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Cavernous haemangioma of the facial nerve

PEDRO ESCADA, M.D.*, CLARA CAPUCHO, M.D.*, JOSÉ MADEIRA SILVA, M.D., PH.D.*,
CARLOS BENTES RUAH, M.D., PH.D.*, JOSÉ PRATAS VITAL, M.D., PH.D.†, RUI SILVA PENHA, M.D., PH.D.*

Abstract
Facial nerve haemangiomas are probably the most frequent benign tumours involving the facial nerve in its intratemporal portion. Usually facial nerve dysfunction is present when these tumours are of extremely small size, the average tumour being less than 10 mm. We present a case of a 15 mm diameter cavernous haemangioma of the geniculate region, with histological findings of nerve infiltration, without facial nerve symptoms. The atypical clinical presentation justifies the report and subsequent literature review.

Key words: Haemangioma; Facial nerve; Temporal bone

Introduction

There are two main groups of intratemporal vascular tumours: glomus tumours and haemangiomas. Temporal bone haemangiomas are more often called facial nerve haemangiomas because of their predilection to involve the facial nerve. They were once thought to be rare, but they have been recently seen in some series with a incidence close to better-known facial nerve neuromas. It is unclear whether they are true tumours or vascular malformations. They develop extraneurally and originate in the vascular plexuses distributed along the facial nerve.

Haemangiomas of the facial nerve present with facial nerve paralysis and/or hearing loss, of different severities. The most important diagnostic investigations consist of appropriate imaging of the temporal bone. Magnetic resonance imaging (MRI) is effective in detecting tumours in the internal auditory canal and computed tomography (CT) is effective to rule out a possible geniculate ganglion lesion. Differential diagnosis from other intratemporal tumours, particularly facial nerve neuromas, is mandatory. A more frequent and severe facial nerve dysfunction points to the diagnosis of facial nerve haemangioma. CT can display typical findings in 50 per cent of cases. In the remainder, definite diagnosis is obtained at surgery or after histological examination.

Surgical removal is the definitive form of treatment, and early resection offers the best chance for good facial recovery. Surgical approach is chosen according to the location and size of the tumour and pre-operative hearing level. Facial nerve repair, by primary anastomosis or grafting, is necessary in some instances with variable post-operative facial nerve function.

Case report

A 32-year-old healthy Caucasian male presented with a six-month history of a progressive hearing loss on the left ear. Otolaryngological examination and audiometric evaluation revealed a middle ear mass and a conductive hearing loss in the left ear, and the patient was referred to our hospital for further care.

At admission, he presented normal facial function. Otoscopy showed a middle ear red bluish mass bulging the intact superior pars tensa and pars flaccida. The lesion was not pulsatile at microscopic examination but became slightly pale with positive pressure on pneumatic otoscopy. Mastoid ausculation was negative. Pure-tone audiometry disclosed a 30 dB conductive hearing loss on the left. Tympanometry, performed using a 226-Hz probe tone, showed a Lidén and Jerger type A_d admittance tympanogram, with an abnormal notch when using a 678-Hz probe tone. A CT scan of left temporal bone revealed a well-circumscribed mass adjacent to the medial wall of the middle ear cavity. The mass appeared confluent with the geniculate ganglion and tympanic segment of the facial nerve, and had eroded the geniculate area and neighbouring ossicular chain (Figure 1a and 1b). The lesion was consistent with a haemangioma or neuroma of the facial nerve.

A surgical procedure was then scheduled to remove the tumour. A combined transtemporal supralabyrinthine and transmastoid approach exposed a 15 mm long vascular lesion which had developed from the distal labyrinthine segment to the second genu. The stapes superstructure and the long process of the incus were absent. The remaining ossicular chain was displaced. In the medial tympanic cavity wall, both the processus cochleariformis and adjacent portion of tensor tympani canal were eroded. The tumour invaded and adhered firmly to the posterior tympanic cavity wall. To expose appropriately both the tumour with the adjacent second genu and mastoid segment of the facial nerve, we chose to perform a canal wall down technique (Figure 2).

The facial nerve adjacent to the tumour had a normal appearance and merged in the tumour mass without an identifiable dissection plane. The tumour was removed with the proximal part of the greater superficial petrosal nerve, the geniculate ganglion, the distal part of the labyrinthine portion and all the length of the tympanic portion of the facial nerve.

From the Departments of Otolaryngology* and Neurosurgery†, Egas Moniz Hospital, Lisbon, Portugal.
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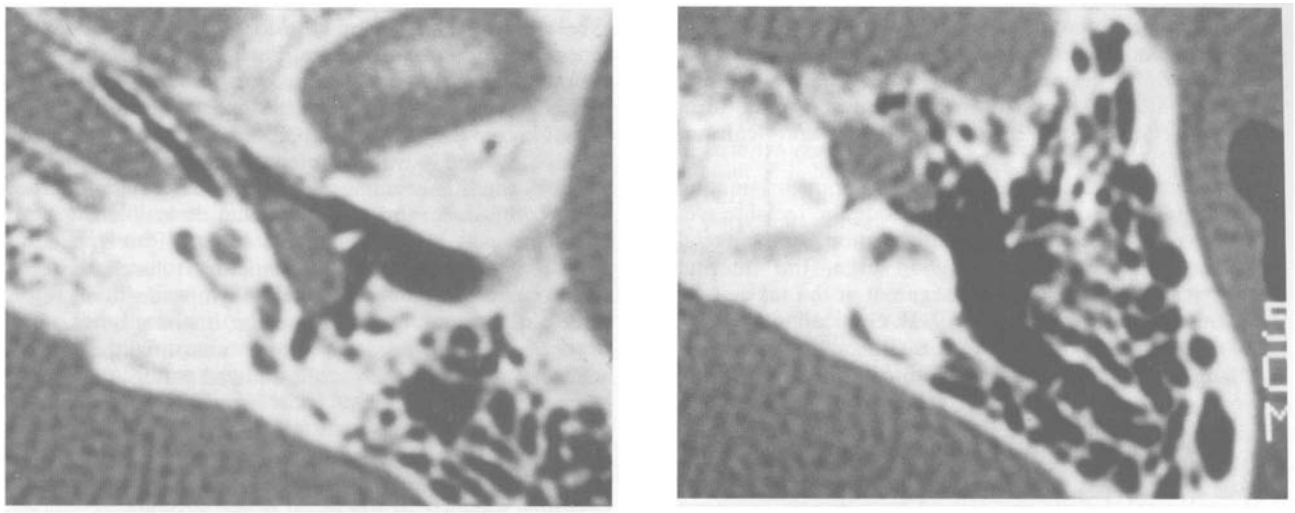

(a) (b)

FIG. 1a and 1b

CT scan of the left temporal bone showing a soft tissue tumour (haemangioma arrowed) in the geniculate ganglion and tympanic portion of the facial nerve, with associated erosion of the ossicular chain (a) and erosion of the bone around the geniculate fossa (arrow) (b).

The facial nerve was reconstructed with a 3 cm sural nerve graft. Both proximal and distal anastomosis were fixed with one 10/0 nylon suture, and the reconstruction was covered with fibrin glue and an autogenous temporalis fascia graft.

Ossicular reconstruction was not performed. The bone defect in the floor of the middle cranial fossa was reconstructed with a piece of craniotomy flap. The posterior two-thirds of the temporalis muscle was rotated to cover the operative cavity, the Eustachian tube was obliterated and finally a meatoplasty is performed.

Histological examination (Figure 3) showed features compatible with the diagnosis of cavernous haemangioma. The tumour showed loss of pseudocapsule with infiltration of the nerve and some spicules of bone are seen at the periphery.

Four years after surgery the patient has no clinical or radiological evidence of recurrence. The facial function is House-Brackmann grade III.

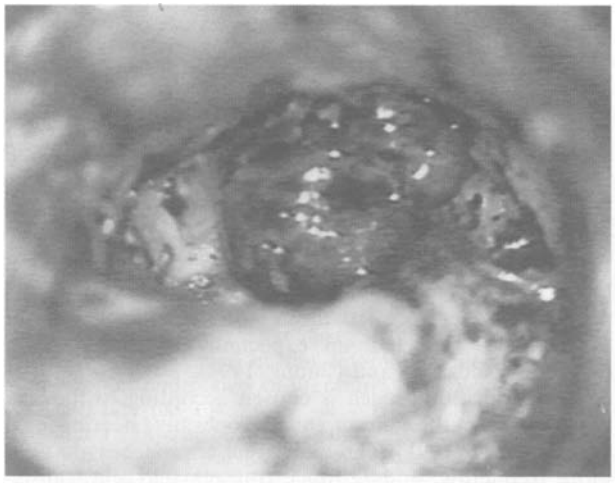

FIG. 2

Surgical view: a highly vascular tumour was found filling the middle-ear cleft and epytimpanum. It originates in the second genu and filled all the geniculate region.

Discussion

Haemangiomas of the facial nerve are thought to be rare. Glasscock *et al.* (1984) found 42 cases of skull base haemangiomas in the literature including those of the temporal bone. Shelton *et al.* (1991) reported 34 cases from the House Ear Institute and Eby *et al.* (1992) reported eight cases from the ENT department of the University Hospital of Zurich. Dufour *et al.* (1994) found 48 cases of facial nerve haemangiomas reported in the literature since 1949 and added six new cases. According to the literature, these tumours have an incidence comparable to the facial nerve neuromas, and some authors recognize temporal bone haemangiomas as the benign tumour most commonly involving the facial nerve in its intratemporal portion (de Amesti *et al.*, 1995).

Most of these tumours have a unique histological appearance, peculiar to the temporal bone, characterized by large vascular spaces lined with thin endothelium, surrounded by thick walls filled with uniform fibrous tissue. It is unclear whether they are true haemangiomas, vascular malformations or a different benign vascular tumour (Shelton *et al.*, 1991). They are usually classified as

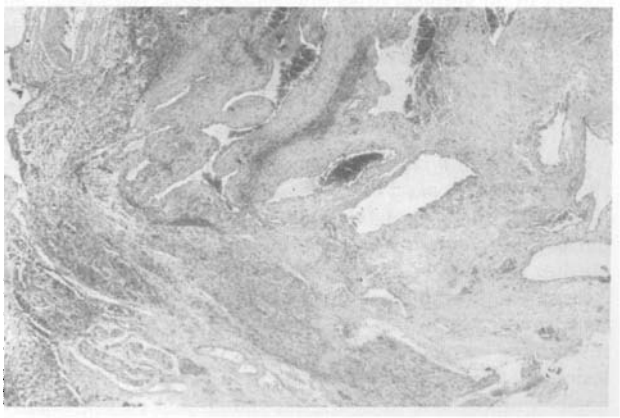

FIG. 3

Photomicrograph of the tumour: thick-walled large vascular channels are lined with flat endothelial cells. There are some areas with loss of pseudocapsule and infiltration of the nerve (H & E; × 60).

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cavernous, capillary or mixed type by the predominant size of the vascular spaces. Some haemangiomas may grow as expansile bony lesions with a honeycomb of bony trabeculae filled by vascular channels, justifying the term ossifying or osseous haemangioma (Fisch and Rüttner, 1977).

The haemangiomas are extraneural tumours originating in the vascular plexuses distributed along the facial nerve. The areas of most intense vascularization of the facial nerve are the geniculate ganglion area, the internal auditory canal and the mastoid segment at the take-off of the chorda tympani (Eby *et al.*, 1992). Geniculate capillary plexus is the most exuberant plexus (Balkany *et al.*, 1991). Haemangioma of the internal auditory canal originates more likely in the vascular plexus surrounding Scarpa's ganglion, and therefore should be named vestibular, rather than facial nerve haemangiomas (Fisch, 1968; Eby *et al.*, 1992).

Neurological deficits are caused by extrinsic compression of the nerve (Martin *et al.*, 1992) consequently the tumour potentially can be removed with preservation of neural continuity. Preservation of the intact facial nerve is less frequent with geniculate ganglion tumours than with internal auditory canal ones because geniculate ganglion haemangiomas are often associated with an intense perineural reaction (Shelton and Brackmann, 1989) or direct facial nerve infiltration (Eby *et al.*, 1992).

The most common clinical features of temporal bone haemangiomas are facial nerve dysfunction and hearing loss. Other symptoms, like tinnitus and dizziness are rarely present.

Characteristically, severe facial nerve dysfunction is present when these tumours are of extremely small size, contrasting to facial nerve neuromas, which are usually relatively large when they produce symptoms. Symptomatic facial nerve haemangiomas are usually less than 10 mm in size, but only 10 per cent of symptomatic acoustic neuromas measure less than 10 mm (Shelton *et al.*, 1991; Dufour *et al.*, 1994). Clinical presentation of facial nerve dysfunction may follow several different patterns (Eby *et al.*, 1992); some patients manifest a progressive facial paralysis, suggesting a tumoral process, while others present with sudden facial nerve paralysis. Other possible patterns are hemifacial spasm, synkinesis and facial weakness. Recurrent facial paralysis has been reported in patients with these tumours. Often each episode recovers completely after treatment with steroids, and incomplete recovery is seen usually after several episodes over several years. This pattern points out the need to consider tumoral lesions in all atypical presentations of facial paralysis. The admonition of Sir Terence Cawthorne 'all that palsies is not Bell's' must be remembered (Cawthorne, 1969). Electromyography may be helpful in establishing the difference, showing findings of simultaneous denervation and reinnervation, not expected in Bell's palsy, and consistent with a compression neuropathy (Ylikoski *et al.*, 1984; Shelton *et al.*, 1991).

Clinical features of facial nerve haemangiomas vary in those tumours arising at the geniculate ganglion and those in the internal auditory canal. Haemangiomas of the internal auditory canal present with slowly progressive sensorineural hearing loss with retrocochlear findings, resembling acoustic neuroma or any other expansive lesion of the internal auditory canal or cerebellopontine angle. Unlike those tumours, haemangiomas do not usually present with tinnitus or dizziness (in spite of the abnormal response to caloric stimulation in the electronystagmogram), and exhibit facial nerve dysfunction more often, starting with a discrete hemifacial spasm, evolving to a progressive and frequently complete facial paralysis (Shelton *et al.*, 1991; Eby *et al.*, 1992). Literature review reports a high but variable rate of facial nerve abnormalities in different internal auditory canal series: Eby found facial nerve symptoms in all of his three patients, Pappas in two of his seven patients (Pappas *et al.*, 1989) and Shelton in three of the House Ear Institute series with 15 internal auditory canal haemangiomas. Facial nerve symptoms are usually the first and predominant clinical features of haemangiomas of the geniculate region. Rarely patients with geniculate ganglion haemangiomas have no facial symptoms. Eby found facial nerve symptoms in all of his three patients and, in the House Ear Institute series, all of the patients showed facial nerve abnormalities. Facial nerve dysfunction may follow two patterns: a rapid progressive and complete facial paralysis (less than one year) and recurrent sudden facial paralysis. Hearing may remain unaffected but conductive hearing loss will occur if the tumour extends to the middle-ear cleft or involves the tympanic segment of the facial nerve, eroding or contacting with the ossicular chain.

Appropriate imaging of the temporal bone is extremely helpful in establishing the diagnosis. High-resolution CT and MRI are the two most useful diagnostic modalities available. Angiography is not helpful because these tumours are angiographically occult (Babu *et al.*, 1994). MRI probably detects all tumours of the internal auditory canal and 40 per cent of tumours in the geniculate ganglion region; CT rarely detects haemangiomas of the internal auditory canal but probably shows most of the geniculate ganglion haemangiomas (Lo *et al.*, 1989). Consequently, an ideal diagnostic strategy consists of MRI of the temporal bone in the first instance. If MRI findings are negative, CT should then be performed to rule out a possible geniculate ganglion lesion. The diagnostic contributions of MRI and CT in the evaluation of these tumours differ in both the topographical diagnosis, and the diagnosis of the nature of the lesion (Quevedo *et al.*, 1996).

MRI reveals a high signal intensity of both T1- and T2-weighted images, resembling other neoplastic lesions in the same location (Lo *et al.*, 1989; Pappas *et al.*, 1989). In some cases gadolinium enhanced MRI can be useful in diagnosis, displaying a strong enhancement of the lesion and the adjacent portions of the facial nerve, with poorly defined lesion margins. Cholesteatoma are non-enhancing lesions and schwannomas are more sharply demarcated on the MRI; in acute Bell's palsy, enhancement is limited to the course of the facial nerve (Tien *et al.*, 1990; Martin *et al.*, 1992).

CT can provide a diagnosis of the nature of the mass in about 56 per cent of cases (Quevedo *et al.*, 1996), showing characteristic intratumoral calcifications. CT findings may be spicules or stippling of bone within the tumour or a border of thin new bone seen around the tumour. Both aspects are related to extensive remodelling of bone (Curtin *et al.*, 1987; Lo *et al.*, 1989; Eby *et al.*, 1992).

Surgical removal is the definitive form of treatment in intratemporal facial nerve haemangiomas. Because these tumours are extremely aggressive locally, early resection offers the best chance for good facial recovery (Gavilán *et al.*, 1990). Therefore, in those instances where a non-neuroma is suspected, an expectant attitude is to be deplored (Nedzelski and Chiong, 1994).

The surgical approach is chosen according to the size and location of the tumour and pre-operative hearing level (Shelton *et al.*, 1991). Three different approaches are used more frequently: middle fossa, transmastoid and translabyrinthine approaches (Nedzelski and Chiong, 1994). The middle fossa approach is useful in haemangiomas of the internal auditory canal when hearing is good and translabyrinthine in haemangiomas of the internal auditory

canal when hearing is poor. The transmastoid approach which combines canal wall up mastoidectomy with posterior tympanotomy is proposed in tumours confined to the tympanic and mastoid segments of the facial nerve. Geniculate ganglion haemangiomas usually extend along the labyrinthine segment, and, to a varying degree, along the tympanic segment, and are exposed with a combined transmastoid and middle fossa approach. Combined transtemporal supralabyrinthine and transmastoid approach has been an alternative to this last situation, proposed by Fisch (Fisch, 1982; Fisch, 1988), and differs from the transmastoid-middle fossa approach because most of the exposure is obtained by removing bone between the dura mater and the otic capsule, minimizing temporal lobe compression and bleeding. Another advantage is that it allows a continuous and complete exposure of all the length of the facial nerve, from the internal auditory meatus to the stylomastoid foramen. In all instances tumour removal is initiated only after complete local exposure.

In spite of the extraneural origin of haemangiomas, often surgical removal of the tumour entails facial nerve resection, particularly in the geniculate ganglion lesions, requiring nerve repair. The House Ear Institute series reports nerve repair for 11 of 15 geniculate ganglion haemangiomas (73 per cent) and for four of eight internal auditory canal lesions (22 per cent). Reconstruction options include (Nedzelski and Chiong, 1994): (1) primary anastomosis if sufficient redundancy of the nerve is present; (2) rerouting of the nerve; and (3) grafting with the use of either the greater auricular nerve or the sural nerve. Functional nerve results are variable. It is advisable to adopt a universal standard for grading facial nerve recovery, such as the House-Brackmann facial nerve grading system (House and Brackmann, 1985) to support a better understanding of the meaning of the results and a objective comparison of various forms of treatment. Better overall facial nerve function recovery is achieved when facial nerve continuity is preserved. When facial nerve repair is necessary the reparative technique seems to be less important than the duration of the palsy (Eby *et al.*, 1992). When facial nerve continuity cannot be achieved the use of facial-hypoglossal anastomosis is a reasonable option (Nedzelski and Chiong, 1994).

Conclusion

Facial nerve haemangiomas are rare. Their predilection to involve the facial nerve makes facial nerve symptoms the predominant clinical feature of haemangiomas of temporal bone, particularly at the geniculate region. Severe neurological deficits characteristically occur when these tumours are of extremely small size. We report a case of facial nerve haemangioma with unexpected normal facial function, considering the large tumour size, geniculate ganglion location and infiltration of the nerve by the tumour, documented in histological examination.

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Address for correspondence:
Dr. Pedro Alberto Escada,
Departamento de Otorrinolaryngologia – Hospital de Egas Moniz,
Rua da Junqueira, 126,
1300 Lisboa – Portugal