International Journal of Pediatric Otorhinolaryngology (2005) 69, 1703–1707

ELSEVIER

International Journal of Pediatric Otorhinolaryngology

www.elsevier.com/locate/ijporl

CASE REPORT

Infantile congenital petrosal cholesteatoma: A case report and literature review

Kazuharu Yamazaki [a,*], **Hiroaki Sato** [a], **Kazuo Murai** [b], **Kaoru Ogawa** [c]

[a] *Department of Otorhinolaryngology, Iwate Medical University, Uchimaru 19-1, Morioka, Iwate 020-8505, Japan*
[b] *Department of Otorhinolaryngology, Iwate Rosai Hospital, Iwate, Japan*
[c] *Department of Otorhinolaryngology, Keio Medical University, Tokyo, Japan*

Received 25 March 2005; accepted 8 April 2005

KEYWORDS
Congenital petrosal cholesteatoma;
Facial nerve paralysis;
Infant

Summary Congenital cholesteatoma of the middle ear is frequently seen in younger children including infant. By contrast, that of the petrous bone is rarely seen in younger children. We report an exceptional case occurring in 23-month-old infant who presented with unilateral facial nerve paralysis at about 18-month-old. Computed tomography and magnetic resonance imaging revealed congenital cholesteatoma measuring approximately 2 cm in the area of the right geniculate ganglion. The mass was completely eradicated via the middle fossa approach, which allowed for preservation of hearing. The facial nerve maintained intact during surgery and paralysis showed partial recovery after the operation. To our knowledge, the present case seems to be the youngest case of congenital petrosal cholesteatoma reported, and also demonstrates congenital petrosal cholesteatoma could exhibit facial nerve paralysis in early childhood.

1. Introduction

Tumors occurring in the petrous part of the temporal bone are predominantly vestibular schwannoma, with cholesteatoma only comprising 4% [1]. Moreover, petrosal cholesteatoma accounts for only 1–3% of all cholesteatomas and is comparatively rare [2,3]. The petrous bone is considered a "silent area", and tumors in the region tend to be asymptomatic in the early stage. Many cases thus display extensive expansion of the tumor by the time symptoms appear.

Compared with cholesteatomas in other regions, facial nerve paralysis is considered characteristic for petrosal cholesteatoma. Cawthorne [4] described facial nerve paralysis as progressing gently and quietly in petrosal cholesteatoma, representing a key point of difference to vestibular schwannoma.

We encountered a case of petrosal cholesteatoma in a 23-month-old girl who developed facial nerve paralysis at 18-months-old. The tumor was removed via the middle cranial fossa approach. This case is thought to be very rare given the age of the patient at

* Corresponding author. Tel.: +81 19 651 5111; fax: +81 19 652 8642.

0165-5876/$ – see front matter
doi:10.1016/j.ijporl.2005.04.027

appearance of facial nerve paralysis, and we discuss the case with reference to the existing literature.

2. Case report

The patient was a 23-month-old girl with no family or previous medical history of note, and no previous episodes of myringitis. Her mother had noticed that movements of the right face decreased at about 18-months-old, and consulted a public health nurse. However, symptoms were attributed to a habit, and no further action was taken. The patient developed chicken pox at 21-month-old, and underwent a checkup at a nearby pediatric clinic. At this time, right facial nerve paralysis was noted, and the patient was referred to our clinic and hospitalized in early 2001 for surgical intervention.

3. Laboratory findings

3.1. Otorhinolaryngological findings

No abnormalities were identified in the ear, nose or pharynx. Movement of the right face was poor, but lagopthalmos was not seen and no leakage of food from the angle of the mouth was present. The facial nerve paralysis was Grade IV by the House—Brackmann classification.

3.2. CT

An expansive lesion was identified above the petrous bone on coronal section and forward of the petrous bone on axial CT (Fig. 1A and B). No relation to the middle ear was apparent. Development of the mastoid air cells was good. A round destruction image was seen above the acoustic meatus in the middle fossa of the petrous bone, on the superolateral aspect of the bone on three-dimensional CT (Fig. 1C).

3.3. MRI

The tumor displayed intermediate intensity on axial sections from T1-imaging, with one region of high intensity. On coronal T2-imaging, the tumor displayed a region of slight hyperintensity above the cochlea and involving the dura mater. Invasion into the brain tissue was not seen (Fig. 2A and B). Cholesteatoma, cholesterin granuloma, mucocele,

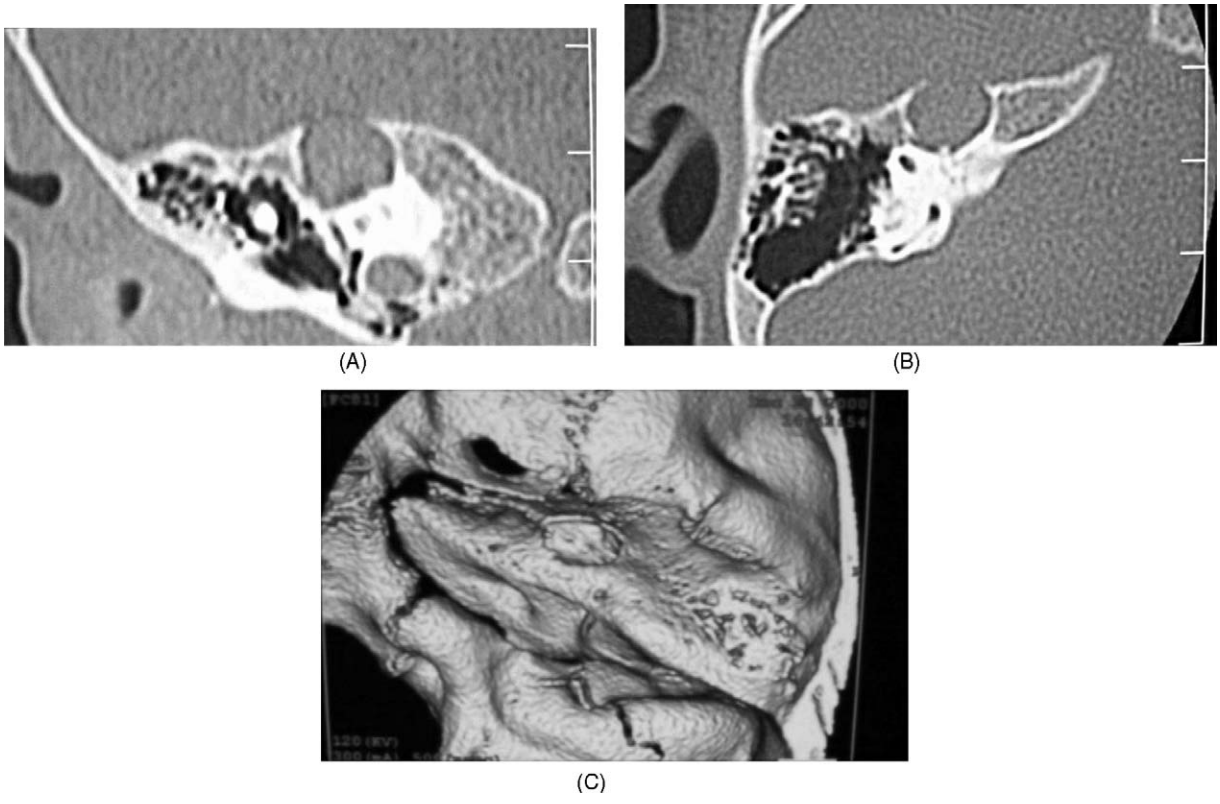

Fig. 1 CT scan of the temporal bone. An expansive lesion was identified above the petrous bone on coronal section (A) and forward of the petrous bone on axial section (B). A round destruction image was seen above the acoustic meatus in the middle fossa of the petrous bone, on the superolateral aspect of the bone on three-dimensional CT (C).