Facial Paralysis Due to Recurrent Cholesteatoma Seventeen Years After First Surgery

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Abstract:
We present a case of facial nerve palsy induced by recurrent cholesteatoma in a quiescent ear, 17 years after initial mastoidectomy. The pathogenesis, clinical and radiological findings and the management strategies are discussed with an emphasis on regular long-term follow up.

Key words: Cholesteatoma, Recurrent, Facial palsy, Mastoidectomy

Introduction:
Recurrent disease still remains a problem to be solved in the management of middle-ear cholesteatoma to ensure a long-lasting remission. Complete removal of the cholesteatoma matrix is the aim that is not always feasible at the initial surgery even with the advent of modern microsurgical techniques. The spectrum of complications induced by cholesteatoma is very wide ranging from mild quiescent to severe life-threatening ones. We report a case of a recurrent cholesteatoma presenting as facial nerve palsy 17 years after the initial surgery.

Case Report:
A 27 year-old female presented to the A & E Department of Hamad General Hospital (Doha) with a history of rapidly progressing left facial weakness of ten days duration. She had no ear pain, discharge, swelling or recent hearing loss but there was a past history of ear surgery 17 years back in the same hospital when, aged ten, she had an attic cholesteatoma that was managed by a modified-radical mastoidectomy, canal-wall-down procedure. There had been no facial weakness before or after the surgery and she had disappeared from follow-up after the second post-operative visit. Clinical examination revealed a grade 4/6 House-Brackman left-sided facial weakness, an old post-auricular scar and severe external auditory canal stenosis. Retained wax was difficult to remove and made it impossible to inspect the tympano-mastoid cavity but the ear was totally dry.

The patient was investigated by audiometry (50 db conductive hearing loss) and by temporal bone CT scan (Figure 1) which demonstrated the presence of an expansile soft tissue mass within the external canal, middle ear and mastoid cavities, eroding their walls and incorporating the facial canal. The inner ear structures and the tegmen seemed to be spared. That picture raised the possibility of a malignant neoplasm and so an MRI scan was requested (Figure 2) which confirmed the presence of a mixed signal intensity mass (in T1 and T2-weighted images) in the middle ear cavity suggesting either a cholesteatoma or a poorly vascularized tumor.

Figure 1: Axial temporal bone CT scan showing an extensive lytic soft tissue lesion within the left petrous bone

A revision mastoid exploration using a post-auricular approach confirmed the presence of an extensive keratin-filled sac (about 3 cm in diameter) within the pre-existing tympano-mastoid cavity (Figure 3) plus a retained piece of cotton wool and a narrowed external auditory meatus. The cholesteatoma matrix was peeled easily from the intact bony walls of the cavity. The bony facial nerve canal was partially eroded in its tympanic segment but with preserved nerve continuity in its tympanic and mastoid portions. There were no identifiable middle ear structures apart from the foot-plate of the stapes.

After complete removal of the disease, the bony contours were re-fashioned and a wide meatoplasty was done. The cav-
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Discussion:

Cholesteatoma in children has been the subject of numerous publications in the past two decades but significant controversy remains concerning the natural history and optimal management of this challenging disorder. Specific issues relate to the differences between cholesteatoma in children and adults, canal wall up (CWU) versus canal wall down (CWD) mastoidectomy, the timing and the need for a second-look procedures, the length of post-operative follow up and the potentially unique behavior of congenital cholesteatoma.

To avoid residual cholesteatoma and hence recurrent ear disease or complications, complete removal of the cholesteatoma matrix is necessary at the initial procedure otherwise a second-look operation is indicated. Many authors revised the possible factors predicting residual/ recurrent cholesteatoma: Ogawa et al. divided the expectation of residual cholesteatoma into four groups (‘none’, ‘low-rate’, ‘high-rate’, and ‘certain’) based on the maintenance of continuity of the cholesteatoma matrix during the initial surgery.

While lino et al. determined the risk factors for recidivism...
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by male gender, pars flaccida type of cholesteatoma and the association of otitis media with effusion. The rate of residual/recurrent cholesteatoma after an average of 10 years from the initial surgery (CWD) varied in the reviewed literature from 6%, 17%, 29%, to 38%. However, most authors agree that the rate of residual/recurrent cholesteatoma is directly related to the duration of follow-up; the longer the follow-up, the higher the chance of developing recidivism and the necessity of a lifelong observation especially in the first six years of surgery. In our case the patient was lost to follow-up a few months after her first operation until she presented with a major complication 17 years later; a condition that proved that the cholesteatoma was growing slowly over the years and was possibly aggravated by poor cavity ventilation due to the missed piece of cotton-wool. However, recurrence usually manifests more rapidly in the form of otorrhea, aggravating hearing loss, or inner ear disease.

It is important not to miss cholesteatoma as a primary cause of facial nerve palsy, although neoplastic factors and tumour-like conditions such as cholesteatoma constitute only 5-13% of the aetiology of facial nerve palsy. The most likely pathogenesis for facial palsy in cholesteatoma is a direct compression or enzymatic resorption of the nerve and its bony canal, although there has been a single reported case of nerve transection by a middle ear cholesteatoma. The most vulnerable portion is the tympanic segment, in a case of acquired cholesteatoma (as was our case), or the area of the geniculate ganglion, in a case of congenital type.

CT and MRI scans were very helpful in establishing the pre-operative diagnosis which showed an expansile lytic lesion in the tympanomastoid cavity (on CT scan), and typical features of cholesteatoma namely hypointensity on T1- and hyperintensity on T2-weighted images. If we consider the CT scan findings alone, less common pathologies should also be considered in the differential diagnosis (e.g. facial neuroma, glomus tumors, meningioma, adenoma, adenocarcinoma, osteosarcoma, rhabdomyosarcoma, and metastatic tumors). Whatever was the case, a mastoid exploration was necessary to decompress the facial nerve and to identify the pathology by a tissue diagnosis. The principle of surgical management is to exteriorize and, if possible, to completely excise the cholesteatoma sac without opening the perineural sheath of the facial nerve in case of chronic infection as in our case. Most of the cavity was found to be nicely lined with squamous epithelium, where only a small piece of temporalis fascia was necessary to cover the denuded part of the facial nerve followed by a wide meatoplasty, to allow an easily accessible post-operative cavity for inspection and toilet.

The facial weakness recovered within a few weeks of removal of the compressing mass, as is the experience of most authors. However, there is a role for a primary facial nerve repair (by nerve graft) in patients with complete facial palsy of significant duration pre-operatively and secondary grafting should be considered when facial nerve function does not recover at all within 6-9 months or when only minimal function is present at 12-15 months after operation.

Conclusion:

The recurrence rate of childhood cholesteatoma can approach up to 70%; therefore a second planned surgery is highly advisable in 6-12 months regardless of the type of primary procedure (CWD or CWU) which is decided on an individual case-by-case basis. Intense follow-up is of prime importance especially in the first three years for early detection of residual/recurrent disease which rarely manifests itself as facial palsy. Radiological diagnostic tools are very helpful to establish the pre-operative diagnosis. The prognosis of facial nerve insult depends on the exact neuropathology (compression, resorption or transection), the delay in ear exploration, and complete eradication of the disease.

References: