CASE STUDY

A New Entity in the Differential Diagnosis of Geniculate Ganglion Tumours: Fibrous Connective Tissue Lesion of the Facial Nerve

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Received 18 September 2011; accepted 16 January 2012

Abstract Differential diagnosis of geniculate ganglion tumours includes chiefly schwannomas, haemangiomas and meningiomas. We report the case of a patient whose clinical and imaging findings mimicked the presentation of a facial nerve schwannoma. Pathological studies revealed a lesion with nerve bundles unstructured by intense collagenisation. Consequently, it was called fibrous connective tissue lesion of the facial nerve.

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Introduction

The differential diagnosis of lesions in the geniculate ganglion mainly includes schwannomas, hemangiomas and meningiomas.

We report the case of a 5-year-old boy who presented unilateral facial paralysis of sudden onset without hearing loss. Imaging studies showed a lesion in the geniculate ganglion suggestive of facial nerve schwannoma. A comprehensive review of the anatomopathological findings did not enable...
them to be matched with any of the usual entities in the differential diagnosis of geniculate ganglion tumours.

**Case Report**

We report the case of a 5-year-old male with no history of interest who suffered complete facial paralysis with a sudden onset and 1 year of evolution. He reported no hearing loss, vertigo symptoms or impaired balance. He presented no significant findings on otoscopy. The audiometric studies determined normoacusis.

We conducted a petrosal computed tomography (CT) scan with multiplanar reconstructions, which revealed an osteolytic lesion, with sharp contours and multiple lobes, without sclerosis or permeation of the surrounding bone. This lesion affected the area of the right geniculate ganglion, in communication with the tympanic cavity, and showed signs consistent with an origin in the facial nerve (Fig. 1A). The T1 sequence of the cerebral magnetic resonance imaging (MRI) scan showed a limited hyperintense lesion in the geniculate ganglion, of approximately 8 mm in diameter (Fig. 1B).

Given the complete facial paralysis and the evolution of over 1 year, we decided to excise the tumour through a transtemporal approach. The lesion was identified in the geniculate ganglion and virtually no extension to the first and second portions of the facial nerve was observed. The margins of these portions were resected and the greater superficial petrosal nerve was sectioned. For the reconstruction we used a graft from the right great auricular nerve.

The anatomopathological study of the tumour revealed nerve fascicles which were notably destructured by intense collagen infiltration, thus preventing recognition of the nerve by conventional haematoxylin–eosin techniques (Fig. 2A). The immunohistochemical study was positive for neurofilaments and vimentin, and negative for S-100 and alpha-actin, thus confirming the existence of nerve fibres (Fig. 2B).

The patient improved after surgery and presented facial function of grade III in the House-Brackmann scale, 10 months after surgery.

**Discussion**

The preoperative study of this case oriented the diagnosis towards a facial nerve schwannoma located at the level of the geniculate ganglion. Facial nerve schwannomas are relatively rare tumours. The clinical presentation of these tumours depends on their location. The first presentation symptom is usually facial paresis, which may be acute or have a slow onset. Depending on the location, transmissive or sensorineural hearing loss may also appear.

The imaging diagnosis of these lesions is based on the joint use of cerebral MRI, which provides information about the origin and extent of the tumour, and CT scans, which give details on the adjacent bony structures. There are no pathognomonic radiological findings. Facial nerve schwannomas are usually iso- or hypointense on T1-weighted MRI sequences, with notable gadolinium uptake.

![Figure 1](http://www.elsevier.es) (A) Computed tomography image showing an osteolytic lesion with sharp contours and multiple lobes in the area of the right geniculate ganglion, compatible with a facial nerve lesion. (B) Magnetic resonance imaging scan in T1-weighted sequence showing a hyperintense lesion of about 8 mm in diameter located in the right geniculate ganglion.

![Figure 2](http://www.elsevier.es) (A) Preparation of haematoxylin–eosin technique showing intense collagen infiltration and unable to distinguish nerve fibres. (B) Immunohistochemical technique showing positivity for neurofilaments and vimentin, thus confirming the existence of nerve fibres.
The differential diagnosis of geniculate ganglion lesions through imaging techniques is complex and only the anatomopathological study can offer a definitive diagnosis. Infiltration of collagen fibres with intense destructuring of nerve fibres which was only defined through immunohistochemical techniques did not fit, after an exhaustive literature review, with any type of tumour included in the differential diagnosis of the geniculate ganglion. At first, this suggested another rare tumour of the temporal bone, such as inflammatory pseudotumour or inflammatory myofibroblastic tumour. However, the characteristic myofibroblastic proliferation was absent. Therefore, the denomination of fibrous connective tissue lesion of the facial nerve, was more correct.

A case with similar findings in the surgical specimen has been described (collagen infiltration with destructuring of nerve fibres), but associated to the vestibular nerve, mimicking the presentation of a schwannoma, as in our case.

Regarding the therapeutic management of facial nerve tumours, several authors have established a response pattern which depends on the degree of facial function. Observation is indicated up to grade III on the House-Brackmann scale, after which, surgery is the most widely accepted option. This algorithm is based on the fact that the best result achieved by all facial nerve reconstruction techniques is grade III on that same scale. Some authors have debated between observation and decompressive surgery for cases of facial neurinomas with good facial function (grades I–II).

The literature contains very few works on the use of radiotherapy and steroids for facial schwannomas: 1 study describes the use of stereotactic radiosurgery in 2 patients with tumour growth control at 29 and 56 months.

In our case there was no doubt about the use of surgery, since we were faced with a patient with suspected facial nerve schwannoma, with complete paralysis and over 1 year of evolution.

Conflict of Interests

The authors have no conflict of interests to declare.

References