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Sudden deafness and cerebellar tumour

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Abstract
We report the case of a female patient who presented with sudden deafness as the first symptom of a cerebellar tumour which was not localized strictly in the cerebellopontine angle and did not show direct compression on the extrabulbar portion of the VIIIth cranial nerve. The clinical picture contained a number of signs and symptoms typical of cerebellar involvement.

Surgical intervention restored the hearing and caused the symptoms to disappear.

We also review the association between tumours and sudden deafness in the literature.

Key words: Deafness, sensorineural, sudden; Cerebellar neoplasms, medulloblastoma

Introduction

Sudden deafness, whether or not accompanied by vertigo, is a relatively infrequent occurrence which causes the patient great anxiety and presents the physician with a diagnostic and therapeutic problem.

Whilst in the majority of patients no cause is found, there are a small number of patients in whom the deafness is a symptom of an underlying process such as a tumour in the bulbocerebellopontine angle. When we speak of angle pathology we tend to think of sudden deafness as the initial symptoms of acoustic neuromas (Higgs, 1973; Neely, 1981) but we must not forget that there are other processes that can first present in this way.

We report the case of a female patient with a cerebellar tumour who presented with sudden deafness as the first symptom.

Case report

A 25-year-old female, with no relevant personal history, presented with left-sided sudden deafness of two months duration attributed to a common cold.

The patient said that two weeks after the onset of the sudden deafness she had started to suffer from a persistent sensation of rotatory vertigo accompanied by nausea, vomiting, unsteadiness and difficulty in concentrating. She also complained of left-sided facial hypoaesthesia which had started with the onset of deafness, and of nonlocalized slight cephalalgia of one-year's standing. She did not mention tinnitus or otalgia.

On otorhinolaryngological examination, otoscopy was normal. Tuning-fork testing showed severe left-sided sensorineural hypoacusis. Spontaneous nystagmus was not seen.

Examination of the cranial nerves showed a left-sided facial hypoaesthesia and an area of anaesthesia in the left external auditory canal. The patient showed a positive Romberg sign and a deviation to the right when walking. The fundi were normal.

Pure tone audiometry showed a left-sided sensorineural hypoacusis (Figure 1). Impedance audiometry revealed a normal curve on both sides. The ipsilateral acoustic reflex was normal on the right side but absent on the left; the contralateral acoustic reflex was normal on the left and absent on the right. Békésy audiometry was type I and the tone decay test was negative. Electronystagmography showed a spontaneous nystagmus to the right and saccadic pursuit. Brain stem evoked response audiometry (BERA) on the left side showed retrocochlear hypoacusis. We observed preservation of waves I and II and disappearance of the remainder (Figure 2).

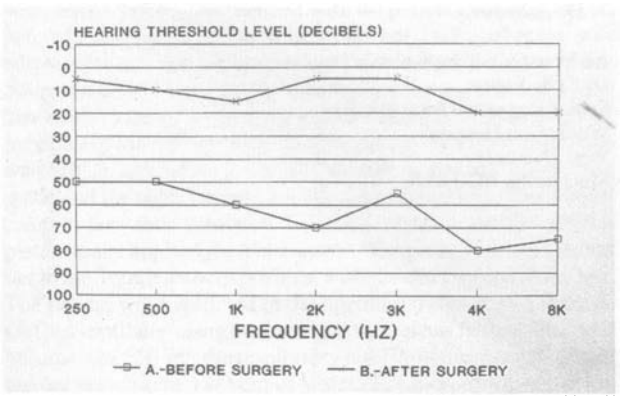

Fig. 1
Left side audiogram showing a substantial hearing level increase after surgery.

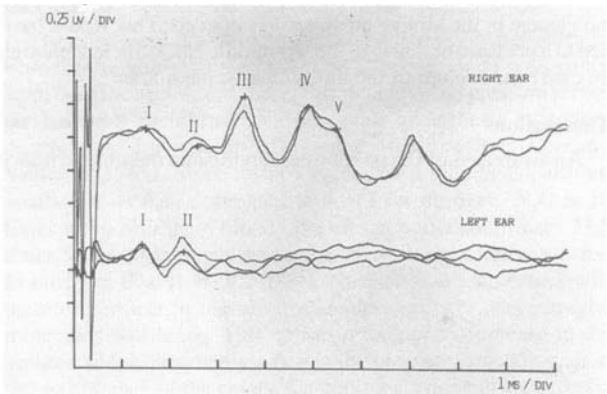

Fig. 2
Pre-surgical BERA register on both sides.

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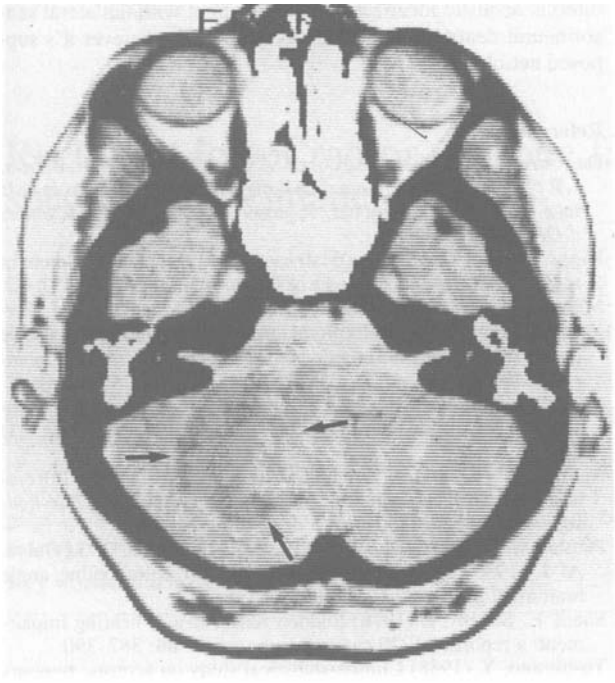

FIG. 3

Axial computed tomogram (CT), using contrast material, showing a mass of irregular density (arrowed) in left side of the cerebellar hemisphere. Internal auditory canals are of normal size and morphology.

A CT study showed a mass of irregular density in the left cerebellar hemisphere, displacing the fourth ventricle to the right. After administration of contrast material, a homogeneous enhancement of the entire tumour was seen, with peripheral oedema. The internal auditory canals appeared normal (Figure 3).

As a result of the clinical picture, the patient was referred to the neurosurgical department of this hospital where she underwent a suboccipital craniectomy with atlas laminectomy. A well-delineated tumour in the lower half of the left cerebellar hemisphere was removed. The histological diagnosis was cerebellar desmoplastic medulloblastoma (Figure 4).

Post-surgical follow-up has been satisfactory, with recovery of normal hearing (Figure 1).

Discussion

Neely (1981) describes four aetiopathogenic theories which may explain sudden deafness when there is a tumour in the internal auditory canal or in the cerebellopontine angle: (i) compression on the VIIIth cranial nerve; (ii) vascular involvement; (iii) biochemical changes in the inner ear; and (iv) diminution of the total number of auditory nerve fibres.

If there is a tumour of the cerebellopontine angle we immediately think of compression mechanisms either of the nerve fibres themselves or of the vessels that feed them. Vascular obstruction would have caused an early and permanent loss of neuronal function and therefore in our patient the indications pointed to nerve compression, either extra- or intraencephalically.

In Figure 3 we can see the internal auditory canals apparently compression-free and consequently it would be reasonable to presume the existence of compression at a point cephaled to the

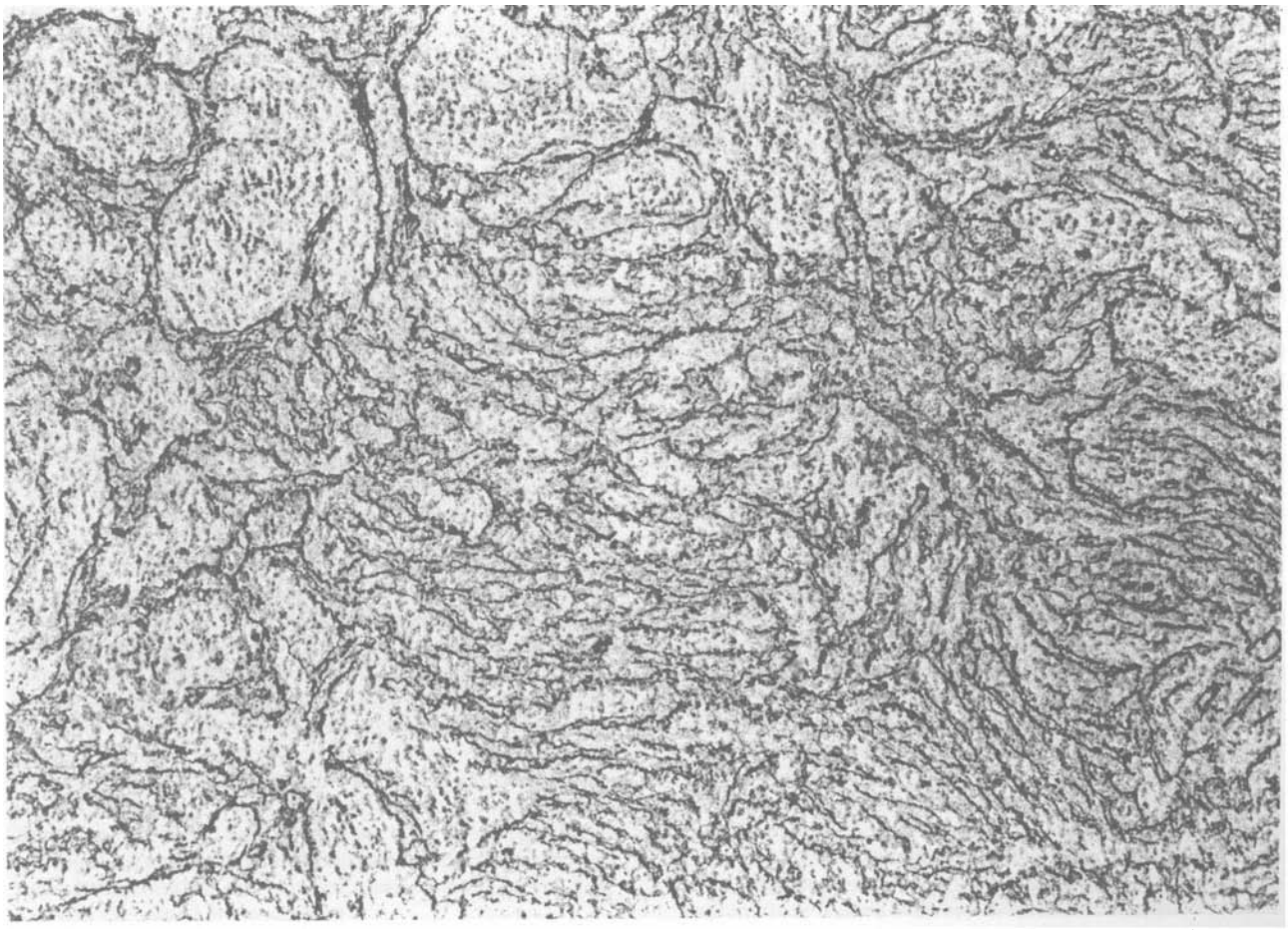

FIG. 4

Low power silver reticulin stain micrograph showing desmoplastic pattern of desmoplastic modulloblastoma.

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cochlear nuclei, or at the start of the acoustic striae and trapezoid body.

In the literature we have been unable to find the description of any case of a tumour of cerebellar origin with sudden deafness as its first symptom. In the great majority of cases sudden deafness, as the first symptom, announces an acoustic neuroma in the cerebellopontine angle. Other much less frequent pathologies, such as meningioma (Darrouzet *et al.*, 1989), angle cholesteatoma (Pensak *et al.*, 1985) and epidermal cysts (Marquez *et al.*, 1988), also exist where sudden deafness is the first symptom, whilst other angle tumours, such as glomus jugulare, facial neurofibroma and arachnoid cysts, have presented only as progressive loss of hearing (Nedzelski and Tator, 1982).

Acoustic symptomatology was similar to that described in the literature for angle tumours. Both sensorineural hypoacusis and absence of acoustic reflex are to be expected in this type of pathology, and the absence of recruitment pointed us towards the cause of a retrocochlear location.

The pattern of the BERA in the left ear was clearly pathological, with only the first two waves being of normal latency, and this also pointed to a retrocochlear origin. In the literature consulted, we have been unable to find this type of pattern. In their series Pensak and his coworkers describe three cases with patterns conserving waves I–II, but with prolongation of their latency (Pensak *et al.*, 1985).

The presence of a spontaneous nystagmus, together with the presence of saccadic movements and dysmetric saccades in the electronystagmograph suggested a central pathology. Left-sided facial paraesthesia and anaesthesia in the left-sided external auditory canal are indicative of Vth cranial nerve involvement.

Whilst the tentative diagnosis was made by means of the acoustic tests, principally the BERA, it was the CT scan which gave definitive confirmation in this case, as happens in the majority of cases (Yoshimoto, 1988).

Finally, one must always remember the dictum of Shaia and Sheely (1976) arrived at after the study of more than 1000 cases of sudden deafness, 'never discount the presence of a tumour of internal acoustic localization in any patient with unilateral sensorineural deafness, however this starts and whatever it's supposed aetiology'.

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