliocytoma of the cerebellum or Lhermitte-Duclos disease (LDD) was first described in 1920 (Lhermitte and Duclos, 1920). The pathogenesis of this rare condition is thought to be the result of a congenital abnormality in granule-cell migration leading to a hamartomatous malformation of the cerebellar hemisphere (Roski *et al.*, 1981). The cerebellum appears enlarged with pale and prominent folia. Histologically there is a hypertrophy of the granular cell neurones and an increase in the myelination of the axons in the molecular layer with a loss of the Purkinje cells and the central core of the affected cerebellar folia (Reznik and Schoenen, 1983). More recently there have been cases reported of LDD occurring in adult patients with Cowden's disease (multiple hamartoma syndrome) suggesting that it might represent one of the phacomatoses (Wells *et al.*, 1994; Liaw *et al.*, 1997).

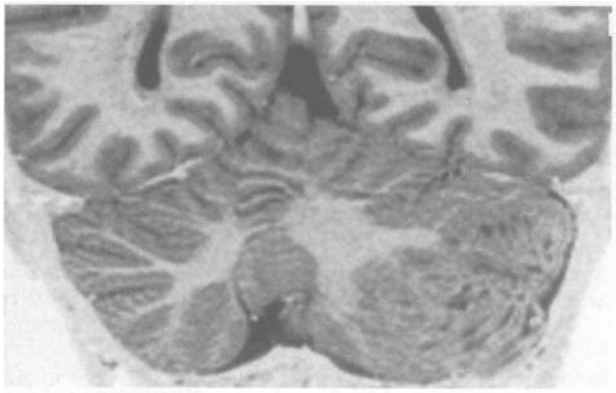

Fig. 1
Coronal inversion recovery sequence image through the cerebellum showing dysplastic folia.

Case report

A 50-year-old white Caucasian male was referred by his general practitioner with high pitched tinnitus and sensation of blockage in his left ear. An ENT examination was unremarkable and normal hearing thresholds and speech discrimination were obtained on pure tone and speech audiometry. However, there were mild left-sided cerebellar signs of dysdiadochokinesia and dysmetria. No other neurological deficits were detected. In view of the cerebellar signs, magnetic resonance imaging (MRI) was performed to identify any central lesion.

MRI findings

Axial T2 (T_E 120 ms, T_R 4158 ms), pre- and post-contrast T1 (T_E 20 ms, T_R 641 ms), and coronal inversion recovery (T_E 24 ms, T_R 2811 ms, and T_I 300 ms) sequences were performed (T_E = time to echo, T_R = time to repetition, T_I = time to inversion). Scans demonstrate left cerebellar hemisphere enlargement with a poorly demarcated mass-like thickening of the cerebellar folia (Figures 1 and 2). These folia have a characteristic laminated pattern of increased signal on T2W images diagnostic for LDD. The mass effect of this lesion has produced clockwise rotation of the brainstem with stretching of the VIIth and VIIIth nerve complex in the cerebello-pontine angle cistern. A vascular loop is seen in contact with the VIIIth nerve in the cistern (Figure 3).

Discussion

Lhermitte-Duclos disease is a rare entity, which has been well described in the literature. Patients usually present in young adult life with a median age of 34 years (± 14 years). There is however, a wide range at presentation from new-born to 74 years. This condition usually presents with signs of raised intracranial pressure secondary to mass effect. Cerebellar signs are minimal and may be absent in half of the cases; whilst only rarely are cranial nerve deficits or long tract signs present (Milbouw *et al.*, 1988; Vinchon *et al.*, 1994; Kulkantrakorn *et al.*, 1997).

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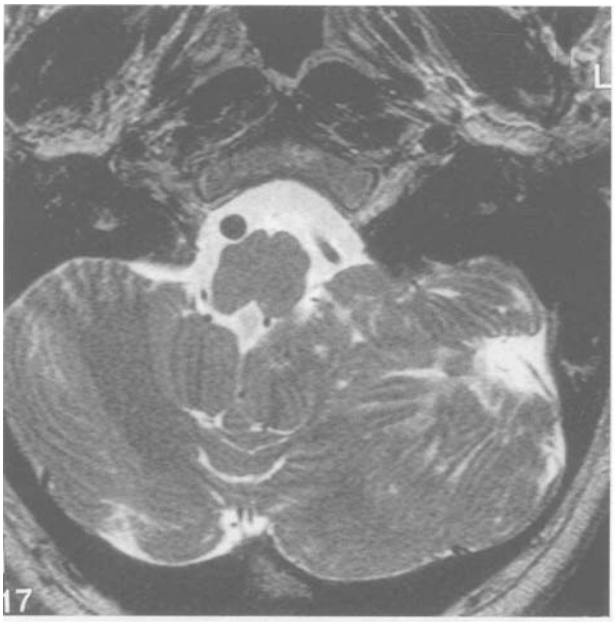

FIG. 2
T2-weighted axial MRI scan of cerebellum shows the laminated pattern of increased signal.

In this case the left vestibulocochlear nerve is seen to be stretched due to a clockwise rotation of the brainstem secondary to mass effect from the dysplastic gangliocytoma within the left cerebellar hemisphere. A vascular loop is also noted to run across this stretched nerve (Figure 3). We propose that the tinnitus may be a result of neural irritation due to a combination of a traction injury to the nerve and compression by a previously asymptomatic vascular loop. The absence of any associated hearing loss suggests minimal damage to cochlear nerve.

Support for such a theory is provided in previous reports of traction injury to the vestibulocochlear nerve secondary to syringobulbia (Morgan and Williams, 1992) and the Arnold-Chiari malformation (Ahmmed *et al.*, 1996). In these cases, the tinnitus was relieved following surgical decompression. Although there is widespread scepticism about the clinical significance of vascular loop compression, there has been histological evidence of compressive injury to the VIIIth nerve by adjacent vascular loops (Herzog *et al.*, 1997).

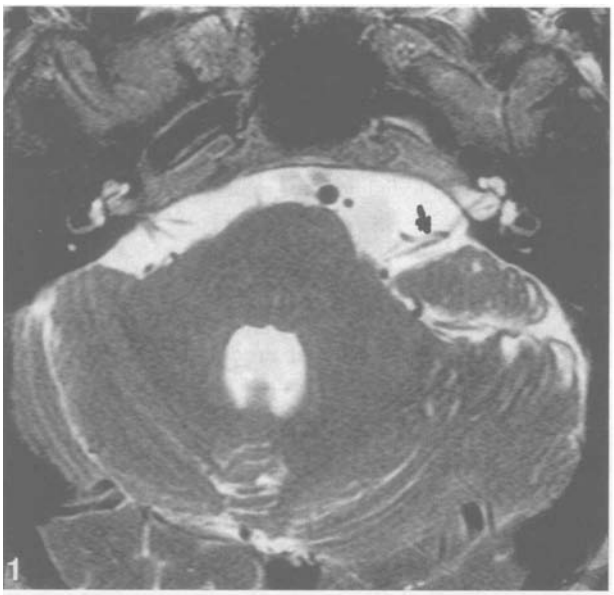

FIG. 3
T2-weighted axial MRI scan showing the vascular loop (arrow) in contact with VIIIth nerve.

The characteristic appearance of the cerebellum on the MRI in this case obviates the need for histological confirmation of this hamartomatous condition (Kulkantrakorn *et al.*, 1997).

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