

Haemangiomas of the Geniculate Ganglion

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About 60 cases have been published since Pulec first described haemangiomas of the geniculate ganglion. They usually cause facial weakness even when they are very small. In cases of insidious evolution of facial paralysis, MRI, and CT are very helpful to rule out these tumors. The treatment is based on complete surgical removal, although it has to be individualized, depending on pre-operative facial function and the possibility of complete surgical removal with preservation of the facial nerve.

Key words: Facial paralysis. Haemangiomas. Geniculate ganglion.

Hemangiomas del ganglio geniculado

Desde su descripción por Pulec en el año 1969, se han publicado unos 60 casos de hemangiomas del ganglio geniculado. Suelen producir debilidad de la función facial, incluso cuando son de muy pequeño tamaño. La resonancia magnética y la tomografía computarizada ayudan en su diagnóstico en los casos de parálisis facial de evolución tórpida. El tratamiento está basado en la resección quirúrgica completa, aunque debe ser individualizado, y las decisiones dependen de la función facial preoperatoria y la posibilidad de resecar totalmente el tumor con preservación del nervio facial.

Palabras clave: Parálisis facial. Hemangiomas. Ganglio geniculado.

INTRODUCTION

Pulec first described a haemangioma of the geniculate ganglion (GG) in 1969.¹ So far, around 60 more cases have been published (Table). These tumours are believed to have originated in the vascular plexi around the GG and they may lead to a deficit in the facial function even when very small in size.

The physiopathology of this phenomenon seems to be a sequestration mechanism whereby the blood is directed towards the tumour instead of to the nerve, which becomes damaged.² These tumours are extraneural in origin and cause symptoms due to progressive compression of the facial nerve. Since the first description of this kind of lesion, a few cases have been reported of direct infiltration of the facial nerve by the tumour, as well as erosion of the otic capsule.³ Since it is not possible to achieve the complete excision of an infiltrating lesion without sacrificing the nerve, the presence or otherwise of histological infiltration is a critical element affecting the choice of the most appropriate

therapeutic strategy. There are somewhat contradictory papers on the handling of this kind of lesion, and early excision of the tumour while sparing the nerve is the most recommended.⁴

Previously Published Cases of Haemangiomas in the Geniculate Ganglion

Authors	Year	Cases
Pulec ¹	1969	1
Mangham et al ⁵	1981	3
Glasscock et al ¹⁶	1984	2
Mazzoni et al ³	1988	1
Lo et al ¹⁷	1989	1
Gavilan et al ¹³	1990	1
Shelton et al ⁴	1991	15
Eby et al ¹⁸	1992	3
Bhatia et al ¹⁹	1995	9
Pulec ¹¹	1996	4
Asaoka et al ²⁰	1997	1
Escada et al ⁸	1997	1
Friedman et al ⁷	2002	1
Piccirillo et al ¹⁰	2004	3
Isaacson et al ¹⁵	2005	6
This paper	2007	2

The authors have not indicated any conflict of interest.

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Received March 14, 2006.

Accepted for publication February 1, 2007.

We present here 2 cases of patients with haemangiomas of the GG and we discuss the most important aspects in the handling of this pathology.

CLINICAL REPORTS

Case 1

Female, 31 years old, without any personal history of interest, attended our clinic with complete left facial paralysis

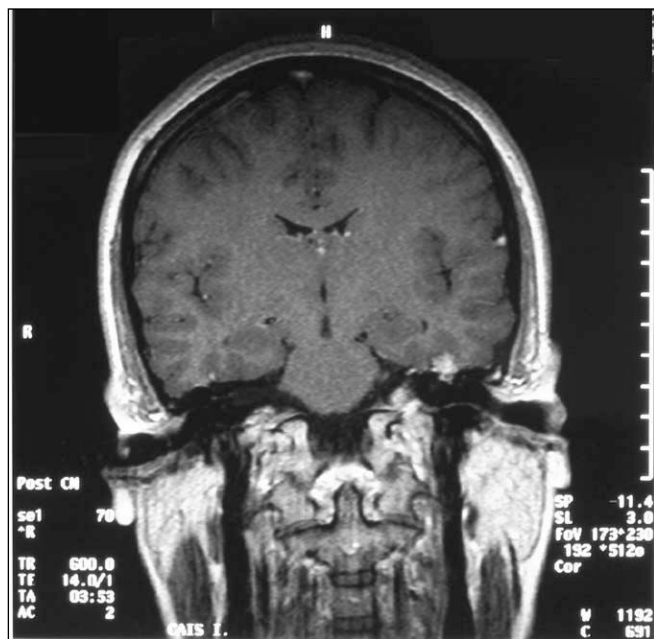


Figure 1. Lesion at the level of the geniculate ganglion region with uptake of gadolinium producing a slight alteration in the signal from the adjacent parenchyma.



Figure 2. Patient with complete eye closure and good tone at rest.

lasting for 8 months (grade VI on the House-Brackmann scale). She had been treated as having Bell's palsy, without any clinical improvement despite the medical treatment. The otoscopy was normal, as was the audiometry test. The other otoneurological examinations gave results within normal ranges. The computerized tomography (CT) of the petrous bone showed signs of bone erosion around the GG. The magnetic resonance (MRI) scan revealed a lesion at the level of the GG region with uptake of gadolinium and producing a slight alteration of the signal in the adjacent parenchyma (Figure 1). The electromyogram (EMG) showed a severe involvement (axonotmesis) of the facial nerve. In view of the suspicion of a tumour in the facial nerve and given the degree of its functional involvement, a transtemporal approach was employed on the left ear. During surgery, a bony housing was observed on the GG with an osteitic appearance. No macroscopic alterations were seen on the facial nerve in the first portion, GG nor on the second portion. It was possible to separate the tumour from the nerve and decompression was effected on the first portion, the GG and the start of the second portion. The pathology report on the tumour indicated haemangioma of the GG. No changes occurred in the hearing level after the procedure. The patient began to recover functionality after the sixth month, and the facial function 1 year after surgery was grade IV on the House-Brackmann scale. A gold weight has now been placed on her upper eyelid to help achieve complete eye closure (Figure 2). No relapse has been observed in the tumour 2 years after surgery.

Case 2

Female, 29 years of age without any personal history of interest, attended our clinic complaining of recurrent right facial paralysis lasting for 1 year, with grade V on the House-Brackmann scale. The otoscopy was normal, as was the



Figure 3. Lesion around the geniculate ganglion surrounding the cochlea.

audiometry. The EMG revealed signs of severe axonal and demyelinating lesion to the right facial nerve. The CT showed a lesion around the GG surrounding the cochlea (Figure 3). The EMG findings, together with the clinical history, indicated a compressive lesion. In view of the suspicion of a tumour on the facial nerve, exeresis of the lesion was performed through a transtemporal approach on the right ear. During surgery, the anterosuperior face of the petrous bone was seen to be deformed by a structure of vascular appearance that was resected. The pathology result was haemangioma of the GG. No changes were observed in the post-operative audiometry test. The EMG 1 year and a half after the procedure showed an improvement in the nerve conduction. An increase in the voluntary pattern was seen together with the evoked motor potential obtained. Facial function 4 years after the procedure was grade II. No relapse has been observed so far

DISCUSSION

Haemangiomas of the facial nerve represent 0.7% of intratemporal tumours.⁵ They may present as a recurrent facial paralysis that worsens with each bout, like a gradual deterioration of the facial function or a hemifacial spasm. Other symptoms include earache; pulsing tinnitus, similar to that of a vestibular schwannoma; vertigo, and conductive hearing loss. The dizziness and pulsing tinnitus are rare and may stem from the angiomatous invasion of the cochlea or vestibule, or compression of the vestibular nerve. Conductive hearing loss occurs due to the spread of the tumour into the middle ear, although this happens only very rarely. In an extensive review of the literature, only 6% of haemangiomas in the GG were seen to produce hearing loss.⁶

MRI with gadolinium and a CT scan are the imaging tests necessary for early diagnosis of this kind of lesion. Apart from its ability to show pathological changes in the nerve, MRI is useful to discard other causes of paralysis of the facial nerve, such as parotid tumours or other kinds of intracranial lesions. High-resolution CT of the temporal bone may be very useful when it comes to differentiating haemangiomas from other pathological entities and it can also provide important anatomical information for surgery.⁷

The differential diagnosis for haemangiomas of the GG includes lesions such as schwannomas of the facial nerve, meningiomas, cholesteatomas, and metastatic tumours. Pre-operative differentiation between schwannomas and haemangiomas of the facial nerve may be extremely difficult, unless the image shows "ossification" with irregular bony margins and bony spiculae within the tumour or the typical "salt and pepper" image of haemangiomas on CT. It has been calculated that this happens in 50% of intratemporal haemangiomas.⁸ This typical image is the origin of the name of ossifying haemangioma with which this kind of tumour is also known. On the other hand, facial schwannomas appear in the CT scan as masses with well-defined margins. There is also no specific symptom that helps us to differentiate these 2 conditions and the only noteworthy clue is that

haemangiomas tend to produce symptoms at a smaller size than schwannomas.⁹

Meningiomas of the GG are very rare tumours in this location and they have no specific appearance with CT or MR imaging. Finally, congenital cholesteatomas present a more defined and smoother erosion of the bone on the CT scan and do not show up as enhanced with the administration of contrast medium.¹⁰

An important characteristic of this kind of tumour is that it tends to lead to weakness in the facial function, even when very small, due to the phenomenon of vascular sequestration.² For this reason, in cases of facial paralysis without the typical characteristics of Bell's palsy, a CT scan of the temporal bone and a MRI with gadolinium of the fossa posterior, ear and parotid region must always be ordered to exclude the possibility of a tumour of the facial nerve.¹⁰

The most accepted treatment for haemangiomas of the facial nerve is surgical exeresis.¹¹ Complete surgical exeresis and primary reconstruction minimize the opportunities for relapse. Nonetheless, the therapeutic attitude must be individualized, depending on the pre-operative facial function and the possibility of totally resecting the tumour with conservation of the facial nerve. In our cases, it was possible to dissect the tumour from the facial nerve and conserve the anatomy, and function of the nerve.

The timing of the surgery is 1 of the most controversial points. Most surgeons believe that an early diagnosis and prompt surgical resection entail a better post-operative facial function. This is due to the fact that small haemangiomas produce less compression of nerve, less inflammatory reaction, and less fibrosis.^{12,13} In this way, it is possible to achieve surgical separation of the tumour from the nerve and conserve its function, as we did in our 2 patients. Large-sized haemangiomas are intimately attached to the facial nerve, enormously hindering the separation of tumour from the nerve. As a result, it is often necessary to resect the nerve and repair it by means of a primary termino-terminal anastomosis or the placement of a graft. Patients and surgeons have to understand that the optimal result of reconstruction by means of primary termino-terminal anastomosis or the placement of a graft does not allow the achievement of a better than grade III result on the House-Brackmann scale.¹⁴ Not all patients have such good outcomes in terms of their facial function after the repair. Shelton et al⁴ reported that 85% of patients requiring nerve repair obtained a grade IV or worse result on the House-Brackmann scale. These facts have led those authors to propose the delaying of haemangioma resection until the patient loses the ability to close the eye (grade IV on the House-Brackmann scale).

The type of approach depends on the location of the tumour, pre-operative hearing levels, and the size of the tumour. In general, the conservation of hearing is a realistic goal in patients with haemangiomas of the GG. For this reason, the transtemporal or fossa media approach is suitable for exeresis.¹⁵

In conclusion, haemangiomas of the GG are slow-growing benign tumours and for this reason decisions about its

handling must be based on the long-term maintenance of the facial nerve's function and hearing. Surgery is the treatment of choice for these tumours and the final outcome is satisfactory in terms of the facial function, depending to a large extent on their prompt resection. Otorhinolaryngologists must make a critical assessment of patients presenting with persistent and progressive facial paralysis, with or without hearing alterations, as this is of vital importance for discarding facial nerve tumours.

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