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Dermoids of the Eustachian Tube: CT and MR Findings with Histologic Correlation

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Summary: Two patients with congenital dermoids of the eustachian tube presented with recurrent otitis media and chronic otorrhea resistant to antimicrobial therapy. CT demonstrated fat density, homogeneous lesions, filling and expanding the eustachian tube. On MR, signal from the lesions was consistent with fat, and the relationship with the internal carotid artery was better delineated than by CT. Microscopically, the masses consisted of a conglomeration of ectodermal and mesodermal elements.

Index terms: Dermoid cyst; Nasopharynx, cysts

Although dermoids of the head and neck region are well-recognized clinical and histologic entities, reports of dermoids involving the temporal bone are rare, with 24 cases described in the literature (1). Of these cases, 10 originated from the eustachian tube (1–5). Even if histologically benign, these lesions can cause considerable morbidity because of their location. We report the computed tomography (CT) and magnetic resonance (MR) findings in correlation with the histologic features in two cases, and discuss their histopathogenesis, clinical features, differential considerations, and treatment, emphasizing the role of preoperative imaging in choosing an appropriate surgical approach.

Case Reports

Case 1

A 12-month-old girl presented with a history of recurrent episodes of otitis media. At that time, the patient underwent bilateral myringotomies with insertion of pressure equalization tubes. Thick, gluelike material was aspirated from each middle ear space, more on the left than the right side. After pressure equalization tube placement, persistent left otorrhea developed. Because of the suspicion of a cholesteatoma, an exploratory tympanotomy was performed through a postauricular approach. A white, fibrotic mass was seen extending into the posteroinferior quadrant of the middle ear. The mass could not be removed in toto and, after obtaining a specimen for biopsy, the procedure was terminated. Because of the extent of the tumor, radiographic evaluation was scheduled to plan appropriate intervention for a more definitive excision.

CT of the temporal bones with 1.5-mm-thick direct coronal and axial sections, demonstrated a 2.0 × 2.5-cm mass with predominantly fatty attenuation, surrounded by a smooth rim of soft tissue located along the course of the left eustachian tube. The bony origin of the eustachian tube was expanded and the carotid canal was flattened. The mass extended superiorly to the inferior aspect of the middle ear cavity. The middle ear and mastoid air cells were almost completely opacified. The middle ear ossicles were normal in appearance and orientation. On MR, the lesion was homogeneous and followed the signal intensity of fat on T1- and T2-weighted as well as fat-suppressed sequences. Tiny hypointense strands in the center of the mass gave the impression of thin septations. The mass was surrounded by a smooth, thick, hypointense capsule, which enhanced slightly after contrast administration. The left jugular vein and carotid artery were seen posterior and medial to the mass, respectively (Fig 1).

The mass was approached through a combination of a postauricular tympanotomy and a modified neck dissection. The pharyngeal component of the lesion was located under the area of the digastric muscle and was surrounded by a thick capsule that, when dissected, revealed a cystic, fleshy mass containing hair follicles. From the parapharyngeal space, the mass extended superolaterally into an enlarged eustachian tube, from which it was resected. A fleshy mass filled the mesotympanum and appeared to originate from the area of the hypotympanum and eustachian tube. The mastoid was filled with mucoid fluid; however, no mass was noted in the epitympanum or the mastoid. To ensure complete removal of the lesion, the eustachian tube was cannulated from the middle ear to the parapharyngeal space with a suction catheter.

Microscopic examination showed a polypoid mass of connective tissue, covered by keratinizing stratified squa-

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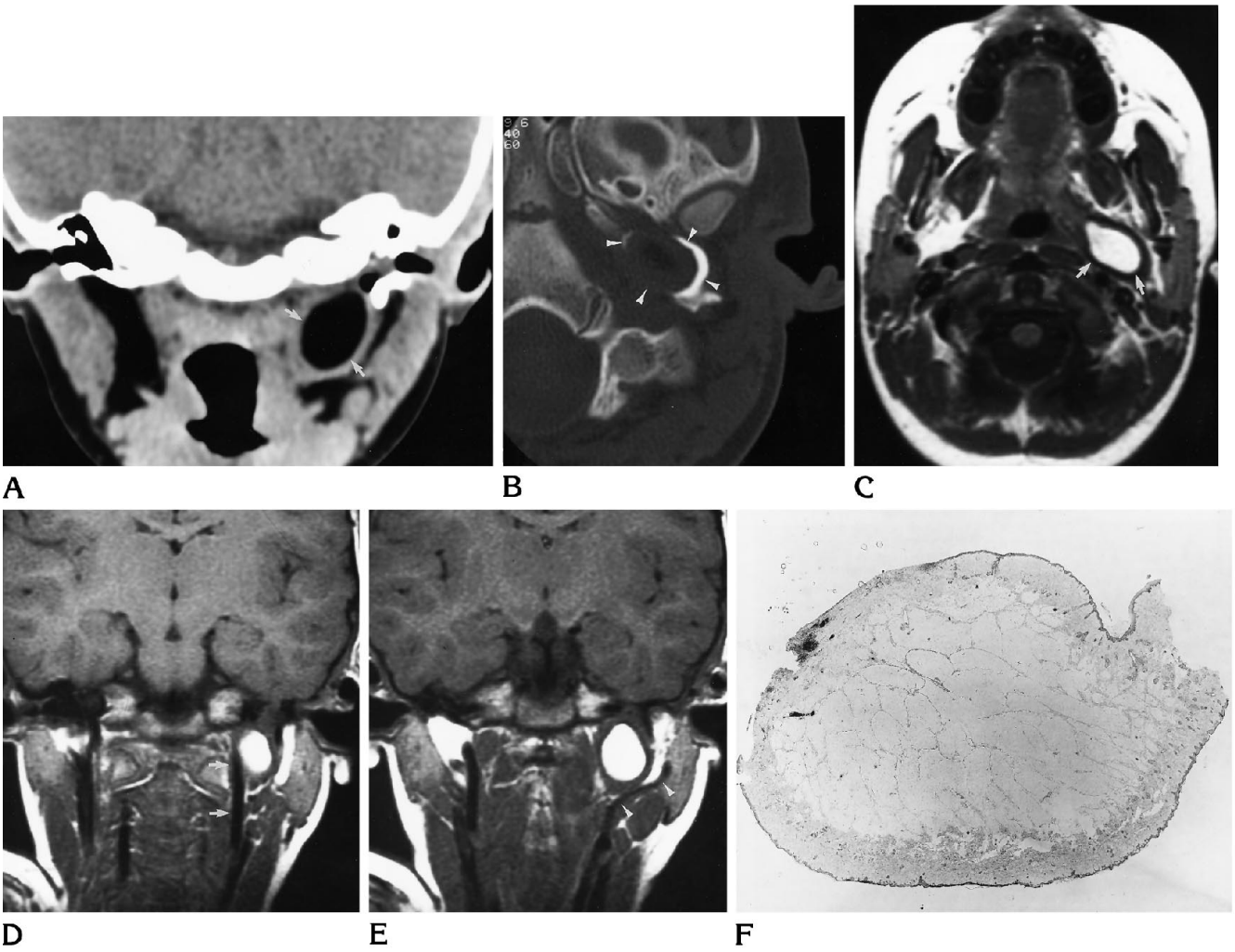

Fig 1. Case 1.

A, Coronal CT scan shows a low-attenuation mass surrounded by a soft tissue capsule (*white arrows*), extending along the left eustachian tube from its bony origin to the left parapharyngeal space.

B, Bone algorithm axial CT scan demonstrates the expansion of the eustachian tube canal by the mass (*white arrowheads*).

C, Axial T1-weighted MR image shows the high-intensity matrix of the lesion and soft tissue capsule around it (*white arrows*).

D and *E*, Coronal T1-weighted MR images reveal the relationship of the mass with internal carotid artery running medially (*white arrows*) and the external carotid artery running laterally (*white arrowheads*).

F, Section through the resected specimen shows that the surface is mature hair-bearing skin and the center consists of lobular adipose tissue.

mous epithelium. Skin appendage structures, including sebaceous glands, hair follicles, and sweat glands, were present.

Case 2

This patient originally presented at 15 months of age with a history of recurrent otitis media. Treatment with multiple antimicrobial agents proved ineffective, and 2 months later the patient was taken to surgery for a tympanoplasty and possible mastoidectomy, for a possible congenital cholesteatoma. A large mass was found filling the entire middle ear space, surrounding the ossicular chain, eroding the incudostapedial joint, and extending into the epitympanum and sinus tympani. The mass appeared more firm and fibrous than expected for a cholesteatoma. A frozen section was submitted and was consistent with a dermoid. The mass was pedunculated and extended down the eustachian tube orifice, which was markedly dilated. A pulsating mass was noted just medial to the fibrous lesion and was thought to be the vertical portion of the internal carotid artery. As much of the lesion as possible was removed before the procedure was terminated because of inadequate exposure.

Radiologic evaluation was undertaken for estimation of the extent of the lesion. A CT of the eustachian canal

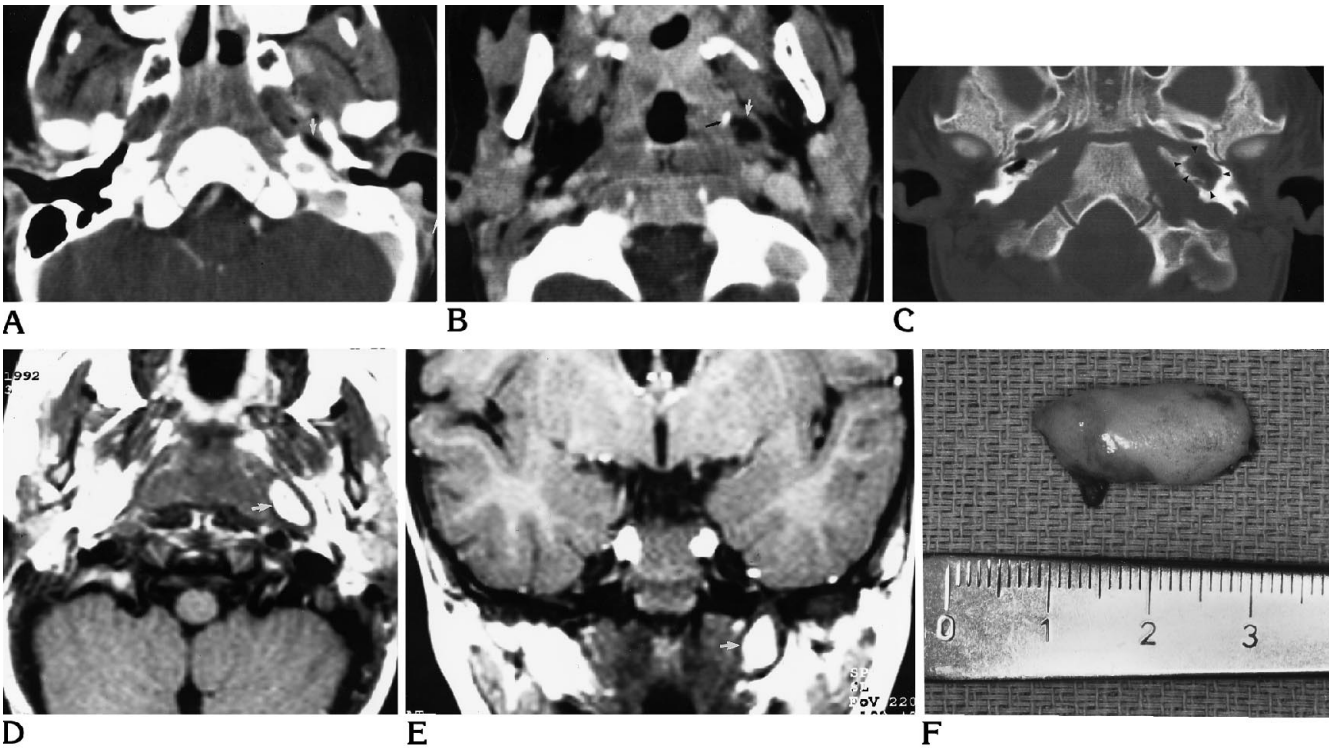

Fig 2. Case 2.

A and *B*, Contrast-enhanced axial CT scan shows a low-attenuation mass (*white arrows*) that extends along the left eustachian tube canal from the middle ear cavity to the left parapharyngeal space. A high-density structure (*black arrow*) inside the mass corresponded histologically to bone formation.

C, Bone algorithm axial CT scan demonstrates expansion of the bony origin of the eustachian tube (*black arrowheads*).

D and *E*, Axial and coronal T1-weighted MR images show the high-intensity lesion (*white arrows*) along the left eustachian tube.

F, Gross specimen of the resected mass shows a finely granular white surface and pedicle attachment.

revealed a fatty-attenuation mass with a thin enhancing capsule expanding the bony portion of the canal and extending down the cartilaginous portion to the left parapharyngeal space. A small, high-density structure near the base of the mass corresponded histologically to bone formation. The mass was in contact with the left carotid artery, but did not encase it. MR better demonstrated the relationship of the mass with the carotid artery beneath the temporal bone. The signal intensity of the mass followed the intensity of fat on all imaging sequences. No intracranial extension or involvement of the inner ear was present (Fig 2).

Through a postauricular approach, a large, firm mass was revealed in the protympanic space. Complete removal was achieved through the tympanotomy in combination with simultaneous flexible fiberoptic nasopharyngoscopy, open exploration of the neck, and exposure of the parapharyngeal space. The tumor was removed easily from the middle ear, but was adherent to the eustachian tube, giving the impression that it originated from the tubal canal.

Pathologic examination showed a polypoid mass with a pedicle attachment. The mass was covered by skin with numerous sweat glands and occasional immature hair follicles in the dermal tissue. The center consisted predominantly of mature fibroadipose tissue. A small area of bone formation was present near the base of the mass.

Discussion

Congenital dermoids are benign developmental anomalies rather than true neoplasms (6). They originate during early embryogenesis and are composed of a disorganized conglomeration of mesodermal and ectodermal derivatives (7). They are uncommon in the head and neck, comprising approximately 7% of all dermoids. More than 50% of these are found in the periorbital region, 25% are located in the oral cavity, and 13% occur in the nasal cavity (8). Twenty-four cases of dermoids of the temporal bone have been reported in the otolaryngologic literature and are summarized in a review article by Vrabec and Schwaber (1). Multiple sites of involvement within the temporal bone have been described, including the middle ear cavity,

the mastoid air cells, and the petrous apex, and 10 were noted as originating specifically from the eustachian tube (1–5).

The most popular hypothesis, explaining the histogenesis of dermoids, suggests that the lesions originate from inclusion errors at the site of contact between the endodermal lining of the first pharyngeal pouch and the ectodermal surface of the first branchial cleft (2, 7, 9, 10). The tympanic cavity and the eustachian tube form from an outward expansion of the endoderm-lined first (and perhaps the second) pharyngeal pouch. The blind distal end of this pouch expands to form the tympanic cavity. The proximal portion of the pouch undergoes a constriction and lengthens to become the eustachian tube. Around the fourth week of gestation, the endodermal lining of the first pharyngeal pouch comes in contact with the overlying ectoderm of the first branchial groove, which later deepens to form the external auditory meatus (Fig 3). This contact between endoderm and ectoderm is soon lost by interposition of mesoderm from which the auditory ossicles develop (11, 12). An inclusion of ectoderm into the endodermal lining during this short period of contact between the two layers is believed to be the underlying mechanism for the development of dermoids in this region.

Some confusion exists in the literature regarding the terminology used to describe these masses. *Dermoid cyst*, *teratoid tumor*, *hamartoma*, and *hairy polyp* have all been used to describe the same lesion. According to the histologic classification for teratomas in the head and neck region most commonly used today, the term *dermoid* as it was proposed by Arnold in 1870 is most appropriate (13). This implies an origin from epidermal and mesodermal elements, which differentiates them histologically from teratomas composed of elements from all three germinal layers and from cholesteatomas, which are only of ectodermal origin. Grossly, dermoids are usually polypoid, pedunculated, and rarely sessile masses. They are grayish-white or pink in color, and covered by skin, often including hair, hence their alternative name, "hairy polyp." Microscopically, the surface layer consists of stratified squamous epithelium that contains epidermal appendages. The stroma is primarily fibrofatty material, but may also contain smooth and striated muscle, cartilage, bone, minor salivary glands, nerves, and lymph nodes (14).

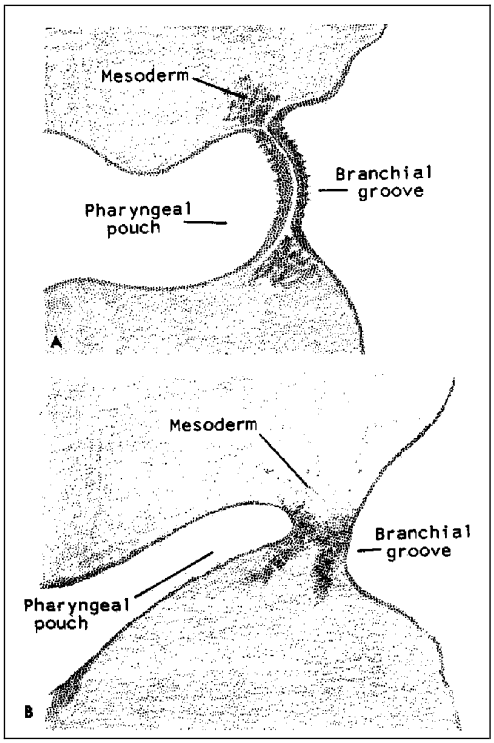

Fig 3. Pathogenesis of eustachian tube dermoids (from Williams [12]).

A, Around the fourth to fifth weeks of gestation, the endodermal lining of the first pharyngeal pouch (from which the tympanic cavity and the auditory tube form) comes in contact with the overlying ectoderm of the first branchial groove (which later deepens to form the external auditory meatus). An inclusion of ectoderm into the endodermal lining during this short period of contact between the two layers is the underlying mechanism for the development of dermoids in this region.

B, By the sixth week of gestation this contact between endoderm and ectoderm is lost by interposition of mesoderm from which the auditory ossicles develop.

The typical clinical presentation of dermoids involving the eustachian tube consists of chronic ear drainage with recurrent episodes of otitis media (8, 9). This is caused by the obstruction of the eustachian tube by the dermoid. Negative pressure is created within the tympanic cavity and the mastoid air cells, associated with low oxygen and/or high carbon dioxide tension. Such abnormal tension produces alterations of the middle ear mucosa with an increase of the mucus-secreting cells (15). Tympanic membrane perforation and inflammatory polyps or granulation tissue may be accompanied findings (4, 5). Upper airway obstruction as a result of extension of the dermoid into the nasopharynx is a rare complication (5). Although the number of reported cases is limited, a marked predominance in female subjects

is noted, with a ratio of 6 to 1 (female-to-male) existing for all dermoids of the head and neck (4, 8). The age at the time of presentation is usually during early infancy, with 5 of the 10 reported cases being before the age of 12 months, and only 2 cases reported after the age of 10 years.

In all previous reports, the diagnosis was made at the time of surgical exploration because of the characteristic macroscopic appearance of the lesion. A lack of adequate preoperative imaging may have contributed in several cases to an inadequate initial surgical approach, necessitating a second, more radical surgery. Although the definitive diagnosis is histologic, CT and MR may predict this lesion based on the imaging characteristics, anatomic location, and involvement of the eustachian tube. The radiographic findings of eustachian tube dermoids are not well described. Both CT and MR demonstrate involvement of the tubal canal and the extension to the middle ear and the parapharyngeal space. The characteristic histologic consistency of the mass is equally well demonstrated by both imaging modalities. CT allows better evaluation of the bone changes with expansion of the osseous portion of the eustachian canal and occasional erosion of the petrous bone. It is also superior in demonstrating involvement of the ossicular chain within the middle ear. Soft tissue algorithms reveal the fatty content of the mass, suggesting the correct diagnosis. Typical MR appearance consists of a well-defined, nonenhancing mass, surrounded by a smooth capsule (corresponding to the skin covering), and a relatively homogeneous matrix that follows the signal intensity of fat. The presence of septations reflects the presence of other mesodermal derivatives within the mass. The anatomic relationship of the lesion with the internal carotid artery beneath the temporal bone is better demonstrated by MR to assist in selection of the appropriate surgical approach. Associated inflammatory changes, with opacification of the middle ear and mastoid air cells, may create some confusion regarding the exact extent of the lesion to these areas. Imaging is of further importance to rule out any intracranial extension, and extension to the parapharyngeal space.

Differential considerations of pathologic conditions involving the eustachian tube are usually limited to processes of inflammatory and/or allergic nature. A careful search of the literature reveals only a few records concerning neoplastic or malformative processes involving the eustachian tube. These include temporal lobe meningocele herniating into the eustachian tube (16), meningiomas (17), and lipoma (18). A few cases also have been reported of processes involving the eustachian tube by direct extension from the adjacent structures, including plexiform neurofibroma (19) and nasopharyngeal teratoma (20). Teratomas and dermoids originating in other areas of the temporal bone can also extend to involve the eustachian tube (1). In most of the previously reported cases, the initial consideration, based on the clinical presentation and age of the patients, was of a congenital cholesteatoma. This diagnosis can be easily excluded by the typical CT and MR features of the mass.

The recommended treatment for these lesions is complete surgical excision (4, 5). Close anatomic relationships of the eustachian tube with vital structures, such as the internal carotid artery running medially and the middle meningeal artery laterally, impose some surgical risk.

The prognosis of all dermoids in the head and neck region is very favorable (13). We know of no reports of malignant degeneration or intracranial extension in the literature. These lesions have a limited growth potential, and when complete surgical removal is achieved they do not recur.

In summary, dermoids, although uncommon, need to be included in the differential diagnosis of middle ear lesions. Clinical presentation consists of recurrent otitis media and chronic otorrhea, refractory to treatment with antimicrobial agents. A well-defined fatty mass on both CT and MR, involving the eustachian tube, in a young child indicates the correct diagnosis. CT allows better evaluation of the bone architecture, whereas MR better demonstrates the relationship of the mass with the carotid artery beneath the temporal bone. The origin and extension of the mass are equally well outlined by both imaging modalities, facilitating the appropriate surgical approach and complete removal.

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