GENICULATE GANGLION TUMORS. THERAPEUTIC AND RECONSTRUCTIVE MANAGEMENT

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ABSTRACT

We present five cases of patients diagnosed of a facial nerve tumoral lesion localised at the Geniculate Ganglion and all of which underwent surgical resection. The initial symptoms were in four cases of peripheral facial nerve paralysis and in the fifth case facial paresis. Two transfacial extra-labyrinthine approaches were performed and one through a combined path (middle fossa and transfacial) with reconstruction of the facial nerve through a termino-terminal graft. In the fourth case, an approach through fossa media was done, and did not include nerve reconstruction but palliative treatment with a palpebral gold plaque. In the fifth case, a modified trans-labyrinthine approach with facial-hypoglossus termino-terminal anastomosis. Histological diagnosis was 3 neurinomas and 2 hemangiomas. Of the termino-terminal grafts two managed a functional Grade III of the House-Brackmann classification. The third one sustained a Grade VI and therefore a suspension with temporal muscle was carried out. The patient with facial-hypoglossus anastomosis is in a functional Grade IV. Geniculate ganglion tumors are in their majority benign and their treatment is the total resection of the tumor. Nerve reconstruction can be primary or deferred with the aid of a nerve graft or anastomosis, being necessary palliative techniques when resection is not possible.

INTRODUCTION

Primary facial nerve tumours are rare entities, and usually benign, which first symptom is frequently a peripheral facial nerve palsy. They can be difficult to diagnose and when suspected, image test need to be requested to exclude a tumour.

Amongst the possible causes be can include primary facial nerve tumours, local and distant metastasis. This includes 5% of all facial nerve palsies. The typical clinical history is progressive or torpid peripheral facial nerve palsy, although we must be aware of an “acute” peripheral facial nerve palsy, especially if it is associated to atypical Bell’s palsy symptoms (hemifacial spasm, hearing loss, dizziness, tinnitus) a mass seen in the otoscopy, severe changes on the electromyogram (EMG) or ipsilateral recurrence. Amongst malignant tumours, parotid tumours are included, especially squamous cell carcinoma and cystic adenocarcinoma and tumours of the External Auditory Canal (squamous, cutaneous, primitive neuro-ectodermal……) showing severe otorrhea, otorrhagia and pain, in this case, the facial palsy is a sign of bad prognosis. Temporal metastasis with neuralgia type pain and meningeal metastasis of the PCA are also malignant and diagnosis is mainly done by cytology of the cerebrospinal fluid (CSF).

The majority of the peripheral facial nerve palsies develop from benign tumours, primarily from the Fallopian tube (neurinomas, meningiomas and angioleiomyomas) or from the temporal bone or its surroundings (paragangliomas, meningiomas, adenomas, aquatic neurinomas…).

Neurinomas are the most common type of facial nerve tumours and they typically affect the geniculate ganglion and the mastoid part of the nerve. Their growth is by continuity along the nerve and only in one third of the patients go through the otic capsule, which is explained by the slow growth of the tumour. Conductive hearing loss can also happen when the mass involves the ossicular chain. A low percentage of these patients are enclosed in the two clinical features of neurofibromatosis. The alteration of facial movements is often delayed and in most cases the first symptom is a peripheral facial nerve palsy of progressive evolution.

Hemangiomas of the facial nerve are even more rare and they originate in the perineural vascular plexus that is why they tend to localise in the geniculate ganglion. They produce much earlier symptoms than neurinomas, due to extrinsic compression of the neurological trunk. Conductive hearing loss is more frequent than sensorineural one in advanced tumours as they less frequently affect the otic capsule. Regarding the histology, the hemangiomas are cavernous with big vessels, which are covered by endothelium and bone trabeculae inside the tumour. In the early stages they can be dissected by preserving the whole nerve.

The diagnosis is based on the clinical and radiological findings, being in this case complementary, and not excluding the Computed Tomography (CT) and Nuclear Magnetic Resonance (MRI). It is necessary to evaluate the bone surrounding, which is the temporal bone by CT, but also neurological structures of the pontocerebellar angle and the internal auditory canal in order to find out the real extension of the tumour along the nerve.

We will make a special reference to the surgical treatment of these primary facial nerve tumours localised in the geniculate ganglion and to the possibilities of nerve reconstruction by nerve graft or anastomosis as much as well as the palliative techniques when this reconstruction is not possible.

MATERIAL AND METHODS

We present five cases of primary facial nerve tumours, which originate in the region of the geniculate ganglion in five patients between 26 and 72 years old, three females and two males. In four cases the initial symptom was a progressive peripheral facial palsy of more than 6 months evolution; one patient showed an isolated paresis of the marginal branch of the facial nerve.

The treatment of benign tumours does not depend on the histology but on the CT and MRI radiological findings, which are complementary in the study of facial nerve tumours.

Clinical case 1: A 41 year-old male with a 6-month evolution palsy of the marginal branch of the facial nerve. The CT scan showed an epitympanic lesion medial to the head of the hammer suggesting a primary cholesteatoma but in the MRI the lesion capted paramagnetic contrast. As a neurinoma was suspected, a transmastoid extra-labyrinthine approach was...
used with reconstruction of the ossicular chain using malleolus estapedopexy with autologous incus and reconstruction of the facial nerve using nerve graft from the major auricular nerve. On a second surgical time, and under local anaesthe tic, we proceeded to insert a gold plate in the upper eyelid to protect the eye until we had proved evidence that the graft was functional.

Clinical case 2: A 59 year-old male with cutaneous neurofibromas and a plexiform neurofibroma in the lower left limb which shows grade VI House-Brackmann progressive peripheral facial palsy which had been developing for 3 years. CT scan showed a mass with calcified areas in the geniculate ganglion compatible with a meningioma (figure 1). Surgical approach was a combined middle fossa and trans-mastoid one with reconstruction of the facial nerve with a graft from the major auricular nerve.

Clinical case 3: A 72 year-old female with grade VI House-Brackmann progressive peripheral facial palsy for 7 months and whose MRI (figure 2) showed a primary facial nerve tumour with an intracranial component which is surgically approached through the middle fossa. In this patient, primary nerve reconstruction was not carried out but a gold plate was placed in the upper eyelid.

Clinical case 4: A 58 year-old female with a two year evolution bilateral and symmetric sensorineural hearing loss, that for the last six months also had left peripheral facial palsy grade IV House-Brackmann, who was operated through a modified trans-labyrinthine approach in order to reach a tumour in the geniculate ganglion area, that was extending across the labyrinthine segment to the internal auditory canal and to the pontocerebellar angle (figure 3). On a second surgical time and using a graft from the major auricular nerve, a termino-lateral anastomosis between the facial and the hypoglosal nerve was performed following M. May’s technique

Clinical case 5: A 26 year-old male with a progressive peripheral facial palsy of 3 years evolution and with a CT finding of a tumour coming from the epitympanum and involving the
second and third parts of the facial nerve (figure 4) and in which the MRI showed an intense captation of gadolinium. An exploratory tympanotomy was performed which showed a tumour of the first part of the facial nerve extending to the tympanic and mastoid parts. After assessment in the tumour we proceeded to a transmastoid extra-labyrinthine dissection and to the reconstruction of the facial nerve by a graft from the major auricular nerve.

RESULTS

All the patients were followed-up six months after the surgery and MRI performed and no signs of tumour recurrence were seen.

Histologically cases 1 and 2 were reported as hemangiomas of the facial nerve. Cases 3, 4 and 5 as neurinomas of the facial nerve.

Three termino-terminal nerve grafts were carried out, two of which obtained an optimal result with a grade III on House-Brackmann classification. The third patient did not show signs of reinnervation and had a gold plate implanted in the eyelid and a hemifacial suspension with temporal muscle was performed.

The patient who was operated on a second time by facial-hypoglosal termino-lateral anastomosis also obtained a good result with grade IV on House-Brackmann.

DISCUSSION

In all the cases presented the symptom that took us to the diagnosis was the affectation of facial movements, always with an evolution of more than six months with no auditory or vestibular alteration due to the tumour. Such alterations are less frequent in facial nerve tumours in this location than in tumours located in the internal auditory canal.

In the image tests, schwannomas and hemangiomas show very similar characteristics in MRI, they are hypo or isointense in T1, and hyperintense in T2, and more intense after administering gadolinium. The CT scan is necessary to assess dilations of the intralabyrinthine fallopian tube of the geniculate ganglion area or of the vertical part. Nowadays CT and MRI scans are essential to decide treatment of these tumours. In case 1 the CT was compatible with a primary cholesteatoma but the mass captured clearly the paramagnetic contrast, being this compatible with benign tumours.

Primary hemangiomas of the facial nerve tend to come from perineural vascularisation growing from the periphery of the tumour, and for this reason they are more common in areas where this is rich, as are the geniculate ganglion and the internal auditory canal. The hemangioma in case 1 had an intraneural growth, as the neurological fibres surrounded the tumour completely with the dye of protein S-100 (figure 5). It is not really known if they are true tumours or vascular malformations which makes it difficult to classify them. From their natural history we know that they have a slow growth and that the most common early symptom in all series is a progressive peripheral facial palsy.
Regarding neurinomas, they can originate in any part of the facial nerve but 2/3 of them do so in the geniculate ganglion. The initial clinical sign is the progressive peripheral facial nerve palsy but versus hemangiomas are more commonly associated to another symptom, which in more than 50% is hearing loss or tinnitus. The starting MRI meant an advance in the early diagnosis of neurinomas of the geniculate ganglion as it is more sensitive to tumours in this area.

The differential diagnosis is only possible with CT when the hemangioma shows a “salt and pepper” aspect, being surrounded by new bone. We must differentiate them from the cholesteatoma, which has also been seen as a cause of peripheral facial palsy. The post-traumatic facial neurinoma has been associated to chronic inflammation in the ear. We must always bear in mind that we may found histologies in these tumours such as angioleiomyomas, meningiomas, and perineuromas.

The most delicate aspect in the treatment of these tumours is to decide when to perform surgery and which type we choose. This is even more complicated when the patient has been diagnosed of a facial nerve tumour but does not show any symptoms of nerve palsy. The best functional result we can anticipate after a facial nerve graft is a grade III-IV of House-Brackmann. On the other hand waiting too long before dissection of the tumour could affect significantly the final result of the facial function. It is a well-known fact that those patients with a well-established facial palsy have worse results following dynamic reinnervation techniques than those who had a better function or no facial affection before the surgery. Some authors prefer to carry out a decompression of the nerve and to maintain a good facial function for a few more years.

Before approaching the tumour, one must be prepared to explore the three portions of the facial nerve and the pontocerebellar angle. Likewise the condition of the hearing is of vital importance before deciding the type of surgery.

Tumours, which extend medially to the geniculate ganglion and with a good ipsilateral hearing can be approached via the middle fossa. All tumours with a distal extension to the geniculate ganglion require a transmastoid approach. The combination of both techniques gives access to the nerve from the cerebral trunk to the stylomastoid foramen. When the tumour extends from the geniculate ganglion to the tympanic part of the nerve, the transmastoid extra-labyrinthine approach allows us access to the complete mastoid and tympanic part, the geniculate area and the beginning of the labyrinthine portion. In these cases we must be prepared to, by using intraoperative biopsies, extend the approach to middle fossa in case the tumour extends to the geniculate. As this approach requires the resection of the incus and the head of malleus it is necessary to reconstruct the ossicular continuity using a stapledomealoeolopexy by interposing the autologous incus.

For patients with no useful hearing the translabyrinthine approach would be the most appropriate, being useful also the transotic approach. King and Morrison also used the infratemporal approach in 2 cases with serious hearing problems, tinnitus and vertigo. In our case we used a modified translabyrinthine approach with subsequent obliteration of the cavity with abdominal fat and closure of the external auditory canal.

Regarding the reconstruction of the facial nerve, our first choice was to use a termino-terminal graft from the major auricular nerve, due to the similarity of the calibre, and as it is in a surgical field accessible with a single incision, but other authors use the sural nerve or the internal cutaneous brachial nerve. The best results are obtained in patients with a short-term paralysis and a good facial function before treatment. Another possibility is termino-lateral heteroneurological anastomosis normally with the hypoglossal nerve and following the modified May’s technique of interposing a graft from the major auricular nerve in a termino-lateral position. With this technique we can obtain similar results to those from the termino-terminal graft avoiding the ipsilateral lingual atrophy that may come from direct anastomosis.

When the reinnervation technique fails or cannot be carried out due to muscular atrophy, transposition of the temporal muscle or a cross graft is required. These techniques require to work no the orbicular muscle separately, in order to maintain a better symmetry at rest and on movement.

Usually, while waiting for the facial muscles to work autonomously, we place a gold plate adjusted in weight into the upper eyelid of the affected side. It is removed once the competence of the orbicular muscle is proved.
Other static techniques include ocular tarsorrhaphy when there is a high risk of corneal damage and the advancement of the lower eyelid.

CONCLUSIONS

Primary tumours of the facial nerve are rare but they have certain clinical symptoms, which should take us to suspect the existence of a tumour. Radiological exam must be performed by CT and MRI scans as both help to the differential diagnosis of these lesions. The first election treatment should be dissection and primary or differed interposition of a termino-terminal graft. Other dynamic techniques are facial-hypoglosal termino-lateral anastomosis and the suspension with temporal muscle.

Palliative techniques are essential to avoid complications when reinnervation is not feasible.

REFERENCES