Glial Choristoma of the Temporal Bone in a 7-Month-Old Infant

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Glial choristomas, also known as glial heterotopias, are rare congenital lesions composed of neuroglial tissue connected to the central nervous system. Their origin is speculated to derive from abnormal closure of the neural tube vs sequestration of an acquired encephalocele. These lesions have been documented in the more common perinasal region in the form of nasal glioma but have also been identified in the scalp, oropharynx, nasopharynx, tongue, lip, orbit, chest wall, and lung.1 Scalp lesions are predominantly found in the midline and frequently present with overlying alopecia and a surrounding area of long, coarse hair noted as a “hair collar” sign.2 The middle ear and mastoid regions are especially rare locations for heterotopic glial tissue and have been thought to derive from acquired encephaloceles.3 Nearly all published cases of middle ear and mastoid region heterotopic glial tissue occur in adults. These lesions tend to present later in life in adults with complaints of hearing loss and/or otitis media.4 This case reported herein is unique in that it represents a lateral congenital temporal scalp lesion with extension into the mastoid portion of the temporal bone in an infant.

Report of a Case

A 15-day-old white female infant was referred with a left postauricular soft-tissue mass present since birth. The lesion measured approximately 3 cm in the postauricular scalp and was slightly raised and noncompressible. There was no redness, alopecia, overlying capillary staining, or hair collar sign present. There was no bluish hue or other indication of hypervascularity of the lesion. The skin overlying the lesion appeared uninvolved with hair growth consistent with the adjacent normal regions. She had passed her newborn hearing screen and was otherwise developmentally healthy with no anatomic or other abnormal condition (Figure 1).

We elected to follow the mass conservatively, given the patient’s young age. The mass slowly enlarged and began to displace the left auricle anteriorly and inferiorly. A computed tomographic (CT) scan of the temporal bone obtained at 2.5 months of age demonstrated a soft-tissue and fatty mass posterior to the left ear involving the base of the pinna with an area of conical invagination into the outer cortex of the mastoid.
bone. There was no bony destruction of the left mastoid bone. Some focal soft-tissue penetration was noted in the attic by the radiologist, but her middle and inner ear spaces appeared otherwise unremarkable. No intracranial extension was noted on the CT scan (Figure 2). A needle biopsy was performed under CT guidance, with findings of fibrovascular benign-appearing tissue.

As the child grew, the mass enlarged, causing increasing anterior displacement and deformity of the left pinna. Her mother reported increasing wax impaction, and the left ear canal became increasingly collapsed, resulting in difficulty examining the left tympanic membrane. Because of progressive ear deformity and obstruction, at age 7 months surgical excision was performed with the use of general anesthesia. The lesion was sharply excised. There was no clear capsule, and the overlying skin was fairly thin but otherwise unremarkable. Silverstein facial nerve monitoring was in place in case mastoidectomy was necessary. The lesion was elevated off of the temporal fascia, and centrally a stalk extended into the mastoid cavity, ending in a blind pouch. Periosteum was incised circumferentially around this area, and a sharp elevator was used to remove the residual stalk of tissue. A bony spicule was present in the mastoid extension, which was removed with the specimen. The bone was drilled down 1 to 2 mm with a small diamond burr. There was no visible communication into the intracranial vault or the middle ear space. Postauricular excess skin was trimmed. Left otoplasty with the Mustarde suture technique was performed to reposition the helix and antihelix that had been deformed by pressure from the mass.

In the gross and histopathologic assessment, the specimen consisted of a 3.8 × 2.0 × 1.1-cm, tan, soft-tissue mass and a 3.5 × 1.0-cm ellipse of unremarkable skin. Microscopic examination of the lesion demonstrated mature glial tissue in a background of vascularized fibroadipose tissue. Focal cystically dilated areas contained papillary formations resembling choroid plexus. The glial tissue was composed of a delicate fibrillary network with scattered astrocytes, occasional oligodendrocytes, and microglia. No definitive neurons were seen. Findings from immunohistochemical analysis showed that the glial tissue was positive for glial fibrillary acidic protein. The inner wall of the cystic spaces was lined by cuboidal cells resembling ependymal lining cells (Figure 3).

Postoperatively, she healed well with good position of the left pinna (Figure 4). She was reevaluated at 10 months postoperatively and had good appearance and symmetry of the pinna. There was no evidence of recurrent disease.
Discussion

Though previous reports of mastoid and middle ear glial choristoma suggest these lesions arise from an acquired encephalocele,3 this case truly represents a congenital glial choristoma of the lateral scalp involving the mastoid portion of the temporal bone in an otherwise healthy infant. Only one other published case of temporal bone region glial choristoma in an infant was found during this search of the English-language literature.5 This supports the theory that lateral heterotopic glial tissue of the middle ear and mastoid bone in infants may result from abnormal closure of the neural tube in utero. This is in contrast to most published cases of glial choristoma in the middle ear and mastoid bone, which present in adults with hearing loss.3,6-8 It is possible that the cases in adults represent a sequestered encephalocele instead of a congenital anomaly, suggesting there is a bimodal distribution of heterotopic glial tissue in the middle ear and mastoid. Cases presenting in infancy likely represent abnormal neural tube closure, whereas cases in adults may derive from prior encephalocele.

As this case did not demonstrate clinical signs suggestive of a neural tube deformity, such as the hair collar sign or overlying skin discoloration, the importance of preoperative imaging prior to operating on congenital scalp lesions is highlighted and should include lateral lesions as well as midline lesions. In this instance, preoperative biopsy did not suggest glial tissue, and thus needle biopsy cannot be used to definitively rule out heterotopic glial tissue. Congenital scalp masses must always be examined fully for intracranial extension, as in the case of encephalocele or meningocele, with heterotopic glial tissue always remaining in the differential diagnosis. Recommendations include long-term follow-up, although recurrence rates of heterotopic glial tissue have been shown to be very low despite poor encapsulation of the mass.9

References