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Case report

Middle ear adenoma with neuroendocrine differentiation (MEA-ND) in the pediatric population☆

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ABSTRACT

Middle ear adenoma with neuroendocrine differentiation is an uncommon tumor of the tympanic cavity. The terminology of this entity has suffered due to a poor understanding of the differentiation of these neoplasms, and has included both "adenoma" as well as "carcinoid tumor." Immunohistochemical techniques have helped clarify that these tumors all share elements of neuroendocrine differentiation. Occurrences of this tumor in the pediatric population are extremely rare. In the world literature, only three pediatric cases of middle ear adenoma with neuroendocrine differentiation have been described. We report the youngest case to date of middle ear adenoma with neuroendocrine differentiation, in a 13-year-old boy. We discuss the differential diagnosis, surgical management, and pathology of this entity.

1. Introduction

Middle ear adenoma with neuroendocrine differentiation (MEA-ND) is a rare tumor that was first reported in 1980. It can be difficult to classify histologically as middle ear adenomas may show purely epithelial (adenoma), purely endocrine (carcinoid) or hybrid differentiation [1]. In spite of this variability, some authors suggest that all middle ear adenomas have elements of neuroendocrine differentiation. Support for this viewpoint comes from immunohistochemical studies with neuroendocrine-specific markers. Torske and Thompson, for example, reviewed 48 middle ear adenomas and demonstrated chromogranin positivity in all, implying some neuroendocrine differentiation despite indistinguishable morphology on routine stains [2].

Historically, MEA-ND has been referred to as a carcinoid tumor, reflecting its histologic similarity to intestinal carcinoids, which are classified by site of origin as foregut, midgut or hindgut lesions [3–5]. Middle ear carcinoids are associated with the foregut tumors, which generally produce only small amounts of peptide hormones. As a consequence, the typical carcinoid syndrome of cramping, flushing and diarrhea is absent in MEA-ND.

MEA-ND was considered an indolent lesion until 1999 when Mooney et al. [6] reported the first case with metastases. In this paper, we present the youngest case to date of MEA-ND in a 13-year-old boy. We discuss the surgical management of this patient, including findings at the original surgery and on re-exploration one year later. We also review the pathology and differential diagnosis of this tumor.

2. Case report

A 13-year-old boy followed for chronic otitis media with effusion presented with a left tympanic membrane perforation. His past medical history was significant for ventilation tube placement on several occasions and for recurrent tonsillitis necessitating tonsillectomy at age 11 years.

Examination of the left tympanic membrane was remarkable for a perforation in the attic region and a posterior-superior quadrant white mass. The right tympanic membrane examination was significant only for myringosclerosis. On audiometric testing, hearing in the right ear was normal, while a 20 dB air-bone gap and a speech reception threshold of 20 dB were documented in the left ear. Speech discrimination was 100% bilaterally. The patient denied otorrhea, tinnitus and aural fullness. A computerized tomography (CT) scan of the temporal bone was obtained (Fig. 1).

A middle ear exploration with fat graft myringoplasty was performed and at surgery, a mass was biopsied and note to be moderately vascularized. Routine hematoxylin and eosin sections

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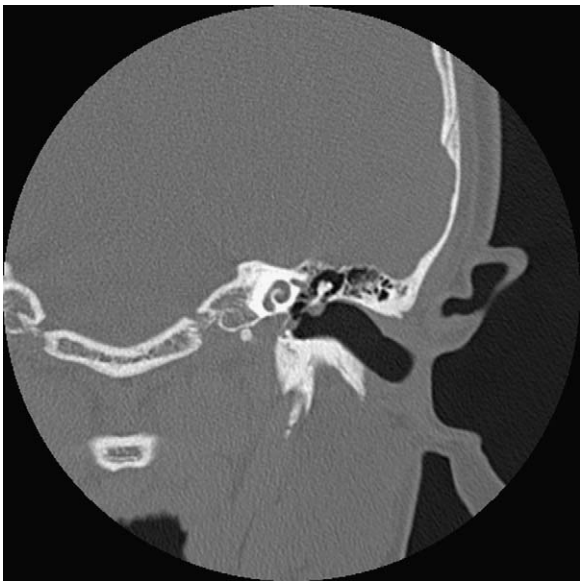

Fig. 1. Coronal CT of the left temporal bone showing opacification in the region of the ossicular mass and the tympanic membrane.

(Fig. 2) and immunohistochemical stains were performed. The latter were positive for S-100, synaptophysin (Fig. 3), pan-keratin antibody (Fig. 4), and chromogranin staining. A neuron-specific enolase stain was negative.

Four weeks later, a definitive resection was completed via tympanotomy through a postauricular approach. The mass abutted but was not attached to the manubrium of the malleus and was easily mobilized over the prominence of the facial nerve above the oval window. The overlying tympanic membrane was excised to improve exposure.

The mass encased the long process of the incus and the attic was visualized by removing the scutum. A small portion of the mass in the facial recess was mobilized using the facial recess excavator. The malleus head and entire incus were removed with the bulk of the mass, with residual remnants then removed piecemeal. Thick silastic sheeting was placed in the middle ear and the ear was closed.

One year later, a second middle ear exploration was performed. Recurrence was suspected based on a preoperative CT of the temporal bones, which showed opacification in the attic region. This suspicion was confirmed at surgery and an intact canal wall tympanomastoidectomy with a facial recess approach to the middle ear was completed to remove the tumor. Silastic sheeting was again placed in the middle ear. Due to the recurrence of the disease, ossicular reconstruction was deferred for two years at which time a CT scan showed no recurrence (Fig. 5).

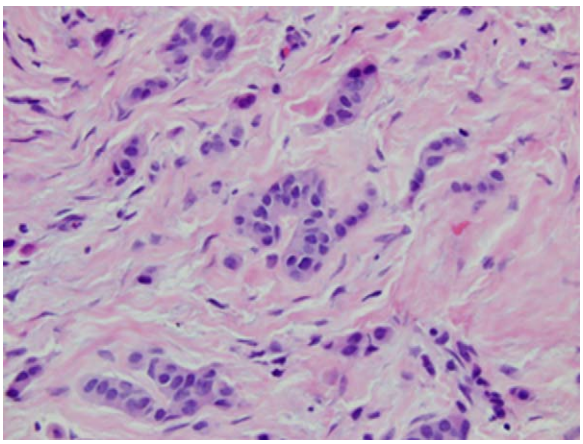

Fig. 2. Microscopic examination revealed epithelial cells having round–oval nuclei with fine chromatin and small–medium size eosinophilic cytoplasm within fibrous stroma. Scattered lymphocytes and mast cells were present (H&E, 600×).

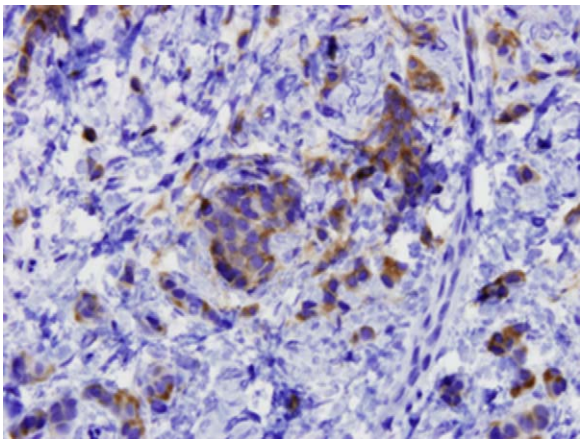

Fig. 3. Synaptophysin antibody stain is markedly positive (600×).

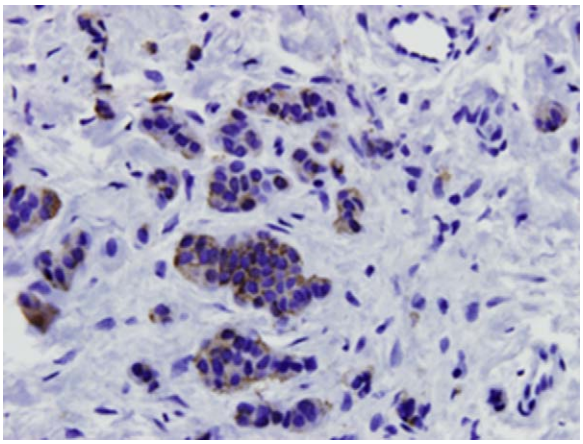

Fig. 4. Pan-keratin antibody stain is markedly positive (600×).

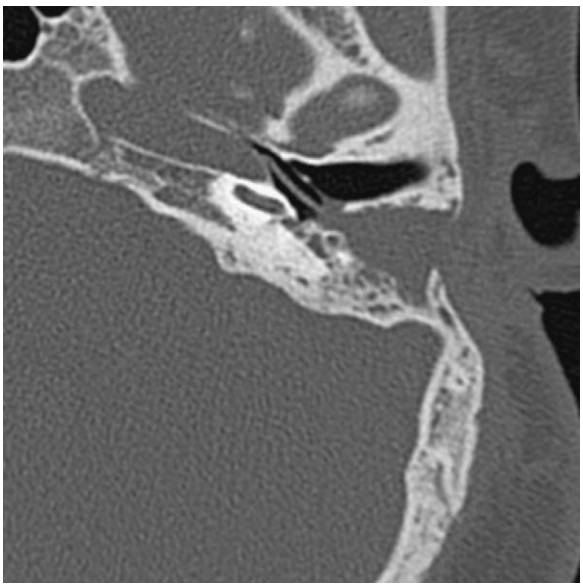

Fig. 5. 2-Year postoperative axial CT of the left temporal bone reveals silastic sheeting in the middle ear space without evidence of recurrence.