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Clinical Records

Cavernous angioma of the internal auditory canal

Kristian Aquilina, F.R.C.S., Jagdeep S. Nanra, F.R.C.S.I., Francesca Brett, M.D., F.R.C.Path.*, Rory McConn Walsh, M.D., F.R.C.S. (Orl)†, Daniel Rawluk, F.R.C.S. (Glas.)

Abstract

Cavernous angiomas of the internal auditory canal are rare lesions. The authors present a case of a 29-year-old lady with multiple infratentorial cavernous angiomas, whose sister had previously undergone surgery for a similar supratentorial lesion. She initially presented with an acute brainstem haematoma, secondary to a pontine cavernous angioma. Three years later she developed progressive right-sided sensorineural hearing loss and facial nerve paresis due to an internal auditory canal lesion. This was removed via the translabyrinthine approach and was found to be a cavernous angioma.

This report underlines the multiple and dynamic nature of familial cavernous angiomas, as well as the importance of follow up to determine whether new symptoms are due to the enlargement of known angiomas or the development of new ones. As far as the authors are aware, this is the first report describing a cavernous angioma of the internal auditory canal in the context of familial and multiple infratentorial angiomas.

Key words: Haemangioma, Cavernous; Hearing Loss, Sensorineural

Introduction

Cavernous angiomas of the internal auditory canal are relatively rare lesions and around 35 cases have been described in the English literature.[1–19] Cavernous angiomas occasionally exist as multiple supra- and infra-tentorial lesions, often in patients whose relatives are also known to have similar lesions. We report a case where a *de novo* cavernous angioma occurred in the right internal auditory canal during follow up for a pontine cavernous angioma. This pontine lesion had been the source of a brainstem haemorrhage three years prior to the development of right-sided progressive sensorineural hearing loss and facial nerve paresis caused by the internal auditory canal lesions. To the best of our knowledge, this is the first case of an internal auditory canal cavernous angioma associated with multiple infratentorial cavernous angiomas, one of which had bled previously. The presence of a cerebellar cavernous angioma on the right side implied that retraction of the cerebellum had to be avoided during the approach to the internal auditory canal lesion.

Case report

A 29-year-old lady, whose sister had previously undergone surgery for a supra-tentorial cavernous angioma, initially presented six years ago with an episode of acute headache, dysarthria, ataxia, bilateral tinnitus and bilateral hearing impairment. A magnetic resonance scan of the brain showed an acute haemorrhage within a cavernous angioma in the superior medulla and lower pons, at the level of the VIIth and VIIIth cranial nerve nuclei (Figure 1 (a) and (b)). Other cavernous angiomas were also identified in the cerebellar peduncle, the right cerebellar hemisphere and the parietal cortex. Her symptoms partially resolved over the subsequent year and serial magnetic resonance imaging (MRI) did not demonstrate any change in the size of the brainstem lesion.

Three years later she developed rapidly progressive right-sided deafness, associated with right facial paresis. These changes were not associated with recurrence of her previous ataxia and dysarthria. Her facial nerve function on the right was Grade III. Pure tone audiometry showed a mean threshold of 25 dB on the right; the hearing in the left ear was normal. MRI at this stage showed no change in her brainstem and cerebellar cavernous angiomas; there was, however, a new lesion 5 mm in diameter in the right internal auditory meatus. This was hypointense on T1- and isointense on T2-weighted images; enhancement was noted after gadolinium diethylenetriaminepenta-acetic acid (DTPA) administration. It was therefore felt to be a vestibular schwannoma, although it was clear that the new facial palsy was discordant with this. It was decided at this stage to delay surgery and to manage the lesion by regular evaluation.

The lesion was noted to enlarge on serial scans and measured 10 mm two years later (Figure 2). There was no change in the signal characteristics, but the lesion now extended into the cerebellopontine angle by 5 mm. There was no significant change in her facial nerve function. Pure tone audiometry showed a mean threshold of 70 dB in the right ear. It was felt that the increasing size of the lesion was an indication for operative management. In view of her right-sided hearing deficit, and considering that a retrosigmoid approach would involve retraction on the

From the Departments of Neurosurgery, Neuropathology* and Otorhinolaryngology†, Beaumont Hospital, Dublin, Ireland.
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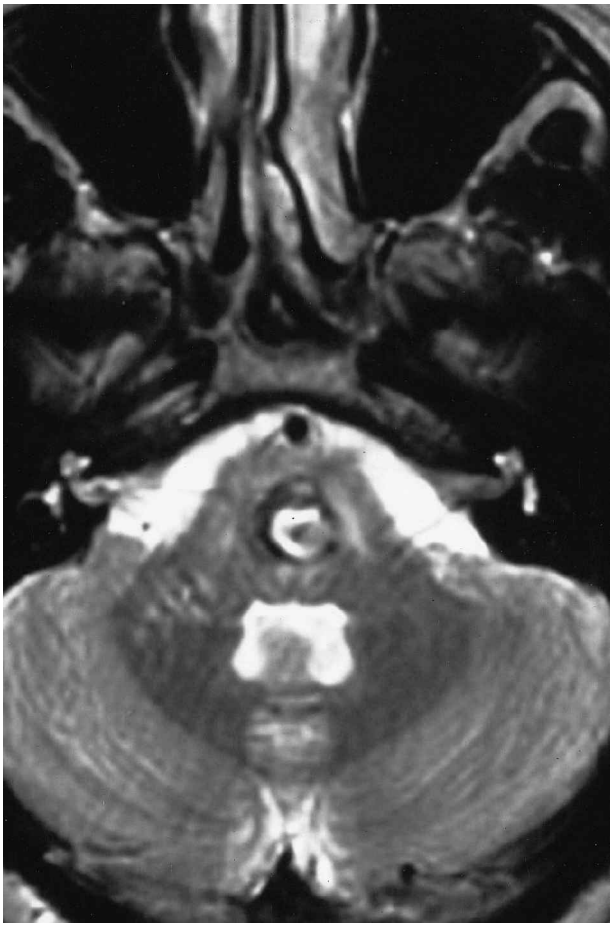
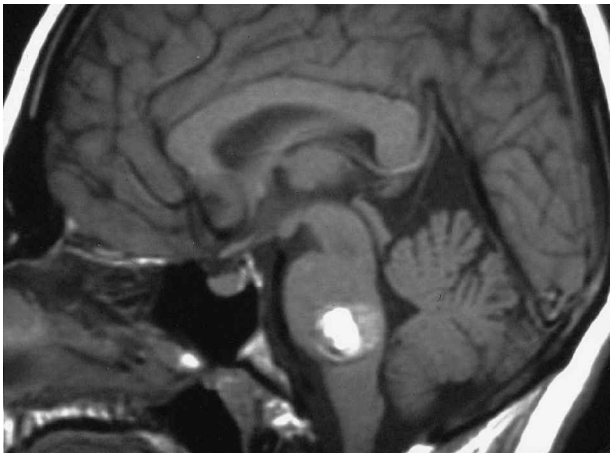


(a)



(b)

FIG. 1

T2-weighted axial (a) and T1-weighted sagittal (b) MRI scan at initial presentation showing acute haemorrhage within a pontine cavernous angioma.

right cerebellar hemisphere, which also contained a cavernous angioma, it was decided that the translabyrinthine approach would be the safest option. The lesion was reddish-brown in colour and was adherent to the VIIth and VIIIth cranial nerves. Complete resection was achieved (Figure 3). Intra-operative facial nerve

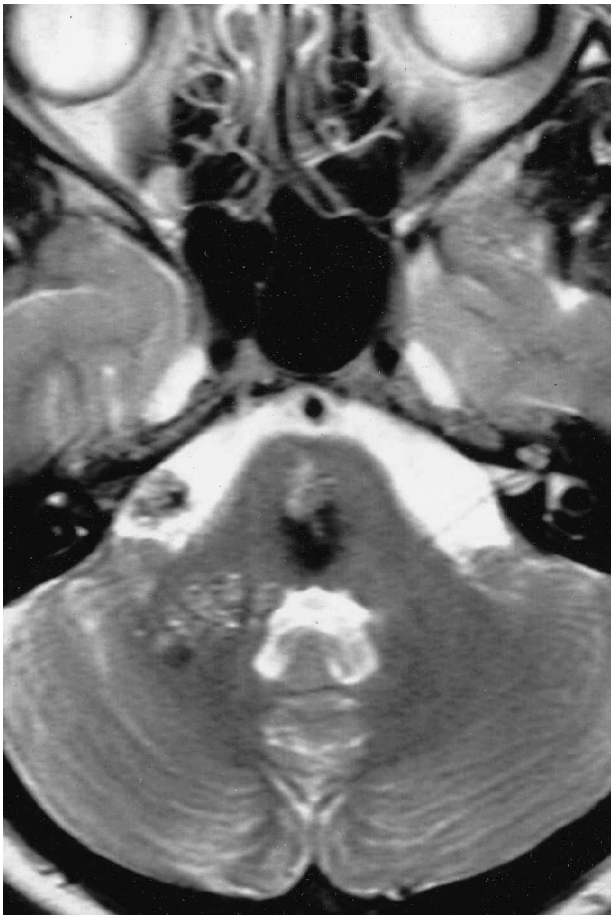


FIG. 2

T2-weighted axial MRI scan showing right internal auditory canal and CPA cavernous angioma. The other angiomas in the pons and the right cerebellum are also evident.

monitoring was used and structural and functional facial nerve integrity was maintained. There was no deterioration in her facial paresis post-operatively. Histologically, the lesion was seen to consist almost entirely of dilated thin-walled sinusoidal vascular channels typical of a cavernous angioma (Figure 4).

Discussion

Cavernous angiomas are malformations of the intracranial vasculature that affect 0.5 to 0.7 per cent of the population.[20] In post-mortem studies they constitute up to 15 per cent of intracranial vascular malformations, which also include arterio-venous malformations, venous malformations and capillary telangiectases. Cavernous angiomas may occur sporadically, when they usually exist as isolated lesions, or may be familial, when the incidence of multiple angiomas is higher.[20,21] About a quarter of cavernous angiomas are situated in the infratentorial compartment; the cerebellum and the brainstem are equally affected. The commonest location of brainstem cavernous angiomas is the pons.

Cavernous angiomas are discrete compact lesions consisting of sinusoidal back to back vascular spaces lined by an endothelial cell layer. The vascular spaces essentially contain thrombosed blood, suggesting that the blood flow through the lesions is slow leading to

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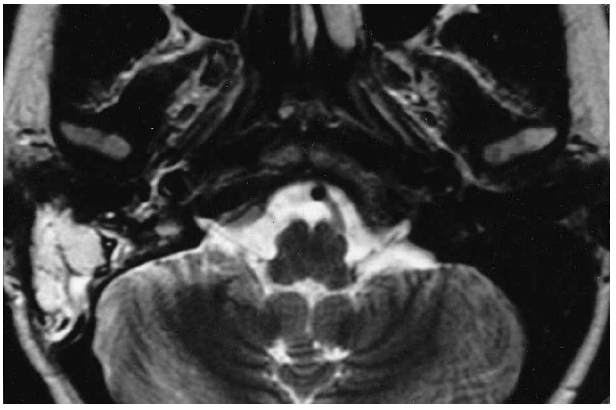

FIG. 3

T2-weighted axial MRI scan showing post-operative appearance, with complete excision of the right internal auditory meatus cavernous angioma.

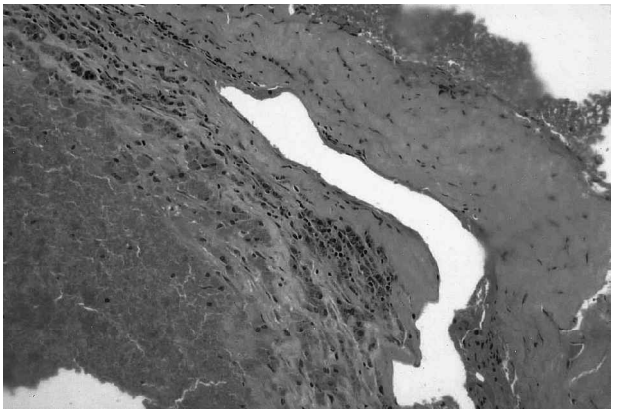

FIG. 4

Photomicrograph of the biopsy specimen. There are numerous large vascular channels arranged in back to back fashion with fibrillar connective tissue in between. This appearance is consistent with a cavernous angioma (H & E; ×160)

- **A patient with multiple familial haemangioma is presented**
- **This is the first reported case of a patient with a familial haemangioma in the internal auditory canal**
- **The patient presented with hearing loss and facial paresis**

stagnation. Unlike arteriovenous malformations, the walls of the vascular spaces do not contain an elastic or a muscular layer.

Cavernous angiomas are symptomatic in three ways. They often cause seizures secondary to the irritative effect of the haemosiderin, the surrounding gliosis, and by compressing the adjacent cortex. Haemorrhage within the lesions causes acute neurological deficits; the risk of haemorrhage has been suggested to lie between 0.1 and 1.1 per cent per lesion per year.[20,22,23,] Cavernous angiomas can also present with a progressive neurological deficit; this is more likely in infratentorial lesions and is due to progressive slow enlargement secondary to chronic or recurrent extravasation of blood or to thrombosis.

The familial occurrence of cavernous angiomas has been described in several studies.[20,21] This is known to be commoner in individuals of Mexican-American (Hispanic) descent. The malformations are often multiple in these patients and have been shown to be more dynamic. Zabramski *et al.*[20] prospectively followed six families for a mean duration of 2.2 years. Six of the 21 patients developed new lesions on MRI of the brain during this period. Our patient's intra-canalicular cavernous angioma had not been identified on the initial magnetic resonance images performed to evaluate and follow up the pontine lesion. It is therefore likely that this represents a new lesion.

Extra-axial cavernous angiomas are rare lesions; they have been reported to occur most commonly in the middle fossa and the cavernous sinus.[24,25] Around 35 cases of internal auditory canal cavernous angiomas have been reported.[1–19] Dufour *et al.*[6] reviewed the 25 cases reported from 1949 to 1994 and added a further five cases to the series. Although the average tumour size of these lesions was less than 10 mm, 88 per cent of patients sustained rapidly progressive hearing loss and up to 72 per cent presented with facial nerve dysfunction. By contrast, in a similar population of vestibular schwannoma patients, there were no cases of facial nerve dysfunction with tumours up to 10 mm in diameter. In the series described by Pappas[17] only two out of five patients presented with facial nerve dysfunction.

Sundaresan *et al.*[19] suggested that sensorineural hearing loss and pre-operative facial nerve palsy in the presence of a small intracanalicular tumour is the typical clinical picture of a vascular space-occupying lesion in the internal auditory canal, with or without extension into the cerebellopontine angle. Our patient would support this hypothesis, however, the early symptoms of the internal auditory canal cavernous angioma could have been easily mistaken to arise from slow growth of the pontine angioma; it was the unilateral hearing loss, confirmed on pure tone audiometry, as well as the absence of ataxia and dysarthria, that pointed to right-sided internal auditory meatus and cerebellopontine angle pathology. The facial nerve weakness could have been caused by involvement of the facial nerve nucleus by the pontine cavernous angioma.

None of the reported cases have been described to occur within the context of familial and multiple cavernous angiomas. Moreover, our patient is unusual in that a magnetic resonance scan three years prior to presentation did not show any evidence of a lesion in the internal auditory canal. This suggests that the rapid growth of new lesions known to occur in patients with the familial form of the disease[20] occurs not only in the intra-axial lesions but also in the rare extra-axial ones.

Conclusion

This report underlines the importance of including cavernous angiomas in the differential diagnosis of internal auditory canal lesions, particularly in patients with multiple and familial cavernous angiomas. It is often difficult to distinguish these lesions from vestibular schwannomas on the basis of imaging alone, and early surgery is therefore indicated to define histology. Patients with multiple cavernous angiomas need to be followed up regularly to identify whether progressive neurological symptoms are due to growing lesions or to the development of new ones.

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Address for correspondence:
Mr Kristian Aquilina, F.R.C.S.,
Specialist Registrar in Neurosurgery,
Department of Neurosurgery,
Beaumont Hospital,
Beaumont Road,
Dublin 9,
Ireland.

Fax: 00353 1 8092309
E-mail address: kristianquilina@hotmail.com

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