Case Report

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CT and MRI Features of Middle Ear Fibrous Hamartoma of Infancy: A Case Report

중이에서 발견된 영유아 섬유성 과오종의 영상 소견: 증례 보고

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Fibrous hamartoma of infancy in the middle ear is extremely rare. We report the case of a 26-month-old male patient who presented with a mass in the left middle ear. A temporal bone CT scan showed complete opacification of the left middle ear and mastoid air cells without ossicular erosion. On MRI, the mass revealed heterogeneous signal intensities indicative of fat and fibrous components. A definitive diagnosis was made postoperatively based on the histological results. Although rare, fibrous hamartoma of infancy should be considered as a differential diagnosis of a middle ear mass during childhood.

Index terms Middle Ear; Hamartoma; Computed Tomography, X-Ray; Magnetic Resonance Imaging

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INTRODUCTION

Fibrous hamartoma of infancy (FHI) is a rare fibroproliferative soft tissue lesion. FHI usually appears within the first 2 years of life and up to 25% of the cases are congenital (1). The usual locations include the upper extremities, axilla, and upper back, but it may occur anywhere in the body. FHI of the middle ear is extremely rare, with less than four cases reported to date (2). Moreover, there are no reports showing both CT and MRI features in a single case of middle ear FHI. We report a case of middle ear FHI with CT and MRI features.

CASE REPORT

A 26-month-old male patient presented to our hospital for evaluation of a left middle ear mass. The patient had visited a local hospital 5 months ago and the provisional diagnosis was congenital cholesteatoma. The patient had passed a hearing screening at birth. His external ear was normal on examination. Otoscopic examination revealed a whitish protruding mass and middle ear effusion behind the intact tympanic membrane (Fig. 1A). High-resolution CT of the temporal bone showed complete opacification of the left middle ear and mastoid air cells, in which the ossicles were not destroyed (Fig. 1B). No inner ear malformations were observed. The extent of the mass could not be distinguished on CT, since the mass and the effusion had similar attenuation values. MRI was performed for better evaluation of the mass using a 3T scanner (MAGNETOM, Vida, Siemens Healthineers, Erlangen, Germany). On MRI, a well-defined mass with a maximum diameter of 7 mm was observed along the cochlear promontory and mesotympanum in the middle ear (Fig. 1C-E). Coronal T2-weighted imaging (T2WI) showed a mass with peripheral very low signal lining and a central high-signal strand. The high signal intensity was suppressed on axial fat-suppressed T2WI. The mass showed intermediate signal intensity on T1-weighted imaging (T1WI) and heterogeneous enhancement. Diffusion-weighted imaging (DWI) revealed no hyperintensity in the mass.

The patient underwent a canal wall up mastoidectomy with excision of the tumor. Intraoperatively, the malleus handle was embedded in a whitish mass without destruction.

Histological examination was indicative of FHI with fibrous tissue, interspersed adipose tissue, and scattered immature mesenchymal cells (Fig. 1F). Immunohistochemistry showed positive staining for CD34 and CD117 (c-kit) and negative staining for S100 protein in the mesenchymal cells, indicating the presence of immature mesenchymal cells.

This case report was approved by our Institutional Review Board, and the requirement for written informed consent was waived (IRB No. DAUHIRB-21-126).

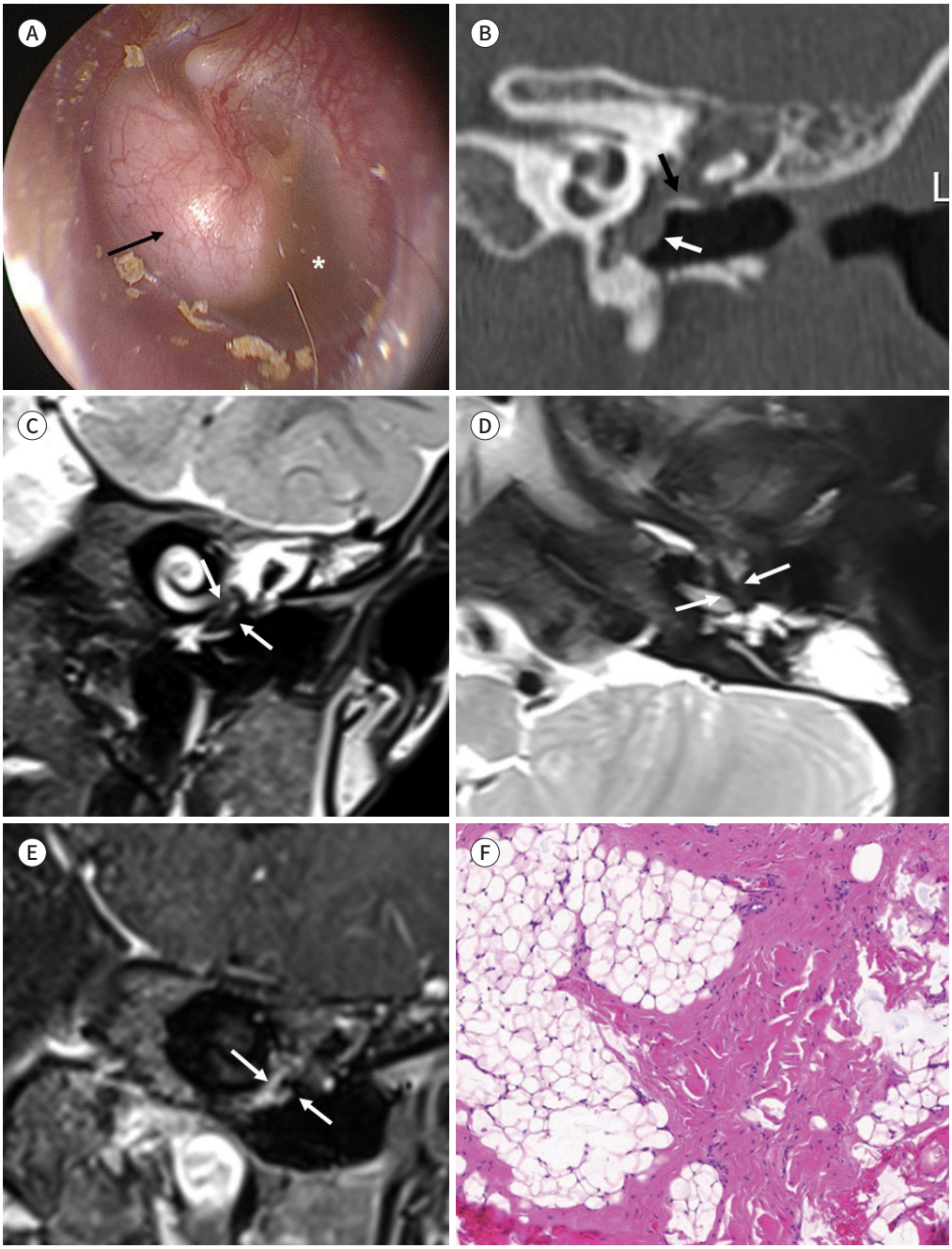
DISCUSSION

FHI is a soft tissue mass with a characteristic morphology including three histologic components: intersecting fascicles of dense fibrocollagenous tissue, loosely textured areas of immature basophilic or myxoid round or primitive mesenchymal cells, and mature adipose tissue (3). FHI is usually diagnosed before the age of 2 years and males are more commonly affected (2.4:1) (4). Most frequently, FHI presents as a solitary nontender soft tissue mass involving the axilla, upper arm, trunk, inguinal region, and chest wall. However, it could also involve a wide variety of anatomical locations including the head and neck region and the distal extremities.

The imaging features of middle ear FHI have not yet been clearly defined due to its extreme rarity. Baget et al. (2) reported a case of a 5-month-old female with middle ear FHI wherein CT demonstrated diffuse opacity of the middle ear without any clearly defined osteolysis. The ossicular chain and the inner ear seemed uninjured.

Ji et al. (5) reported MRI findings of two cases, together with the 14 reported cases in the English literature.

Fig. 1. A 26-month-old male with middle ear fibrous hamartoma of infancy.
A. Otoscopic photograph of the left ear shows a whitish mass (arrow) and effusion (asterisk) behind the tympanic membrane.
B. Coronal CT image shows complete opacification of the middle ear and outward bulging of the tympanic membrane (white arrow). Ossicles are embedded, but not destroyed (black arrow).
C. Coronal T2-weighted MR image shows a predominantly low-intensity mass with a central fine strand of high signal intensity (arrows).
D. Fat-suppressed axial T2-weighted MR image shows signal loss of the high signal intensity portion within the mass (arrows).
E. Coronal contrast-enhanced T1-weighted MR image demonstrates heterogeneous enhancement of the mass (arrows).
F. Photomicrograph of pathologic specimen shows an admixture of islands of adipose tissue, scattered immature mesenchymal cells, and dense bundles of fibrous tissue (hematoxylin and eosin, × 100).



The locations of FHIs in these cases included the subcutaneous layer or muscles in the upper extremity, lumbar area, knee, foot, orbit, abdominal wall, and craniocervical areas. However, there have been no reports describing the MRI features of a middle ear FHI. The usually observed MRI manifestation in these cases was a mass with strands showing signal intensity similar to that of adipose or fibrous tissue. The interspersed strands usually exist in a parallel fashion or even in a whirling appearance. Contrast-enhanced T1WI demonstrated heterogeneous enhancement of the mass.

Similar to the previously reported cases, the mass in the present case showed a predominantly low signal intensity of the fibrous component traversed by the fine strand of high-signal fatty component in a parallel fashion.

Differential diagnosis is essential to distinguish FHI from other masses or tumors in the middle ear. Congenital cholesteatoma could be misdiagnosed due to a similar white and retrotympanic appearance on otoscopy. Unlike our case, the presence of erosion of the ossicle, tympanic tegmen, or scutum on CT is indicative of cholesteatoma. On DWI, cholesteatoma appears hyperintense with low values on the apparent diffusion coefficient map and is not contrast-enhanced. Middle ear adenomas may present a diagnostic dilemma, since the majority of the middle ear adenomas do not show erosion of the ossicles on CT. However, the mass is isointense to the gray matter on T2WI and the signal intensity is higher than that of fibrous tissue (6). Dermoids of the middle ear are exceedingly rare benign lesions and the average age at diagnosis is between 2 and 3 years. Dermoids demonstrate variable imaging features, including fat signal intensity, according to their subtypes on MRI (7). Our case also demonstrated high signal intensity similar to that of fat, which showed a marked signal loss in the fat-saturated technique. However, similar to cholesteatoma, the tumor tends to erode the temporal bone or the ossicles, but not in our case.

FHI has a benign nature in almost all cases, but appears to have the potential to progress to malignancy. Rare cases with rapid growth that clinically mimicked sarcomas have also been reported. Al-Ibraheemi et al. (8) reported two cases with features of a primitive spindle cell or round cell sarcoma as well as genetic changes suggestive of malignancy.

The treatment of choice for FHI is complete excision of the envelope of normal tissue. Although local recurrence of up to 16% have been reported, reoperation is sufficient to cure the disease in these cases (9).

In conclusion, FHI is a very rare middle ear tumor that can be clinically confused with other diseases. Whenever there is an absence of ossicular erosion on CT and MRI demonstrates signal intensity similar to that of fat or fibrous tissue, FHI should be considered in the differential diagnosis of a middle ear mass in childhood.

Author Contributions

Conceptualization, K.S.; data curation, B.S.H.; investigation, all authors; supervision, K.S.; visualization, B.S.H.; writing—original draft, B.S.H.; and writing—review & editing, K.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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중이에서 발견된 영유아 섬유성 과오종의 영상 소견: 증례 보고

백상훈 · 김상현* · 임경재

중이의 영유아 섬유성 과오종은 매우 드물다. 저자들은 26개월 남아의 중이에서 진단된 사례를 영상의학적 소견을 중심으로 보고하고자 한다. CT 영상에서는 귓속뼈의 파괴가 없었으며 MRI에서는 지방과 섬유성 조직과 유사한 신호 강도를 보였다. 수술 후 조직학적 검사에서 영유아 섬유성 과오종으로 진단받았다. 중이의 영유아 섬유성 과오종은 드물지만, 유년기에 중이에서 관찰되는 종괴의 감별진단에 포함되어야 한다.

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