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Cavernous haemangioma of the internal auditory canal

A. M. Shaida, M.A., F.R.C.S., D. J. McFerran, M.A., F.R.C.S., M. da Cruz, F.R.A.C.S.,
D. G. Hardy, F.R.C.S., D. A. Moffat, M.A., F.R.C.S.

Abstract

Cavernous haemangiomas are rare lesions of the cerebello-pontine angle that can mimic the more commonly occurring vestibular schwannoma. A case report involving a patient with a cavernous haemangioma of the internal auditory canal (IAC) highlights this as a diagnostic possibility for lesions of the IAC by comparing and contrasting the clinical and radiological findings with the more commonly occurring vestibular nerve and facial schwannomas.

Symptoms such as hearing loss and facial paralysis that are disproportionate to the size of the lesion or fluctuate with hormonal changes such as those seen in pregnancy are suggestive of haemangioma. Radiological imaging demonstrating a lesion enchancing with gadolinium and containing areas of calcification is also suggestive of haemangioma. It is important to consider the possible diagnosis of haemangioma as early recognition of this entity may improve the chances of preserving the functional integrity of the facial nerve.

Key words: Haemangioma; Facial nerve; Hearing loss, sensorineural

Introduction

Cavernous haemangiomas of the internal auditory canal are rare lesions. Saleh *et al.*[1] found only 27 histologically proven cases in the world literature. The rarity of this pathology means information on the clinical and radiological features is scarce and these tumours can be difficult to differentiate from more commonly occurring lesions of the IAC such as vestibular schwannomas or facial schwannomas. The case report of a patient with a haemangioma of the IAC is presented to highlight this diagnostic possibility early in the management of patients with enhancing tumours of the IAC.

Case report

A 30-year-old female developed a progressive unilateral hearing loss during the third month of pregnancy. At eight months of pregnancy an ipsilateral facial weakness was noticed. This was accompanied by the development of tinnitus and unsteadiness. The facial weakness resolved spontaneously in the post partum period. Clinical examination at this stage revealed marked unsteadiness on tandem gait, a positive Unterberger's test with rotation to the right and a unilteral profound sensorineural hearing loss, confirmed on pure tone audiometry. Cranial nerve examination was unremarkable and in particular facial nerve function was normal.

A T1-weighted gadolinium DTPA-enhanced magnetic resonance image (MRI) scan was performed showing an enhancing lesion confined to the IAC consistent with an intra-canalicular vestibular schwannoma (Figure 1).

Surgical removal was planned via a translabyrinthine approach. At operation an intra-canalicular tumour was found occupying the lateral IAC compressing the facial nerve. The tumour was not particularly vascular and was resected with complete macroscopic clearance. At the conclusion of the resection, stimulation of the brainstem portion of the facial nerve (minimum stimulation current 0.15 mA) confirmed functional integrity. Although there was some initial facial weakness, at six months post-operatively the facial nerve function was normal (House grade 1).[2]

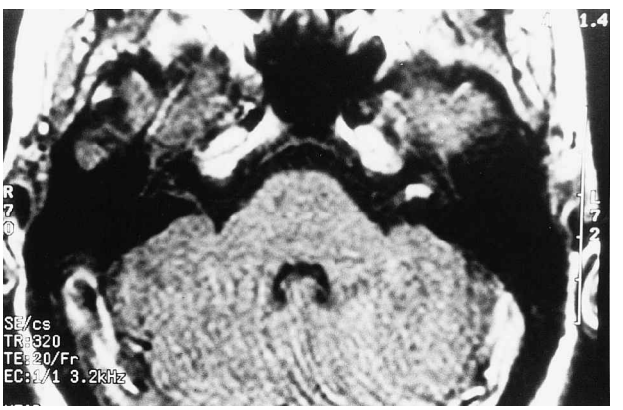

Fig. 1

Axial T1-weighted MRI scan of intracanalicular cavernous haemangioma, post-gadolinium DTPA.

Histological examination of the lesion demonstrated a vascular tumour composed of irregular, dilated vascular spaces with collagenous walls lined by a single layer of vascular endothelium (Figure 2). At one focus within the wall a small fragment of metaplastic bone was seen. There were no features to suggest malignancy. The appearances were those of a cavernous haemangioma.

From the Department of Otoneurosurgical and Skull Base Surgery, Addenbrooke's Hospital, Cambridge, UK.
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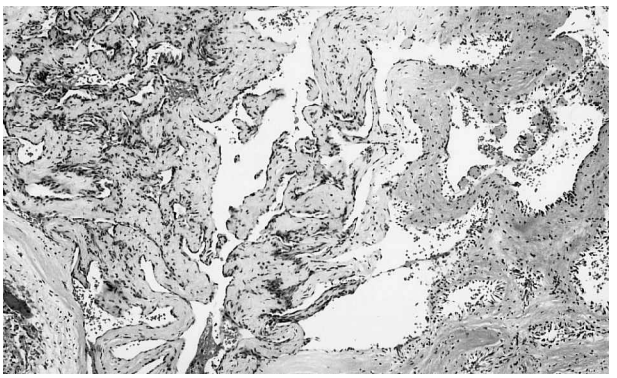

FIG. 2

Low-power view of a cavernous haemangioma, composed of dilated vascular spaces with collagenous walls, lined by a single layer of endothelium (H & E; ×35).

Discussion

Haemangiomas of the IAC are rare lesions. Saleh *et al.*[1] found only 41 cases reported in the world literature, of which only 27 cases were confirmed by histopathology to be cavernous haemangiomas. Two further cases have since been reported.[3,4]

The pathological nature of these lesions is still debated, with the spectrum of suggestions regarding their origin ranging from vascular neoplasms[5] to hamartomatous malformations,[6] while others suggest that the lesions can change their appearance over time and exhibit both capillary and cavernous components.[7] They are thought to arise from the dense vascular networks around the geniculate ganglion and Scarpa's ganglion, and this may explain why the commonest sites for these lesions is the geniculate ganglion region and fundus of the IAC.[8,9] Although haemangiomas are extraneural in origin and benign in nature invasion of the nerve has also been reported.[10]

Intra-cranial haemangiomas may occur in isolation or in conjunction with cutaneous lesions as in the Sturge-Weber syndrome. The haemangioma may be hormone dependent and there is a tendency for enlargement during pregnancy.[11] The patient presented in this report developed symptoms during pregnancy with recovery of facial nerve function in the post partum period suggesting that her lesion was hormone dependent.

There are considerable similarities in the way all tumours of the IAC present with hearing loss being the most common presenting symptom. The presenting symptoms of haemangiomas, vestibular schwannomas and facial schwannomas are summarized in Table I. From this it can be seen that the majority of patients with haemangiomas of the IAC give a history of hearing loss and accompanying facial nerve weakness. These symptoms often occur when the lesion is relatively small.

The lesions can be difficult to detect radiologically due to their small size at presentation, with most tumours being smaller than 10 mm when excised,[12] although larger lesions have been reported.[13,14] Although these are vascular lesions, angiography is usually negative due to slow blood flow through the dilated channels.[15] Computed tomography (CT) may be normal or show a well-circumscribed mass of increased density with occasional calcification and minimal enhancement with contrast.[15] The presence of stippled calcification whilst suggestive of an haemangioma is not diagnostic and is also seen in schwannomas and meningiomas.[16] MRI is more specific and sensitive than CT for diagnosing haemangiomas of the IAC. On both T1 and T2 images there is a heterogenous appearance possibly due to intratumoral calcification, and on T2-weighted images, there is usually a surrounding rim of low signal due to haemosiderin deposition.[15] Signal intensity on T2-weighted images is higher than for vestibular schwannomas.[17] Enhancement is seen with gadolinium.[1] In this case, the MRI findings which may have suggested the diagnosis were not apparent, possibly because such findings are more often associated with larger tumours. Gradient echo MRI imaging would have demonstrated blood degradation products such as haemosiderin more accurately, but this imaging modality was not utilized in this case.

A difference of opinion exists as to the best intra-operative management of the facial nerve associated with haemangioma of the IAC. The extraneural origin makes it possible for the tumours to be removed with anatomical preservation of the facial nerve. Glasscock *et al.*,[18] however, report the presence of a dense perineural reaction which prevents the establishment of a clear cleavage plane and they recommend resection of the nerve with grafting to reduce the incidence of recurrence. As haemangiomas progressively enlarge and do not involute spontaneously, early surgical resection is recommended as this offers the best prospects for preservation of facial nerve function.[10]

Conclusion

Cavernous haemangiomas of the IAC are rare lesions that can mimic other lesions of the IAC clinically and radiologically. Progressive hearing loss with associated facial nerve abnormalities, particularly when the lesion is small, should raise the possibility of haemangioma. The symptoms may arise and progress during pregnancy only to regress in the postpartum period. MRI is the imaging modality of choice with haemangioma being represented by enhancing, calcified lesions in the perigeniculate region.

TABLE I

COMPARISON OF PRESENTING SYMPTOMS FOR VESTIBULAR SCHWANNOMAS, FACIAL SCHWANNOMAS AND CAVERNOUS HAEMANGIOMAS OF THE INTERNAL AUDITORY CANAL

	Haemangioma Saleh *et al.*[1]		Vestibular schwannoma Morrison[19]		Facial schwannoma Lipkin *et al.*[20]	
	n = 37	(%)	*n* = 238	(%)	*n* = 238	(%)
Hearing loss	36	97.3	162	68.1	116	48.7
Tinnitus	6	16.2	117	49.1	32	13.4
Vertigo/dysequilibrium	10	27	117	49.1	24	10.1
Facial weakness	24	64.8	11	4.6	173	72.7
Pain/headache	1	2.7	33	13.9	25	10.5
Aural fullness	6	16.2	5	2.1	–	
Eye problems	–		10	4.2	–	
Ear canal/parotid mass	–		–		40	16.8
Otorrhoea	–		–		15	6.3

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Maintaining a high index of suspicion of haemangioma as a diagnostic possibility for patients with intracanalicular tumours may improve the prognosis for preservation of facial nerve function.

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Address for correspondence:
Mr A. M. Shaida, M.A., F.R.C.S., ENT SpR,
Department of Otolaryngology,
Clinic 10, Box 48,
Addenbrooke's Hospital NHS Trust,
Hills Road,
Cambridge CB2 2QQ.

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