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CASE- REPORT

Solitary Trichoepithelioma of the External Auditory Canal: A Rare Cause of Ear Pain in a Pediatric Patient

Lauren E. Melley, DO [1*] | Eli Bress, DO [2] | Megan Saltsgaver [3] | Judy Pascasio, MD [4] | Alexander Manteghi, DO [5]

1. Department of Otolaryngology -Head and Neck Surgery, Philadelphia College of Osteop -athic Medicine, 4190 City Line Avenue, Philadelphia, PA, USA 19131
2. Department of Otolaryngology -Head and Neck Surgery, Philadelphia College of Osteop -athic Medicine, 4190 City Line Avenue, Philadelphia, PA, USA 19131
3. University of Illinois College of Medicine at Rockford, 1601 Parkview Ave, Rockford, IL 61107
4. Department of Pathology and Laboratory Medicine, St. Christopher's Hospital for Children, Philadelphia, PA, USA
5. Division of Pediatric Otolaryn- gology, St. Christopher's Hospi -tal for Children, Philadelphia, PA, USA

Abstract

Trichoepitheliomas are benign hair follicle tumors that typically present as slow-growing sharply circumscribed, symmetrical, dome-shaped lesions. They can occur as a solitary lesion or as part of the autosomal dominant multiple familial form. Trichoepitheliomas are predominantly confined to the face with a symmetrical distribution along the nose, cheeks, eyelids, forehead, and preauricular area. We present the first pediatric case of a solitary trichoepithelioma in the external ear canal of 12-month-old girl presenting to the ENT office with otalgia and an EAC mass noticed by the patient's Mom 10 days prior. The patient underwent complete excision of a 1.5 cm cyst-like structure without rupture and an acellular graft was used to cover the defect. Histologic evaluation demonstrated numerous hair follicles, each surrounded by a well-defined perifollicular sheath consistent with a trichoepithelioma. Both clinically and histologically, trichoep - itheliomas are similar to basal cell carcinoma and have a low risk of malignant transformation. Excisional biopsy with complete resection of overlying skin is recommended and should be performed promptly to avoid delay in diagnosis.

Keywords: Trichoepithelioma, External auditory canal, Pediatric, Ear canal mass, Tumor, Benign

1 | INTRODUCTION

Trichoepitheliomas are benign hair follicle umtors originating from proliferation of epithelial-mesenchymal origin at the follicle bulb (1),(2). They typically present as sharply circumscribed, symmetrical, dome-shaped lesions on the nose,upper lips, and cheeks (3). While their exact prevalence is is unknown, one study found that trichoepitheliomas represented 0.02% of their total surgical pathology case load over a 19-year period, thus emphasizing their rarity as a diagnosis(4). Both clinically and histologically, richoepithetliomas are similar to basal cell carcinoma and have a low risk of malignant transformation(5). In rare cases, they have been reported to undergo transformation into malignant neoplasms, such as trichoblastic carcinoma or basal

cell carcinoma (BCC) (5),(6),(7),(8). Trichoepitheliomas can occur as a solitary lesion or as part of the autosomal dominant multiple familial form. The familial form, also known as Epithelioma Adenoides Cysticum, is characterized by numerous nodules and papules, which appear around puberty (9).They are predominantly confined to the face with a symmetriccal distribution along the nose, cheeks, eyelids, forehead, and preauricular area but occasionally spread to the scalp, neck, or upper trunk. In rare cases of multiple familial trichoepithelioma, the tumors have also spread to the EAC on both sides, leading to conductive hearing loss(10) There is only one reported adult case of a solitary nonfamilial trichoepithelioma presenting in an EAC, which occurred in a 19-year-old man (11). To our knowledge, we present the first pediatric case of a solitary trichoepithelioma in an EAC. Solitary trichoepitheliomas are typically slow-growing over years and asymptomatic. Contrarily, our 12-month-old patient presented with otalgia and an ear canal mass that appeared rather suddenly, although it is uncertain how long the mass was present before the patient's mother noticed its presence.

2 | CASE PRESENTATION

A 12-month-old girl with no significant past medical history presented to the ENT clinic after her mother noticed a lump in her left external auditory canal (EAC) that appeared 10 days prior to presentation. The mass had not changed since first appearance and was painful without drainage.The child demonstrated ear tugging and became tearful when her mother palpated the mass. The parents reported using antibio -tic ear drops without improvement. Her mother denied any history of trauma, recent upper respiratory infections (URIs), nor ear infections. There was no family history of similar lesions or hearing loss. Physical exam revealed a 1x1 cm flesh-colored cyst, occupying about 75% of the circumfere -nce of the left cartilaginous EAC, abutting the bony EAC junction. She was taken to the operating room for complete excision of a 1.5 cm cyst-like structure without rupture. The defect was not able to be closed primarily, and an acellular graft was used to cover the defect. Histologic evaluation demonstrated numer -ous hair follicles, each surrounded by a well-defined perifollicular sheath. Secondary follicles with abortive pilar differentiation and infundibular keratinization were abundant (Figure 1). Some of the follicles contained primitive sebaceous structures as seen in (Figure 2). There was also an area of adjacent granulation tissue. The aforementioned characteristics were most consistent with a diagnosis of a trichoepith -elioma. The acellular graft was removed two weeks later demonstrating a well-healed surgical site.

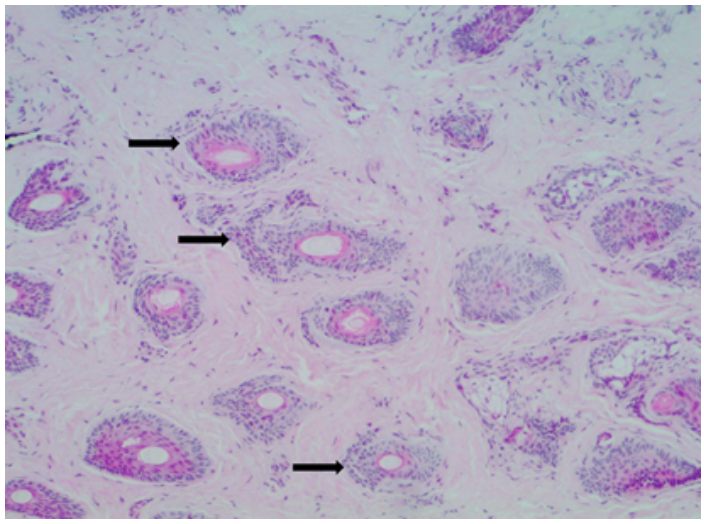

Figure 1. *Trichoepithelioma secondary follicles;100x magnification*

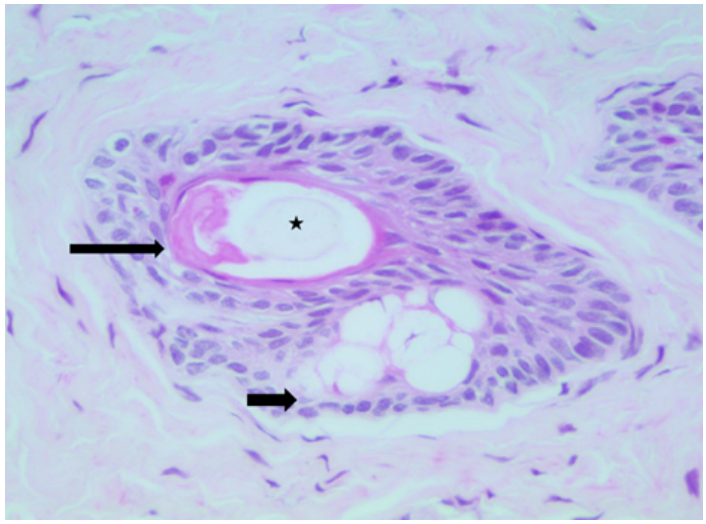

Figure 2. *Trichoepithelioma secondary follicle with hair (star), keratinization (long arrow), and primitive sebaceous structure (short arrow); 400x magnification.*

Supplementary information The online version of this article (https://doi.org/10.52845/JORR/2022/3.4.3) contains supplementary material, which is available to authorized users.

Corresponding Author: *Lauren E. Melley, DO, Department of Otolaryngology- Head and Neck Surgery Philadelphia College of Osteopathic Medicine 4190 City Line Avenue Philadelphia, PA 19131 (215)-871-6100*
Email: lm244130@pcom.edu

3 | DISCUSSION

Considering the rarity of trichoepitheliomas and their even more uncommon appearance in the EAC, this case report highlights the broad differential of EAC masses in the pediatric patient. When evaluating a pediatric patient with an ear canal mass, it is important to consider the extensive list of both benign and malignant diagnoses. More common conditions include otitis externa, osteomyelitis, benign bony tumors (i.e. exostoses, osteoma), sebaceous cyst, granuloma, cholesteatoma, papilloma, keratosis obturans, ceruminoma, and xanthogranuloma. Malignant lesions include carcinoma (i.e. basal cell, squamous cell, or adenocystic), neuroblastoma, and rhabdomyosarcoma.

4 | CONCLUSION

Many of the aforementioned conditions can be identified clinically without the need for extensive workup. However, in cases where involvement beyond the EAC is suspected, computed tomography (CT) of the temporal bones should be considered to evaluate for any associated bony destruction. For the pediatric patient in particular, the benefits of obtaining a CT for accurate early diagnosis or treatment planning should be weighed against the known long-term dangers associated with ionizing radiation. We did not subject our patient to CT imaging, but it is imperative for other physicians to make this decision based on their own clinical judgment. Excisional biopsy with complete resection of overlying skin is recommended. This should be done promptly to avoid recurrence and delay in diagnosis.

CONFLICTS OF INTEREST

The authors report no conflicts of interest.

Patient Consent

Verbal consent was obtained from the patient's parents for the publication of this manuscript.

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