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Melanotic Neuroectodermal Tumor Presenting as Endolymphatic Sac Tumor

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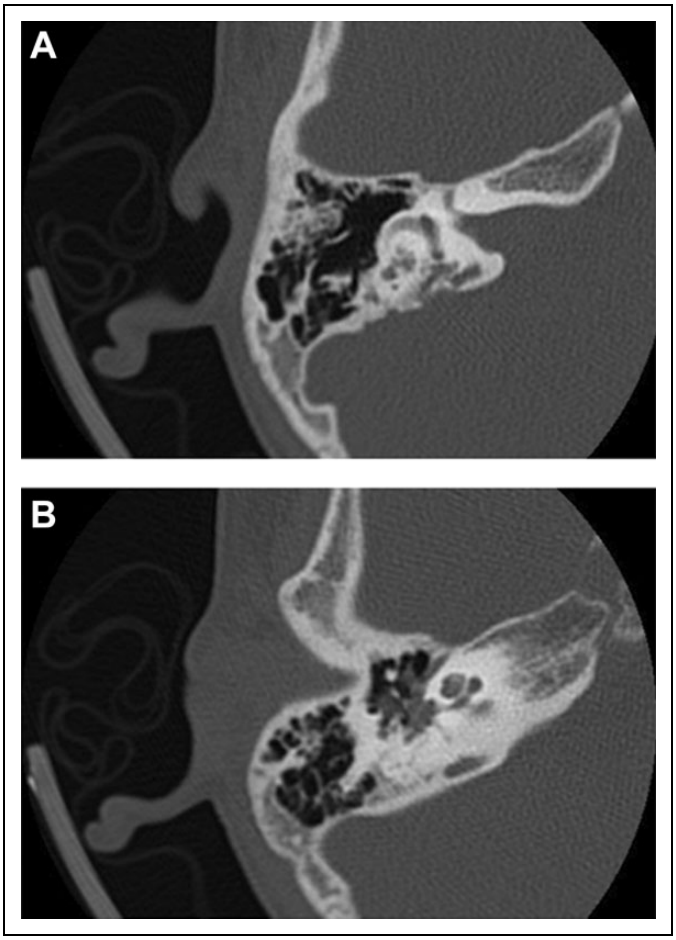

Figure 1. Computed tomography scan of the right temporal bone without contrast shows internal auditory canal widened to 7 mm, indicating an expansile mass with erosion along the posterior petrous bone (A) and the cochlea (B).

Melanotic neuroectodermal tumors (MNTs) are rare neoplasms of neural crest cell origin that usually arise during the first year of life with very few reported cases arising in ages 4 to 69.[1] These tumors most commonly present in the maxilla but are also seen in the skull and mandible and there have been cases reported in unusual regions such as the testes and peripheral bones.[1,2] Here, we report a very rare manifestation of pediatric MNTs presenting as an endolymphatic sac tumor.

A 7-year-old boy was referred to our neurotology clinic for evaluation of right-sided hearing loss, revealed during hearing screening at school. His parents noticed that he also had recurrent attacks of headache and dizziness, episodes of transient vision blurriness, and intermittent epistaxis. Microscopic ear examination revealed a small, nonpulsatile, pink middle ear mass adherent to the tympanic membrane. Audiology testing showed a right-sided severe to profound sensorineural hearing loss. He had a normal past medical and developmental history and his family history was negative for syndromic diseases. Computed tomography (CT) scan of the temporal bone showed widened right internal auditory canal and opacification of the middle ear and mastoid as well as erosion along the petrous bone adjacent to the endolymphatic sac (Figure 1). Magnetic resonance imaging (MRI) scans revealed a mass centered in the endolymphatic sac region with involvement of the internal auditory canal, cochlea and vestibular structures, and the right middle ear (Figure 2).

The patient underwent a transotic approach for the resection of a susceptible endolymphatic sac tumor. Intraoperatively, the tumor appeared melanotic (Figure 3) and had spread to the vestibule and filled the posterior and superior canals as well as cochlea and part of the internal auditory canal. Microscopically the tumor showed foci of alveolar growth pattern in the background of fibrous stroma at low magnification and demonstrated a biphasic tumor with small

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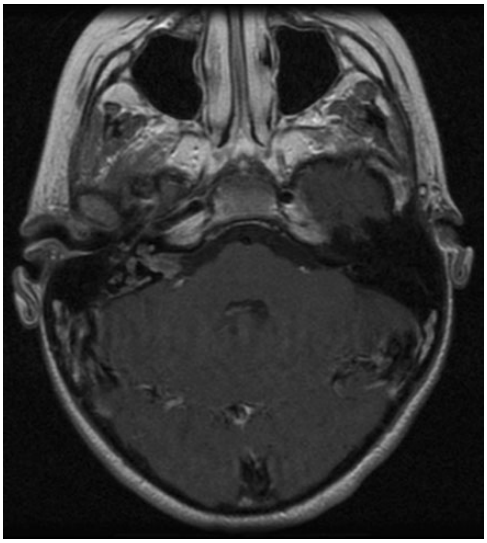

Figure 2. Axial T_1-weighted postgadolinium MRI showing tumor in the endolymphatic sac area with spread into the vestibule, cochlea, and internal auditory canal. MRI indicates magnetic resonance imaging.

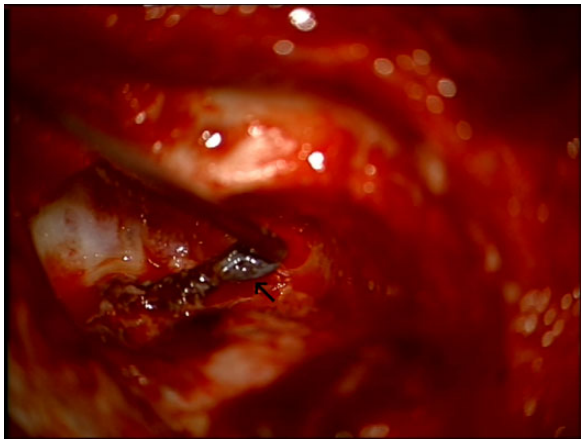

Figure 3. Intraoperative melanotic appearance of the tumor (arrow).

neuroblastic cells and large melanin-containing cells at high magnification (Figure 4). Final pathological examination revealed an MNT with extension into the bone. Postsurgical CT and MRI scans at 3 months showed no evidence of recurrence. Patient's recovery has been uneventful with no further complications 1 year following the surgery.

Melanotic neuroectodermal tumors are rare neoplasms usually presenting in the first year of life with no gender predilection.[1] These tumors have had different names in the past including melanotic progonoma, melanotic adamantinoma, retinal anlage tumor, and pigmented congenital epulis due to the uncertainty of the tumors' origin. Melanotic neuroectodermal tumors are typically benign, although malignant transformation can rarely occur.[1] These tumors generally present in the craniofacial region and, despite their benign nature, can display rapid growth causing destruction of surrounding structures with the need for possible reconstruction at the site of resection.[3] Surgical resection of the tumor is the standard of care and is mostly curative, but these tumors do have an overall recurrence rate of 15% to 27%.[3,4] Although these recurrence rates are reported for a younger age range, this relatively high recurrence rate warrants the need for long-term neuroimaging follow-ups in these patients.

Endolymphatic sac tumors can easily be mistaken for several different malignancies upon clinical presentation because of their location and rarity of the tumor. These tumors are locally aggressive and have been diagnosed over a wide range of age from 15 to 77 years old. In our patient, the location of the lesion, age at presentation, and accompanying sensorineural hearing loss all contributed to the initial clinical diagnosis of an endolymphatic sac tumor. Final diagnosis of MNTs has to be approved pathologically using staining for markers, such as synaptophysin, cytokeratin, and HMB-45.[5] This case illustration highlights the importance of considering MNT in the differential diagnosis list of the craniofacial tumors despite unusual location of the tumor and age range of presentation.

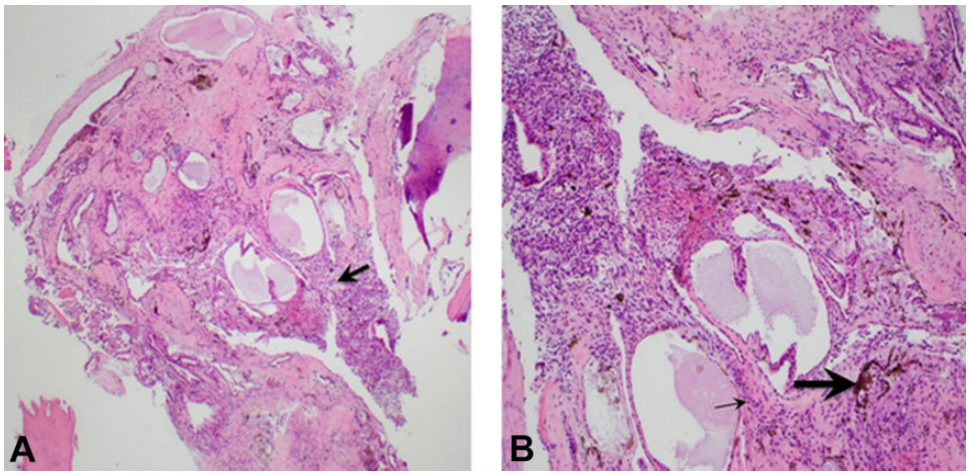

Figure 4. Low magnification displays alveolar growth pattern in background of fibrous stroma (arrow; A). High magnification demonstrates biphasic tumor with small neuroblastic (small arrow) and large melanin-containing cells (large arrow; B).

Authors' Note

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Declaration of Conflicting Interests

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