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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KEYWORDS
Facial nerve; Schwannoma; Inflammatory pseudotumour; Temporal bone; Fibroma

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.

PALABRAS CLAVE
Nervio facial; Schwannoma; Seudotumor inflamatorio; Hueso temporal; Fibroma

Nueva entidad en el diagnóstico diferencial de los tumores del ganglio geniculado: lesión del tejido conectivo fibroso del nervio facial

Resumen Dentro del diagnóstico diferencial de las lesiones del ganglio geniculado nos encontramos principalmente con los schwannomas, hemangiomas y meningiomas. Se presenta el caso de un paciente cuya clínica y hallazgos radiológicos imitaban la presentación de un schwannoma del nervio facial. Los estudios anatopatológicos revelaron una lesión con fascículos nerviosos desestructurados por intensa colagenización, por lo que se denominó lesión fibrosa del tejido conectivo fibroso del nervio facial.

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 sud-
den 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 normoacusia.

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 gan-
glion, 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 res-
onance 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 reconstruc-
tion we used a graft from the right great auricular nerve.

The anatomopathological study of the tumour revealed
nerve fascicules 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 diagno-
sis 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 schwan-
nomas are usually iso- or hypointense on T1-weighted MRI
sequences, with notable gadolinium uptake.

Figure 1  (A) Computed tomography image showing an oste-
olytic 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 diam-
eter located in the right geniculate ganglion.

Figure 2  (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 anatomo-pathological study can offer a definitive diagnosis. Infiltration of collagen fibres with intense destructuring of nerve fibres which was only defined through immuno-histochemical 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.3

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.3 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).4

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.5

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**