MANAGEMENT OF FACIAL PALSY CAUSED BY OTOLOGIC PATHOLOGY – CASE REPORT

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ABSTRACT

Facial palsy is an invalidating condition especially through the associated aesthetic sequelae. Early diagnosis followed by optimal treatment is of crucial importance for functional rehabilitation of the facial nerve. The first step is the establishment of the etiological factor. One first clue regarding the etiology might be represented by the onset mode of the facial palsy but the value of this sign is questionable. There are cited cases in the literature when a benign or malign tumor will be noticed after sudden onset of facial palsy and not as usual after a progressive symptomatology. This study reports a case of peripheral recurrent facial palsy of congenital cholesteatoma etiology. It is described the onset of the palsy, patient evaluation protocol with special emphasis on differential diagnose, stages needed for treatment and post-operative follow-up.

Keywords: congenital cholesteatoma, peripheral facial palsy

INTRODUCTION

Tumors involving the temporal bone have different onset symptoms, but usually can be associated with progressive facial palsy [1; 2]. Nevertheless there are cases when the facial palsy occurs suddenly. Persistent symptomatology or recurrence after medical treatment raises the suspicion on a tumoral mass development along the facial nerve [3; 4].

Regardless of way the facial palsy begins, full ENT evaluation of the patient is mandatory and usually imagistic exams (CT, IRM) indicate the localization and extension of the lesions [5; 6; 7].

The occurrence of facial palsy secondary to temporal bone cholesteatoma is around 20% [4]. Out of all cholesteatoma of the temporal bone, the congenital etiology can be incriminated in up to a quarter of cases (3,7-24%) [8].

CASE REPORT

A 22 years old patient, G.V., was admitted in the ENT Department of the Clinical Rehabilitation Hospital, Iasi, Romania for unilateral facial palsy on the right side.

The medical history shows a previous paralysis of the ipsilateral facial nerve six years ago with sudden onset that benefited from medical treatment (Corticosteroid and Carbamazepin) in the Neurology Department, with full remission of the symptomatology but without establishing any etiology at that time.

There were no further complaints for five years. The facial palsy suddenly reappeared and the patient was admitted again in the Neurology Department. The patient refuses further investigations for a period of one year,

when a CT examination was performed and the patient was diagnosed with peripheral facial palsy on the right side secondary to a temporal bone mass. The patient was transferred in our clinic (ENT Department, Clinical Rehabilitation Hospital in Iasi) for further investigations and treatment.

The clinical evaluation showed a V-th degree - House-Brackmann Score [9; 10] peripheral facial palsy on the right side. The otomicroscopic exam of the right ear showed a normal and permeable external auditory canal and a normal looking tympanic membrane. Through the transparency of the membrane a tumoral mass could be seen, with round shape and white color, located anterior to the malleus handle (fig. 1).

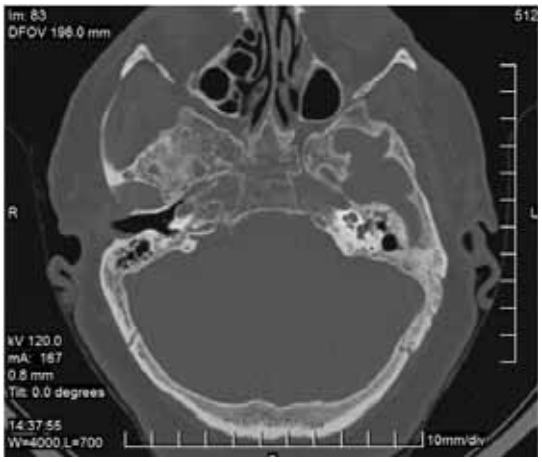

Figure 1. Congenital Cholesteatoma of the left ear

The left ear showed no pathological findings. The patient also suffered from right hemifacial hypoesthesia and neuralgia.

The pure tone audiogram showed an average 50dB mixed hearing loss in the right ear, with an important transmission component.

Electric examinations of the facial nerve showed a 0.5ms difference between the chronaxy of the 2 nerves, suggesting the denervation of the one on the right side.

Summing the data from the clinical exam and paraclinical findings the diagnostic was formulated: Congenital cholesteatoma of the right ear, Secondary recurrent peripheral facial palsy and Secondary Trigeminal Neuralgia.

Surgery of the ear under general anesthesia was performed, by endomeatal approach: a tympanomeatal flap was created in order to visualize the tympanic cavity. A round, pearl white mass is discovered, in close contact with the tympanic membrane without adhering to it, with extensions in the protympanic area obliterating the Eustachian Tube opening, corresponding to a Stage IV cholesteatoma according to Potsic et al., 2002 [11].

Removal of the cholesteatoma was performed but the operating field had to be enlarged to allow the identification of extensions in the tip of the mastoid bone and sinus tympani with erosion on the retrolabyrinthine cells. Finally closed technique mastoidectomy and facial decompression was performed. In the tympanic cavity the intact ossicular chain was identified associated with the dehiscence of the facial nerve canal in the tympanic segment.

Post-operative immediate results were favorable under antibiotic medication (Augmentin). The 7 day post-operative nerve electric evaluation did not show any improvement of muscle excitability.

The patient returns in our clinic after 1 year for right ear otorrhea and unchanged status in the peripheral facial palsy. The otomicroscopy revealed a perforation in the tympanic membrane through which the recurrence of the cholesteatoma could be observed. A revision surgery was performed consisting in open mastoidectomy with obliteration of the tympanic ostium of the Eustachian Tube and elevation of the facial nerve from the bony canal in the mastoid segment. The facial nerve was sectioned between the tympanic and mastoid segments with enough length to be rerouted to reach the

hypoglossal nerve at the level of Farabeuf triangle achieving a termino-lateral suture (fig. 2).

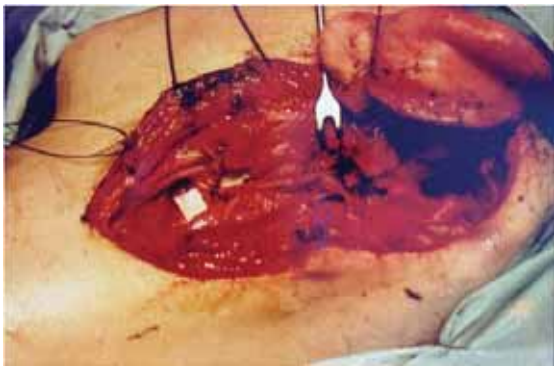

Figure 2. Termino-lateral anastomosis between facial and hypoglossal nerve

At the 6 months follow-up after the last surgery, the mastoid cavity appears free of pathologic elements. The facial muscle tonus is restored and voluntary movement especially in the inferior region of the face is visible (elevation of the right oral commissure and closing of the eyelids). Tongue movement is within normal range.

DISCUSSIONS

Congenital cholesteatoma of the temporal bone is a very rare disease (0.6%) (12). In 1963, Cawthorne [13] suggests that congenital cholesteatoma could be actually a more frequent disease, determining in many cases chronic suppurative otitis media.

It is often considered that facial palsy with sudden onset is the first sign of congenital cholesteatoma.

The criteria for primary cholesteatoma diagnosis were first established in 1965 by Delachi and Clemis [14] and are as follows:

• The presence of a tumor behind a normal tympanic membrane;

• Lack of any previous otic abnormalities (tympanic membrane perforations, acute medium otitis, external auditory conduct atresia);

• Lack of any previous medium ear surgical maneuvers (transtympanic biopsy, miringotomies, transtympanic aerator fixation, etc.)

Cholesteatoma pathogeny is still debated; however, most authors consider cholesteatoma as a congenital disorder that develops from ectodermal embryonic residues that are present in the temporal bone.

In 2002 Potsic et al [11] recommends the classification of congenital cholesteatoma in four stages depending on size relative to the tympanic membrane and involvement of the ossicular chain and mastoid cell system (Table 1).

Table 1. Staging system for congenital cholesteatoma (Potsic et al., 2002) [11]

Stage 1:	Single quadrant: no ossicular involvement or mastoid extension
Stage 2:	Multiple quadrant: no ossicular involvement or mastoid extension
Stage 3:	Ossicular involvement: includes erosion of ossicles and surgical removal for eradication of disease; no mastoid involvement
Stage 4:	Mastoid extension (regardless of findings elsewhere)

In our case the patient had a congenital cholesteatoma that originated from the protympanum with no ossicular chain erosion.

Silent at first, cholesteatoma can sometimes extend towards the middle ear, usually setting out as a progressive facial nerve paralysis [15]. Facial nerve paralysis sets out either through cholesteatoma direct action on the bony canal, or, as in the case of a congenital facial dehiscence through a direct facial nerve cholesteatoma compression. Another possibility would be that of a nerve edema inside the canal through vascular drainage blocking.

In our case, the onset of a recurrent facial nerve paralysis would imply that first it was an intracanalicular facial nerve edema that

healed after anti-inflammatory medication. The persistence of the cholesteatoma in the middle ear induced, in time, an erosion of the bony canal with an irreversible paralysis or a compression on the already dehiscent facial nerve. The patient showed only barely perceptible motion with face asymmetry at rest, no motion in the forehead, incomplete closing of the eye and just slight movement of the mouth receiving a grade V on the House-Brackmann facial nerve grading scale.

An early diagnosis through CT scan followed by an appropriate treatment would have allowed a complete remission of the facial nerve paralysis [16]. The facial nerve paralysis patient needs an immediate, complete exploration. For example, May et al (1984) [17] suggested that a tumoral origin is incriminated in 20% of the ipsilateral recurrent facial nerve paralysis.

Taking into account that the patient did not show a complete Grade VI facial palsy, accompanied by the degeneration of motor end plate, we decided to try the termino-lateral anastomosis between the facial nerve primary trunk and hypoglossal nerve even though more than 2 years passed since the permanent facial palsy onset. This kind of anastomosis was chosen in order to avoid the inconveniences of a lingual palsy [18; 19] that would have been imminent in the case of a termino-terminal anastomosis, even in a partial hypoglossal nerve section [20].

The results after 1 year was encouraging, the patient being able to mobilize labial commissure, close the eye completely with effort and have moderate movement of the forehead with a facial expression with normal symmetry and tone at rest. A good result was also probably achieved due to the fact that the patient was young.

The results are stable at 5 years from the anastomosis.

CONCLUSIONS

Without a clear etiological diagnose in the first moments of the initial facial palsy onset, the patient has lost the opportunity to attempt the surgical removal of the cholesteatoma and hope for normal function of facial motor muscles.

In this case the solution of termino-lateral anastomosis between the facial and the hypoglossal nerve proved to bring benefit regarding facial movements, improving the overall staging from grade V to grade III on the House-Brackmann facial nerve grading scale.

We recommend this type of anastomosis, through the results obtained, to be a good option for the patient considering a proper surgical technique.

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