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Cholesteatoma of the hypotympanum in a patient with Treacher Collins syndrome

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ABSTRACT

In the present article we report a cholesteatoma of the hypotympanum extending to the jugular foramen in a 16-year-old male with Treacher Collins syndrome. Preoperative imaging excluded jugular paraganglioma and set the diagnosis of cholesteatoma. We discuss the operative treatment via a large hypotympanotomy and creation of an open hypotympanic cavity. To the authors' knowledge this is the first description of hypotympanal cholesteatoma with such an extension, being treated through this approach.

1. Introduction

Cholesteatoma is a non-neoplastic, keratinizing lesion which has two forms: congenital and acquired. Congenital cholesteatoma develops behind a normal, intact tympanic membrane, whilst acquired cholesteatoma is associated with a defect in the tympanic membrane. Acquired cholesteatoma may develop by various mechanisms: immigration, basal hyperplasia, retraction pocket and/or trauma (iatrogenic or non-iatrogenic). Cholesteatoma is most likely to develop in the attic, epitympanum or the posterior mesotympanum. Eradication of disease is the primary surgical goal, followed by maintenance or restoration of hearing. Several surgical techniques have been described with modifications and today there is still no general consensus about the optimal procedure [1]. Occurrence or extension of the disease in unusual sites like the petrous apex, internal auditory canal, middle cranial fossa, infratemporal fossa or the upper neck may be diagnostically and therapeutically challenging [2,3].

We hereby introduce a rare case of a patient with Treacher Collins syndrome presenting with a cholesteatoma of the hypotympanum extending to the jugular foramen, which has been exteriorized via a large hypotympanotomy and an infracochlear approach preserving the facial nerve canal, combined with a large mostly inferior meatoplasty resulting in an open hypotympanic cavity for better follow up and in a reduced tympanic cavity with preservation of normal sound conducting mechanism and hearing.

2. Case report

A 16-year-old male with Treacher Collins syndrome presented with aplasia of both mandibular condyles M3 according to the O.M.E.N.S. classification. This resulted in retrogenia, a severe Angle Class II occlusion. The X-ray revealed residual miniplate of an earlier perfomed genioplasty.

Prior to his presentation at the University of Mainz Medical Center he had been operated three times for his mandibular dysplasia at an age of 4, 5 and 9 years. As a sequelae of earlier surgery a "neo temporomandibular joint (TMJ)" was seen on the left side below the coronoid process. The mandibular retrogenia was typically associated with a transversal deficit in the maxilla. Dental findings were retained teeth 15, 25, 37, 38, 47, 48, decayed 46 and a super numerate upper incisor (mesiodens).

Since the age of 15 he developed purulent, sometimes serosanguinous discharge of his left ear with associated hearing loss and otalgia. Otologic examination revealed bilateral pretragal hypertrophic scars after previous surgeries of the condyles. The external ear canal was narrow on both sides however while the right tympanic membrane was visible and the middle ear

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ventilated, there was discharge from the left ear with edema and polyps originating from the anterior upper quadrant of the left tympanic membrane, subtotally obstructing the external auditory canal and impairing inspection of the tympanic membrane. Pure tone audiograms revealed a pantonal conductive hearing loss of 25 dB on the left and normal hearing on the right.

CT scan showed large bony defect of the jugulare foramen and hence jugular paraganglioma has been suspected (Fig. 1). Examination of the lower cranial nerves showed no functional impairment. In order to clarify further the dignity of the lesion, magnetic resonance imaging (MRI) scan and MRI angiogram were ordered. These revealed a hypointense in T1-weighted and hyperintense in T2-weighted images lesion of the temporal bone, measuring $1.6 \times 1.7 \times 1.8$ cm in diameter with diffusion restriction and without contrast enhancement (Figs. 2 and 3). The lesion was located in the triangle between the hypoglossal canal, the jugular foramen and the internal carotid canal and the diagnosis of cholesteatoma was set. Further MRI findings were hypoplasia of the transverse and sigmoid sinus of the left side and fluid retention in the left mastoid cells.

After obtaining written consent from the patients parents, the patient was subsequently taken to surgery where an infracochlear approach preserving the facial nerve canal combined with hypotympanotomy and meatoplasty were performed while neuromonitoring facial nerve function.

A left retroauricular incision, extended into the neck, was performed. The deep jugular vein was looped and the accessory nerve identified. After entering the external ear canal, polyps and granulation tissue were removed. The ear drum was covered with granulation tissue. As sequelae of earlier surgeries of the condyles, the floor and the anterior wall of the external ear canal were partially missing, demonstrating a bony cavity of the os tympanicum which reached the mandibular joint and which was filled with granulation tissue and epithelium. After elevating the tympanic membrane, the tympanic cavity showed granulation tissue which was removed until an intact ossicular chain could be identified. No cholesteatoma could be seen in the middle ear however there was cholesteatoma matrix in the hypotympanum. A complete hypotympanotomy was performed, totally removing the floor of the external ear canal and granting access to hypotympanum. Infracochlear approach preserving the facial nerve canal was performed. The tip of the mastoid cavity was resected and the vertical part of the facial nerve was delineated, preserving a bony shell over the facial nerve. The inferior to posterior ear canal was drilled out, preserving though the posterior to superior ear canal. Infracochlear approach allowed eradication of the disease and provided an accessorily approach to the hypotympanum, medial to the vertical part of the facial nerve. The cholesteatoma was situated medial, anterior and posterior to the facial nerve, reaching the vertical portion of the internal carotid artery and the jugular bulb anteriorly, the jugular foramen and the hypoglossal canal inferiorly.

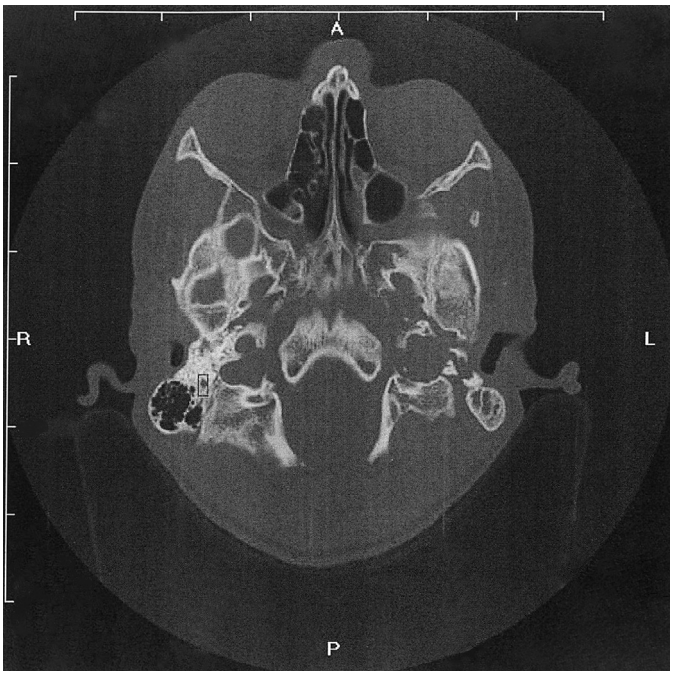

Fig. 1. Axial computed tomography scan showing large bony defect of the left jugular foramen.

The cholesteatoma was removed, the tympanic membrane repositioned and an underlay facia graft was placed to seal the tympanic from the hypotympanic cavity. Subsequently a large mostly inferior meatoplasty was performed sealing opened

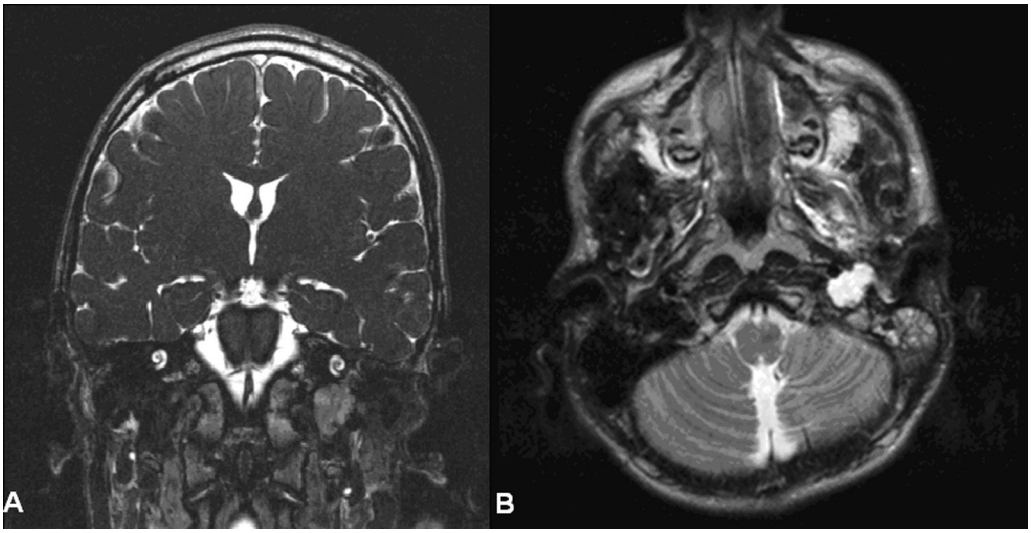

Fig. 2. Magnetic resonance imaging showing the infralabyrinthine cholesteatoma of the left side. (A) Coronal T1-weighted magnetic resonance imaging showing extent of infralabyrinthine cholesteatoma of the left side. (B) Axial T2-weighted magnetic resonance imaging with infralabyrinthine cholesteatoma on the left side adjacent to the internal carotid artery.