Trismus and TMJ disorders as first clinical manifestations in an intracranial acquired cholesteatoma

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SUMMARY

Intracranial extension of acquired cholesteatoma is a rare occurrence that can develop secondary to trauma, chronic otitis media or acquired aural cholesteatoma. The most commonly observed symptoms are headache and hearing loss. The authors report on a rare case of intracranial cholesteatoma presenting with atypic symptoms: swelling and temporomandibular joint disorders.

BACKGROUND

Acquired cholesteatoma consists of epithelial debris that results from the ingrowth of keratinising squamous epithelium from the external ear and the outer lining of the tympanic membrane.[1] Intracranial extension is a rare occurrence that can develop secondary to trauma, chronic otitis media or acquired aural cholesteatoma, with an extension that can go beyond the middle ear, into the middle or posterior cranial fossa. The supratubal cavity is the most common site of spread.[1 2]

The most frequent symptoms are headache and hearing loss, less common are: otorrhea, eardrum perforation, dysequilibrium, facial nerve dysfunction, tinnitus and epilepsy spells.[2 3]

The authors report on a case of intracranial cholesteatoma presenting with swelling and temporomandibular joint (TMJ) disorders.

CASE PRESENTATION

A 22-year-old man presented with a right preauricular swelling, pain localised to the TMJ and trismus.

At the age of 7, for a tuberculous otitis, the patient had undergone a closed right tympanoplasty with reconstruction of the tympanic membrane for anti-inflammatory purposes. The surgical procedure left a hearing reduction. In the subsequent years, the patient did not have of any trouble or symptom until the recent manifestations.

INVESTIGATIONS

The ear examination and otoscopy revealed a cholesteatoma descending from the epitympanic region of the tympanoplasty cavity. No otorrhea was observed. The facial nerve function was normal (House-Brackmann grading score: (1) An MRI and a CT were performed and revealed a right intracranial temporal mass. The lesion appeared to be well circumscribed and capsulated, with no infiltrating aspects. The temporal lobe of the brain was dislocated and constricted without signs of injury or dura invasion (figure 1). The CT scan showed a defect of the squamous temporal bone, anteriorly, and of the anterior aspect of the petrous bone, posteriorly. The erosion of the tegmen tympani and middle fossa plates was also observed, together with the involvement of the glenoid fossa (figure 2). Preoperative pure tone audiometry showed a severe mixed hearing loss with a downward curve in the high frequencies (figure 3).

TREATMENT

The patient underwent surgical resection of the lesion through a combined transcranial and retroauricular approach. A right temporal craniotomy was performed allowing for a wide expose of the multilobulated lesion (figure 4). A 1 cm area of bone erosion was observed in the temporal bone, requiring its partial removal. On the contrary, even though the lesion was adherent to the tegmental dura, it did not infiltrate it and it was possible to preserve it during surgery.

The cholesteatoma sac was opened revealing its content and was completely removed. The erosion of the glenoid fossa was exposed together with the the articular disk and the supratubal recess fistulisation. A temporalis muscle flap was transposed to fill the surgical gap, to protect the dura and to prevent cerebrospinal fluid leakage.

A retroauricular approach allowed to highlight the involvement of the eustachian tube, the presence of a second fistulisation area and the extension of the lesion from the mastoid to the synodural angle. A subtotal petrosectomy was performed, with preservation of the second and third intrapetrosal tracts of the facial nerve. The external auditory canal skin was removed, the abdominal fat was positioned to obliterate the petrosectomy cavity and the Eustachian tube was sealed using a combination of free

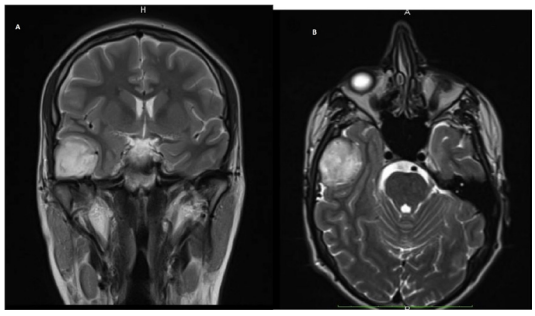

Figure 1 Preoperative MRI, axial (A) and coronal (B) planes.

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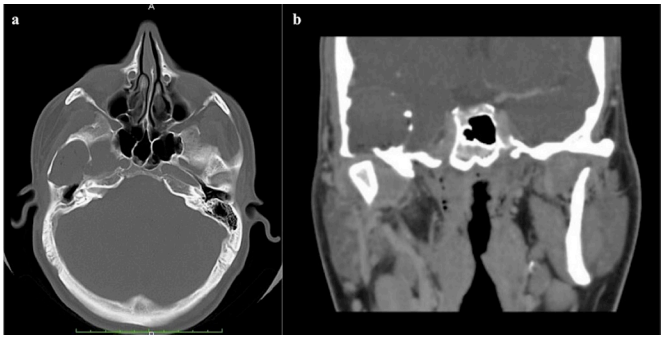

Figure 2 Preoperatice CT-scan, axial (A) and coronal (B) planes showing erosion of the temporal bone and of the middle fossa plate with involvement of the glenoid fossa.

muscle graft and fibrin glue. A cul-de-sac closure of the external auditory canal was finally performed.

OUTCOME AND FOLLOW-UP

The postoperative recovery was uneventful and no cerebrospinal leakage was observed. No changes in facial nerve function were detected, with a postoperative House-Brackmann grading score of 1. The histological examination on the operative specimen confirmed the diagnosis of cholesteatoma. The patient showed no signs of relapse during a 96-month follow-up period.

DISCUSSION

Only case reports and small case series are reported in the literature about the intracranial extension of acquired cholesteatoma, it is in fact a rare occurrence.[1–6] Congenital and acquired cholesteatomas can spread intracranially from the temporal bone into either the middle of the posterior fossa. The fundamental steps involved in the intracranial spread have been reported to be: entrapment of the cholesteatoma in a narrow recess, erosion of the bone of the dural plates, and expansion of the cholesteatoma into the cranial fossae.[7]

The reason for the above is the locally destructive nature and the ability to erode bone that characterises the cholesteatomas. Several mechanisms have been proposed to account for this behaviour, including secretion of osteolytic enzymes, pressure necrosis, osteitis and surrounding chronic granulation tissue.[8–10] A wide variety of symptoms are described basing on the location and the extension of the lesion. Clinical presentation can be characterised by: hearing loss, facial nerve paralysis, headache, tinnitus, vertigo, otorrhoea, eardrum perforation, epilepsy spells.[2 3] Since a gradual intracranial involvement does not

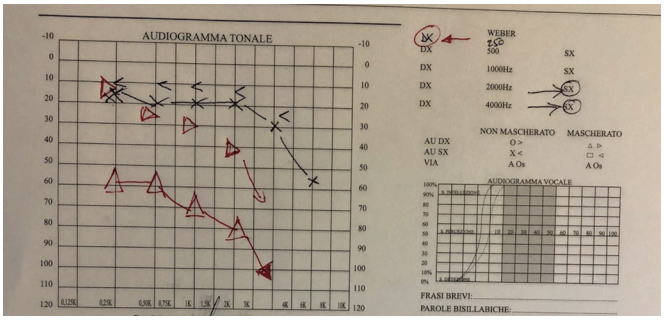

Figure 3 Preoperative pure tone audiometry showing a severe mixed hearing loss with a downward curve in the high frequencies. Dx: right; AU: ear; SX: left; VIA: conduction.

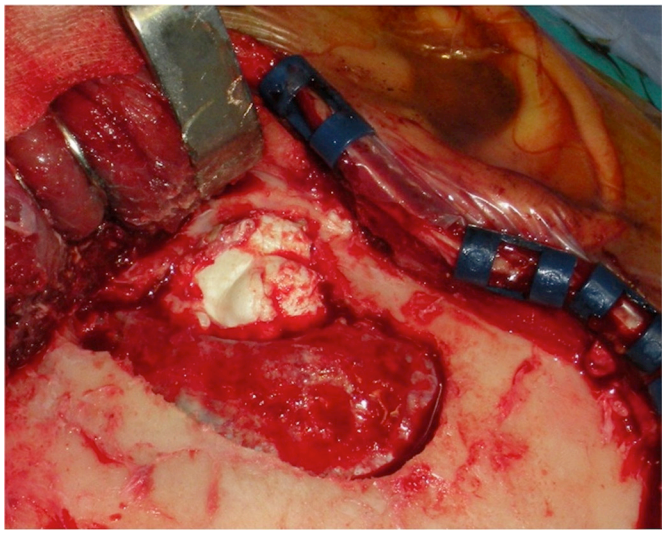

Figure 4 Intraoperative view of the lesion.

usually cause acute symptoms, the correct diagnosis may be difficult and delayed.[4] In the present paper, the authors report on a rare case where the first clinical manifestations of an intracranial acquired cholesteatoma were TMJ disorders. A review of the literature did not show other significant reports about this. The patient arrived the first time to the physicians complaining about a preauricular swelling, pain to the TMJ and trismus. Only otoscopy and radiology allowed to identify the presence of a lesion extending in the middle fossa and eroding the tegmen tympani and middle fossa plates. The glenoid fossa was destroyed and replaced by the lesion. During the surgical procedure, the cholesteatoma removal showed a completely exposed articular disk. The chronic flogosis involving the pterygoid muscles, secondary to the lesion, and the obliteration of the articular space by pathological tissue, were responsible for the symptoms reported by the patient. The complete removal of the lesion and the placement of well-vascularised tissue to fill the defect and to protect the articular disk, allowed to obtain a clinical improvement. Moreover of no less importance was the closure of the eustachian tube, the removal of the canal skin and the blind sac closure of the external ear. One of the main cause of postoperative complications in these patients is, in fact, the cerebrospinal fluid leakage, that leads to an increased risk of infection and to slower recovery times. This is why complete obliteration and cul-de-sac closure is now preferred by many authors, especially in patients with a preoperative severe hearing loss.[8]

Learning points

- The appearance of swelling and disorders of the temporomandibular joint are symptoms of cholesteatoma very rarely.
- Their presence in patients without other problems and with a history of cholesteatoma or ear surgery should prompt the clinician to more in-depth investigations, to exclude the presence of an intracranial cholesteatoma.
- The pathological tissue can erode the glenoid fossa, leading to the obliteration of the articular space, with chronic flogosis and involvement of the pterygoid muscles.

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