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Clinical Record

Glioneural hamartoma of the VIIIth nerve

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Abstract
Hamartomas of the cerebellopontine angle or internal auditory canal are very rare and only four cases have been reported. We report an unusual case of a glioneural hamartoma of the VIIIth nerve with clinical, radiological and audiometric similarity with vestibular schwannoma.

Key words: Hamartoma; Cochlear Nerve; Magnetic Resonance Imaging

Introduction

Hamartomas are not true neoplasms and develop from pluripotential neural crest cells, which are capable of differentiating into a variety of neuroectodermal and mesenchymal cell types.[1] During the third week of gestation, the midline embryonic ectodermal germ cell layer thickens and forms the neural plate, which later invaginates to form the neural groove and two neural folds, located one on each side of the groove. The neural folds fuse to form the neural tube and neural crest.[2,3] Abnormal embryogenic development during arrangement of the neural tube and neural crest is assumed to cause hamartomas, non-neoplastic lesions caused by abnormal migration of the cells. Hamartomas with mesenchymal and epithelial components are uncommon, forming via benign, tumour-like, erroneous tissue development and usually presenting with well demarcated, yellowish, rubbery and solitary masses of enlarged and hypertrophied mature myocytes, collagen and adipose tissue.

Hamartomas of the cerebellopontine angle (CPA) or internal auditory canal (IAC) are rare. Glasscock *et al.* reported one case of tumour composed of ganglion and fat cells, which was designated as a ganglioneurolipoma, in a series of 80 CPA tumours.[4] Nearly 90 per cent of IAC and CPA tumours are acoustic neuronomas. It has been estimated that intracranial hamartomas account for only 0.1 per cent of all central nervous system tumours.[5] Glioneural hamartomas of the VIIIth nerve are uncommon and only four cases have been reported. We report an unusual case of a glioneural hamartoma of the VIIIth nerve with clinical, radiological and audiometric similarity with vestibular schwannoma.

Case report

A 34-year-old man presented with unsteadiness and unilateral left-sided hearing loss of one year's duration. He had had two bouts of acute-onset vertigo before and intermittent tinnitus on the left for six months. Neurological examination revealed nothing remarkable.

The otoscopic view was normal on both sides, and otoacoustic emission and impedance studies were normal. Audiometric evaluation showed bilateral down-sloping sensorineural hearing loss (Figure 1). Speech discrimination score was 100 per cent on the right and 92 per cent on the left. Analysis of auditory brainstem responses at 90 dB click stimulation with alternating polarity and 2000 sweeps demonstrated that I–III, III–V and I–V interpeak latencies were 2.03, 2.07 and 4.10 μV on the right and 2.20, 2.47 and 4.67 μV on the left, respectively (Figure 2). Magnetic resonance imaging (MRI) of the brainstem demonstrated an iso/hypointense, well demarcated, solid, homogenous mass in the most lateral part of the left IAC (Figure 3). Magnetic resonance imaging with gadolinium contrast demonstrated a 6 × 7 mm, tiny, slightly enhancing mass within the IAC.

The patient underwent suboccipital retrosigmoid craniotomy with complete removal of the tumour. Post-operative neurological examination found normal facial nerve function, with preservation of hearing. Post-operative follow up over six months was unremarkable.

Histopathologic examination demonstrated that the tumour was composed of fibrous tissue containing spindle-shaped cells, myelinated nerve fibres, isolated and mononucleated mature ganglion cells, and well differentiated smooth and striated muscle fibres. There were no mitotic figures (Figure 4). Immunohistochemical staining for S-100 protein showed a normal pattern of immunoreactivity in the neuronal cell bodies.

Discussion

The histogenesis of germ cell tumours is controversial. There is a morphologic heterogeneity and difficulty in classifying germ cell tumours, which explains the use in reports of such different terminologies as hamartoma, choristoma, mesenchymoma, ectomesenchymal hamartoma, neuromesenchymal hamartoma, neuromuscular hamartoma, ganglionic hamartoma, glioneural hamartoma, benign triton tumour and neurocristopathy. A hamartoma is an abnormal collection and arrangement

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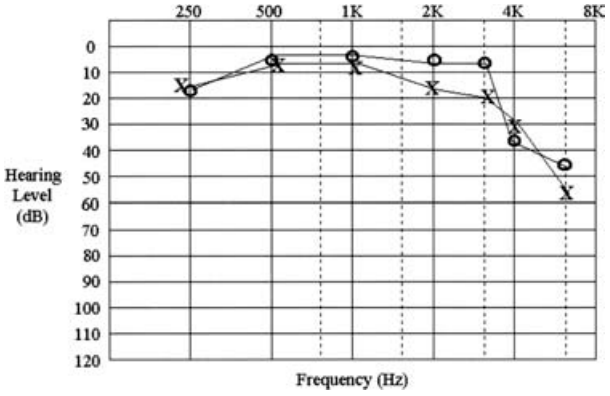

FIG. 1

Pure-tone audiogram (American National Standards of Institute (ANSI)) of the patient, demonstrating mild high-frequency hearing loss on both sides. X = air conduction threshold on the left ear; O = air conduction threshold on the right ear.

of mature tissue elements and cells that are normally present in the tissue from which they arise. A choristoma implies the presence and collection of cells that are normally not found at the tumour site, which seems to be a more proper term.[6] A diagnosis of 'hamartoma', however, was preferred in this case because of the complete absence of cytological features of neoplasia and the presence of mixed tissue elements. Intracranial hamartomas may be divided into lipomatous and glioneural types.[5,7] Lipomatous hamartomas of the central nervous system are usually found in the region of the corpus callosum, hypothalamus and quadrigeminal plate.[8] Lipomas are rarely found at the CPA and IAC. Hamartoma of the IAC was first described by Legent *et al.* in a 14-year-old child with progressive facial paralysis.[9] Bigelow *et al.* reviewed 84 reported patients with lipoma of the VIIIth nerve, including their largest series of 17 cases in 1998.[10] Glioneural hamartoma of the VIIIth nerve is very infrequent and only four cases have been reported.[5,7,11–13]

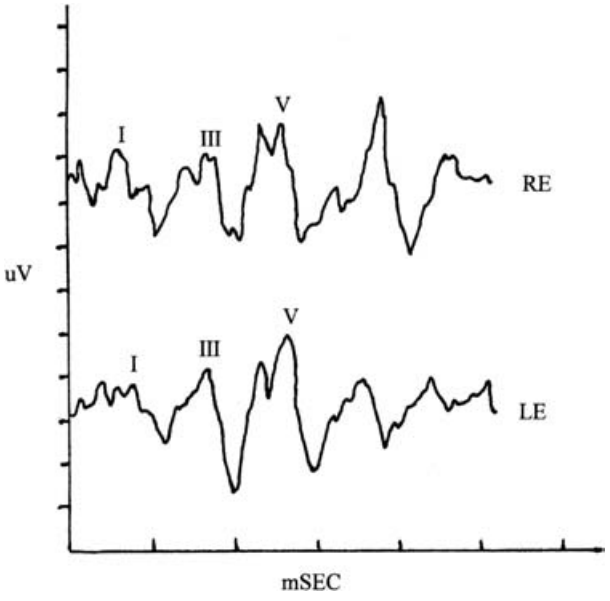

FIG. 2

Auditory brainstem response analysis for the patient shows inter-peak latency prolongation on the left (LE) as compared with the right (RE).

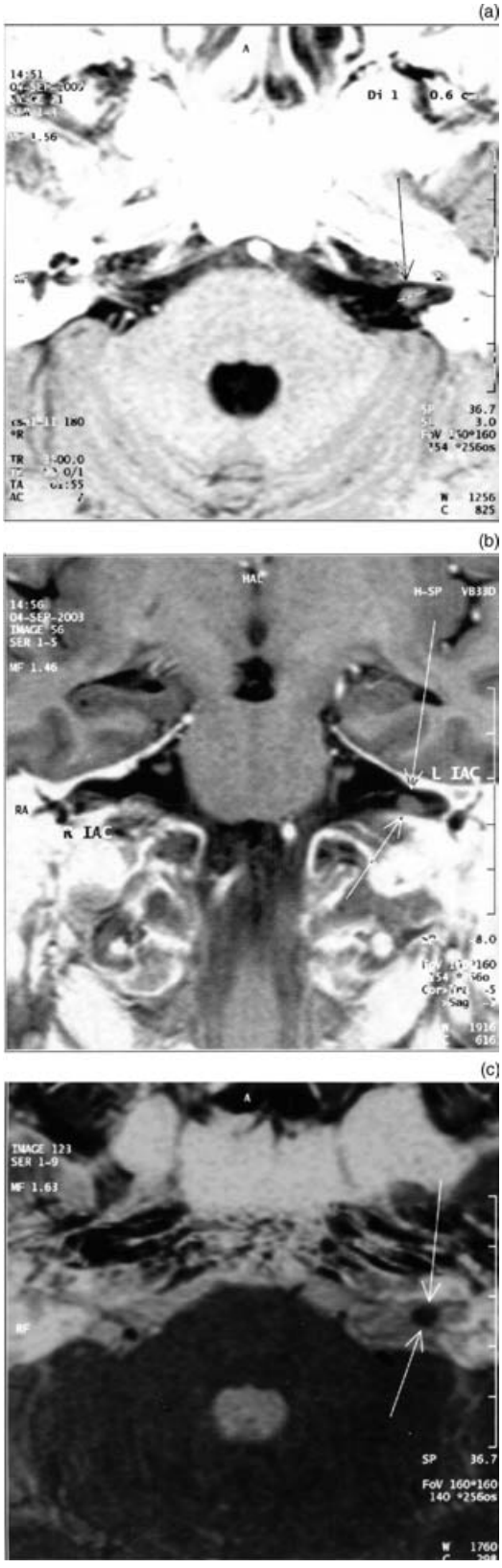

FIG. 3

Magnetic resonance imaging of the temporal bone demonstrates a soft tissue mass within the internal auditory canal. (a) T1-weighted spin echo axial scan demonstrates an oval, isointense lesion (long arrow) in the internal auditory canal (line indicates long axis). (b) After contrast administration, the lesion shows slight enhancement on coronal scan (two long arrows). (c) T2-weighted axial scan demonstrates hypointense lesion (marked with long arrow).

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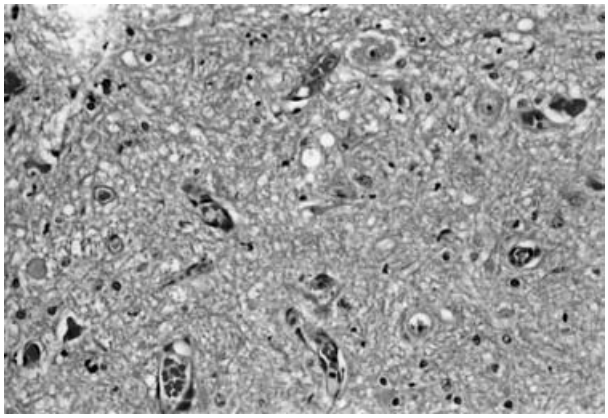

FIG. 4

Histopathologic examination demonstrates that the tumour is composed of fibrous tissue containing myelinated nerve fibres and isolated mature ganglion cells. No mitotic activity is seen (H&E; ×400).

Recent case reports have confirmed that hamartoma is an active, evolving disease, although use of the term 'tumour' for hamartoma has been questioned. On the other hand, the presentation of hamartomas of the VIIIth nerve is similar to that of vestibular schwannoma, being characterized by hearing loss, tinnitus, vestibular impairment and headache.[11] However, the initial symptom can be facial nerve dysfunction or trigeminal neuralgia, and accurate pre-operative diagnosis of hamartoma is often difficult.[9,14] These lesions can, rarely, mimic Ménière's disease, as also seen in the presented case.[15] Magnetic resonance imaging provides pre-operative diagnosis. Compared with normal grey matter and common lesions of the IAC, such as vestibular schwannoma and meningioma, hamartomas are iso/hypointense on non-enhanced T1- and T2-weighted images.[5,16] However, lipomas are hyperintense on T1-weighted images, with or without contrast enhancement.[7] Fat suppression of T1-weighted images on MRI offers the most precise pre-operative diagnostic tool for IAC lipomas.[17]

Pre-operative radiological differentiation of the lipomatous hamartoma of the VIIIth nerve is important, since the cochlear nerve is more in danger during surgical removal of IAC lipoma due to the fact that identification of tumour border is poor and there is always dense adherence to neurovascular structures. Tumoral infiltration of the nerve is more common. Bigelow *et al.* reported that total tumour removal was accomplished in only 33 per cent of patients with lipoma of the VIIIth nerve.[10] Tankere *et al.* reviewed 98 cases, including their four new ones, and reported that total tumour removal was possible in 32.8 per cent of patients.[18] They proposed conservative follow up for patients with a confirmed radiological diagnosis of lipoma, unless significant progressive or disabling symptoms were present, because of the potential for significant morbidity with resection of these lesions. In the case of glioneural hamartoma presented here, total tumour removal and preservation of hearing was possible. However, no comparison can be made regarding surgical compromise of cochlear and facial nerve function because of the rarity of these lesions.

In conclusion, we report a case of IAC hamartoma presenting as an acoustic neuroma. Cerebellopontine angle glioneural hamartomas are unusual lesions, more exceptional than other types of hamartomatous lesions. More knowledge of these tumoral lesions and of their close relationships with cranial nerves will enable establishment of a comprehensive strategy for treatment of IAC tumours, reducing post-operative functional sequelae.

- **This case report describes a glioneural hamartoma of the VIIIth nerve. Clinically, this lesion mimicked an acoustic neuroma, presenting with unilateral hearing loss and unsteadiness**
- **Radiologically, magnetic resonance contrast-enhancement of glioneural hamartomas is poor compared with that shown by acoustic neuromas**
- **These tumours are more infiltrative than schwannomas, making total tumour removal more difficult**

References

1 Apostolides PJ, Spetzler RF, Johnson PC. Ectomesenchymal hamartoma (Benign ectomesenchymoma) of the VIIIth nerve: case report. *Neurosurgery* 1995;**37**:1204–1207
2 Le Douarin N. Migration and differentiation of neural crest cells. *Curr Top Dev Biol* 1980;**16**:31–85
3 Selleck MAJ, Scherson TY, Bronner-Fraser M. Origins of neural crest cell diversity. *Dev Biol* 1993;**159**:1–11
4 Glasscock ME, Hays JW, Murphy JP. Complications in acoustic neuroma surgery. *Ann Otol Rhinol Laryngol* 1975;**84**:530–40
5 Carvalho GA, Matthies C, Osorio E, Samii M. Hamartomas in the internal auditory canal; report of two cases. *Neurosurgery* 2003;**52**:944–9
6 Wu SS, Lo WW, Tschirhart DL, Slattery WH 3rd, Carberry JN, Brackmann DE. Lipochoristomas (lipomatous tumors) of the acoustic nerve. *Arch Pathol Lab Med* 2003;**127**:1475–9
7 Palmer CA, Townsend JJ, Harnsberger HR, Parkin JL, Apfelbaum RI. Cerebellopontine angle glioneural hamartoma. *Surg Neurol* 1996;**45**:467–9
8 Pensak CA, Glasscock ME, Gulya AJ, Hays JW, Smith HP, Dickens JRE. Cerebellopontine angle lipomas. *Arch Otolaryngol Head Neck Surg* 1986;**112**:99–101
9 Legent F, Resche F, Beauvillain C, de Kersaintgilly A, Nomballis M, Nicolas R. Hamartoma of the internal auditory canal and cerebellopontine in a child. Transtemporo-extradural excision method. *Ann Otolaryngol Chir Cervicofac* 1979;**96**:291–3
10 Bigelow DC, Eizen MD, Smith PG, Yousem DM, Levine RS, Jackler RK *et al.* Lipomas of the internal auditory canal and cerebellopontine angle. *Laryngoscope* 1998; **108**:1459–69
11 Hung TY, Litofsky NS, Smith TW, Megerian CA. Ganglionic hamartoma of the intracanalicular acoustic nerve causing sensorineural hearing loss. *Am J Otol* 1997;**18**: 498–500
12 Babin RW, Fratkin JD, Cancilla PA. Hamartomas of the cerebellopontine angle and internal auditory canal. *Arch Otolaryngol* 1980;**106**:500–2
13 Ho KL. Concurrence of ganglionic hamartoma and heterotopic neuroglial tissue in the cerebellopontine angle. *Arch Pathol Lab Med* 1981;**105**:438–9
14 Goda M, Isono M, Karashima A, Kasai N, Kobayashi H. Hamartoma in the internal auditory canal. *J Clin Neurosci* 2003;**10**:111–13
15 Kitamura K, Futaki T, Miyoshi S. Fluctuating hearing loss in lipoma of the cerebellopontine angle. *ORL J Otorhinolaryngol Relat Spec* 1990;**52**:335–9
16 Freeman JL, Coleman LT, Wellard RM, Kean MJ, Rosenfeld JV, Jackson GD *et al.* MR Imaging and spectroscopic study of epileptogenic hypothalamic hamartomas: analysis of 72 cases. *Am J Neuroradiol* 2004;**25**:450–62
17 Hara A, Takahashi K, Ito Z, Kusakari J, Kurosaki Y. Value of fat suppression magnetic resonance imaging in

https://doi.org/10.1017/S0022215105009515 Published online by Cambridge University Press

the diagnosis of lipomas of the internal auditory canal. *Ann Otol Rhinol Laryngol* 1997;**106**:343–7
18 Tankere F, Vitte E, Martin-Duverneuil N, Soudant J. Cerebellopontine angle lipomas: report of four cases and review of the literature. *Neurosurgery* 2002;**50**:626–31

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