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Case Report

# Acute Bacterial Meningitis and Petrous Apicitis in a Child with Aplasia Cutis Congenita: A Case Report

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Petrous apicitis and acute bacterial meningitis are uncommon in the present antibiotic era. The diagnosis of petrous apicitis is seldom considered unless there is cranial nerve palsy. A young child with aplasia cutis congenita presented with acute bacterial meningitis and an incidental opacified left mastoid in brain imaging. During the course, fever persisted, and high-resolution temporal bone imaging showed rapid progression to coalescent mastoiditis, petrous apicitis with erosions of tegmen tympani, and petrous apex. Other findings included bony dehiscences and thinning of left calvaria. Tympanomastoid exploration showed herniated brain and cerebrospinal fluid leak through tegmen tympani, which was closed with temporalis fascia graft. Herein, we report a rare presentation of petrous and tegmen erosion along with aplasia cutis congenita and discuss the challenges in diagnosis and management.

**KEYWORDS:** Petrous apicitis, cerebrospinal fluid leak, congenital cholesteatoma, Langerhans cell histiocytosis, skull lytic lesion

## INTRODUCTION

Petrous apicitis, the inflammation of the mucosa lining the petrous apex (PA) and acute bacterial meningitis (ABM) are severe intracranial complications of otitis media (OM) and acute mastoiditis.[1,2] With the introduction of the pneumococcus vaccine and early antibiotic therapy, both petrous apicitis and ABM are increasingly rare. In a series of 5.8 million emergency visits for OM and mastoiditis, Ren et al[2] report complications in 0.26%.[2] The prevalence of both petrous apicitis and ABM is 0.002% in this series.

The PA is susceptible to infection because of its extensive pneumatization, rich bone marrow, and proximity to the mastoid and middle ear. The Gradenigo's triad of suppurative OM, abducens nerve palsy, and trigeminal neuralgia is considered the hallmark of petrous apicitis, and the diagnosis is seldom considered unless there is cranial nerve palsy.[3] Petrous apicitis can have an insidious course. Also, in children, other PA lesions, including bone disorders and neoplasms, constitute the differential diagnosis.[4]

We present the first report of a child with petrous apicitis and aplasia cutis congenita (ACC). This article also discusses the differential diagnosis of pediatric PA lesions.

## CASE PRESENTATION

A 6-year-old previously well male child presented to the emergency with high-grade fever, headache, vomiting, and irritability of 3 days duration. There was no history of ear pain, discharge, convulsions, or neurologic deficit. On examination, the child was irritable with a temperature of 101°F and tachycardia. General examination showed 3 areas of circumscribed scarring alopecia suggestive of congenitally healed ACC on the left posterior scalp (Figure 1). Otoscopic examination was normal. Meningeal signs were evident on nervous system examination with no cranial nerve palsy or focal neurological deficit.

Blood investigations showed elevated leukocyte count (19 600 cells/mm$^3$) with neutrophilic predominance (93%) and elevated procalcitonin (72 ng/mL). Contrast-enhanced computed tomography (CT) of the brain showed opacification of left mastoid air cells (Figure 2). Left parietal and occipital bones showed thinning and irregular contours (Figure 3), corresponding to the overlying

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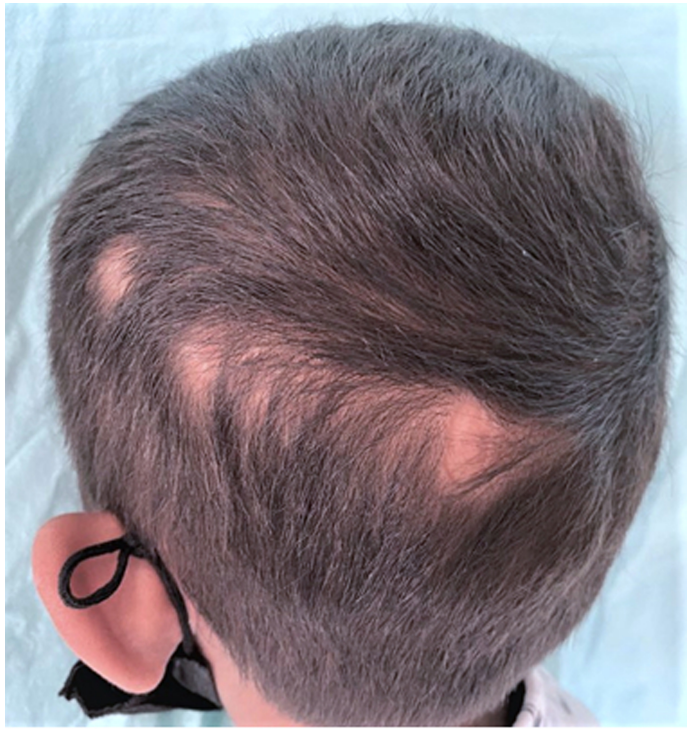

**Figure 1.** Circumscribed areas of scarring alopecia on the left posterior scalp above the plane of the ear.

skin lesions. The cerebrospinal fluid (CSF) analysis showed pleocytosis (500 cells/mm$^3$) with neutrophilic predominance (54%), low sugar (9 mg/dL), and elevated protein (346 mg/dL). The child was started on empirical intravenous ceftriaxone and vancomycin with diagnosis of ABM and acute mastoiditis. The CSF culture grew methicillin-sensitive *Staphylococcus aureus*.

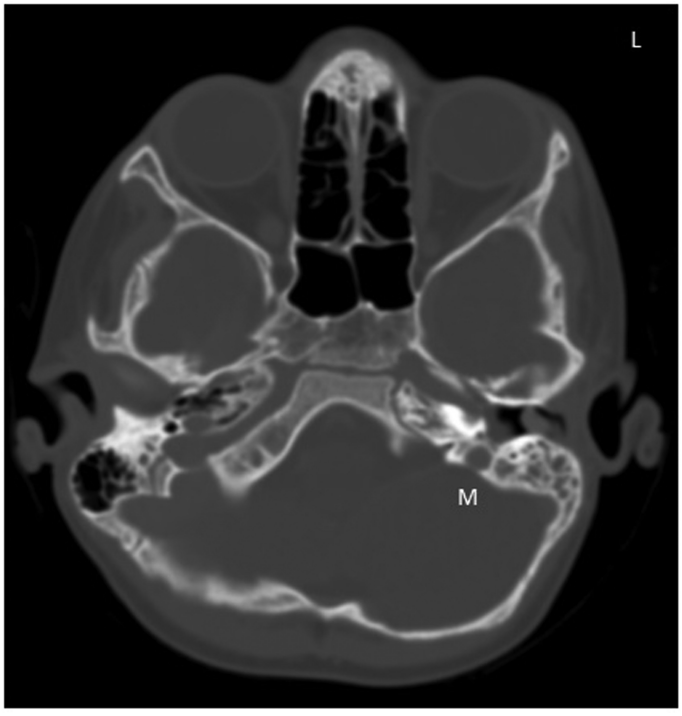


**Figure 2.** Contrast-enhanced computed tomography of the brain showing opacification of left (L) mastoid (M) air cells.

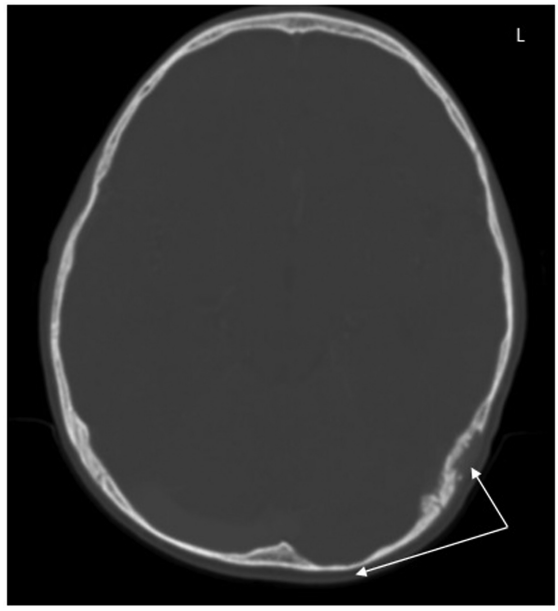


**Figure 3.** Contrast-enhanced computed tomography of the brain showing thinning and irregular contour (arrows) of the left (L) parietal and occipital bones.

The child continued to have fever even after 7 days of antibiotic therapy, though his general condition improved considerably. Repeat otoscopic examination showed bulging left tympanic membrane (TM). High-resolution computed tomography (HRCT) of the left temporal bone showed nonenhancing soft tissue density with air-fluid levels in the tympanic cavity (TC) and mastoid and PA suggestive of coalescent mastoiditis with petrous apicitis. There were erosions of tegmen tympani, PA (Figure 4), and bulging TM (Figure 5). There was dehiscence of superior aspect of semi-circular canal (Figure 6). Left tympanomastoid exploration was done under general anesthesia. On elevation of the tympanomeatal flap, pulsating clear CSF was

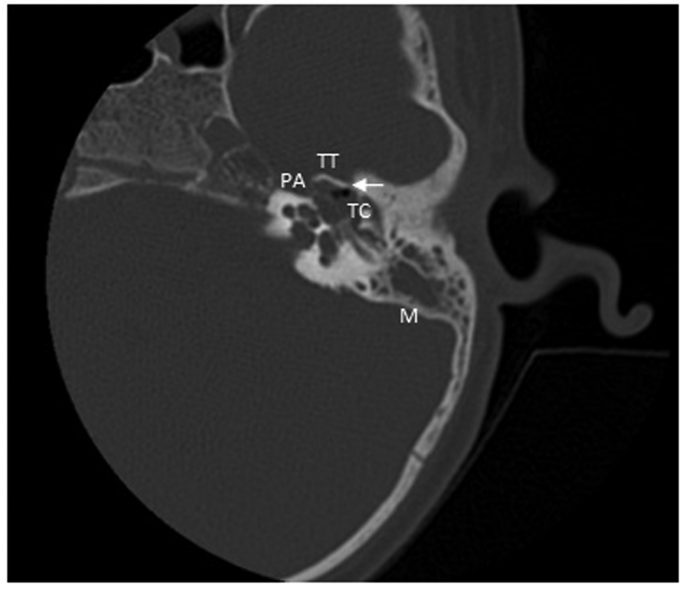


**Figure 4.** Preoperative high-resolution computed tomogram of the left temporal bone showing nonenhancing soft tissue density in the tympanic cavity (TC) with air-fluid level (arrow) and erosions of tegmen tympani (TT) and petrous apex (PA).

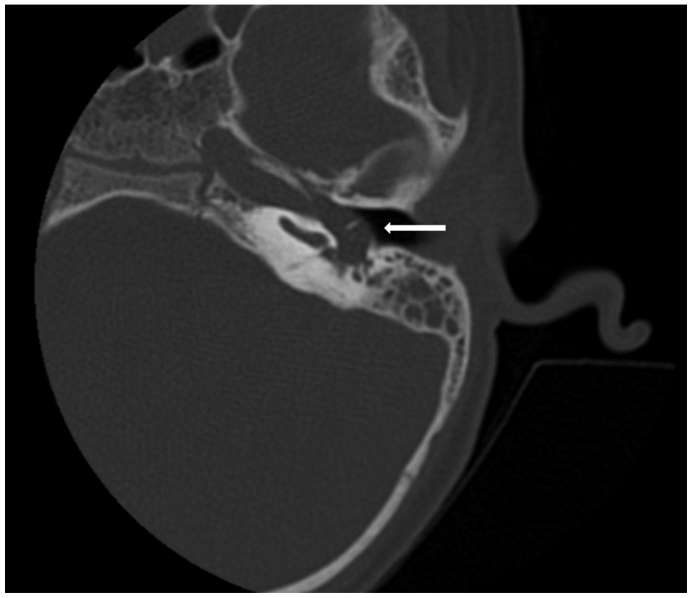

**Figure 5.** Preoperative high-resolution computer tomogram of the left temporal bone showing bulging tympanic membrane (arrow).

noted in the TC (Figure 7). After suctioning, herniated brain tissue was seen through bony defect in the tegmen tympani (Figure 8). The defect was sealed with temporalis fascia and surgicel (Figures 9 and 10), middle ear was packed with gelfoam (Figure 11), and the tympanomeatal flap was replaced (Figure 12). Cortical mastoidectomy showed a normal mastoid antrum. A biopsy was done, and a drain was placed in the mastoid cavity which was removed on postoperative day 2.

The CSF fluid analysis obtained during the procedure showed resolving meningitis. The CSF culture was sterile. The child was continued on antibiotics for 6 weeks. Histopathology of the mastoid biopsy showed bony trabeculae, fibro collagenous tissue, and chronic inflammation. A review on postoperative day 15 and day 30 showed

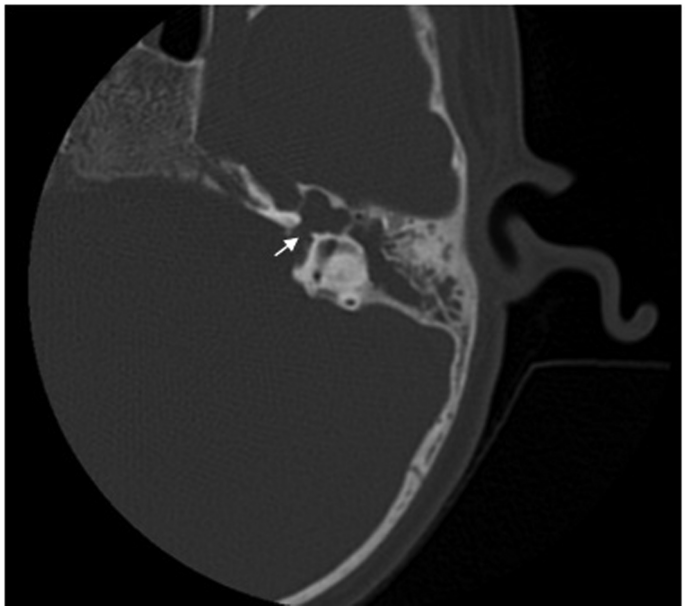

**Figure 6.** Preoperative high-resolution computer tomogram of the left temporal bone showing dehiscence in the superior semicircular canal (arrow).

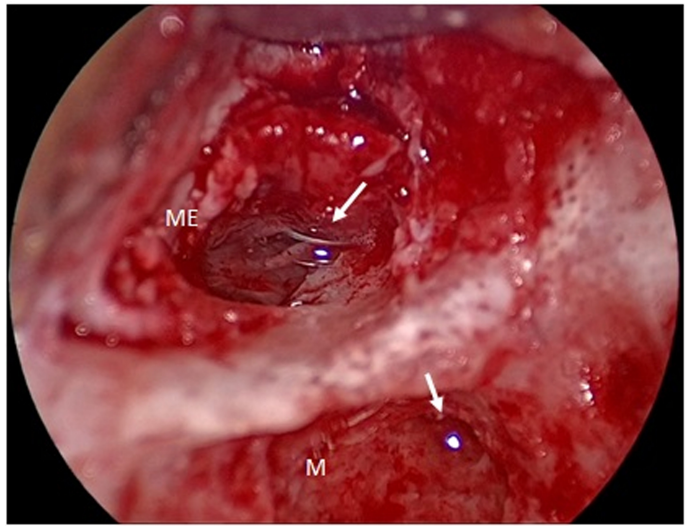


**Figure 7.** Intraoperative photograph showing clear watery discharge filling the middle ear and mastoid.

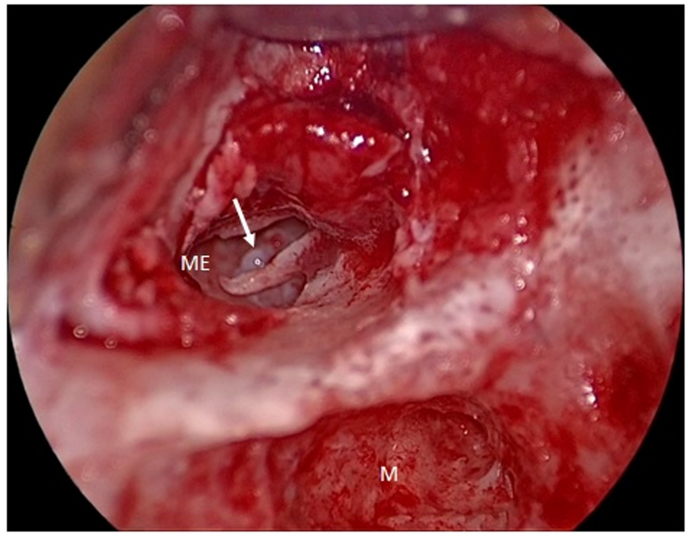


**Figure 8.** Intra-operative photograph showing herniated brain tissue filling the middle ear.

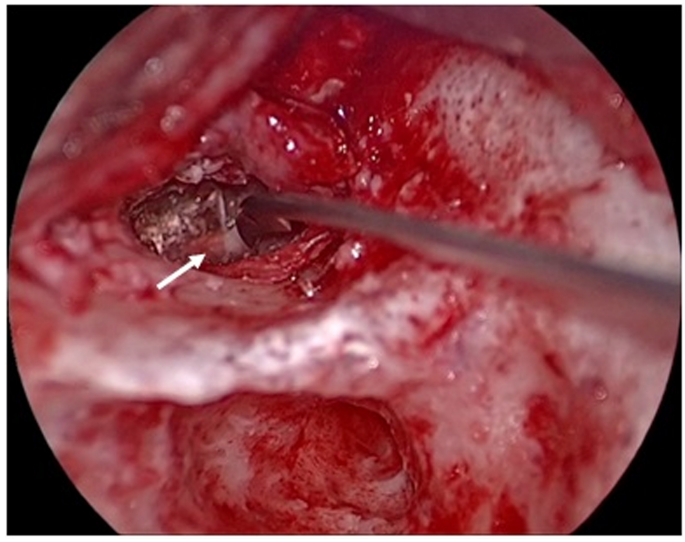

**Figure 9.** Intraoperative photograph showing sealing of the bony defect with temporalis fascia.

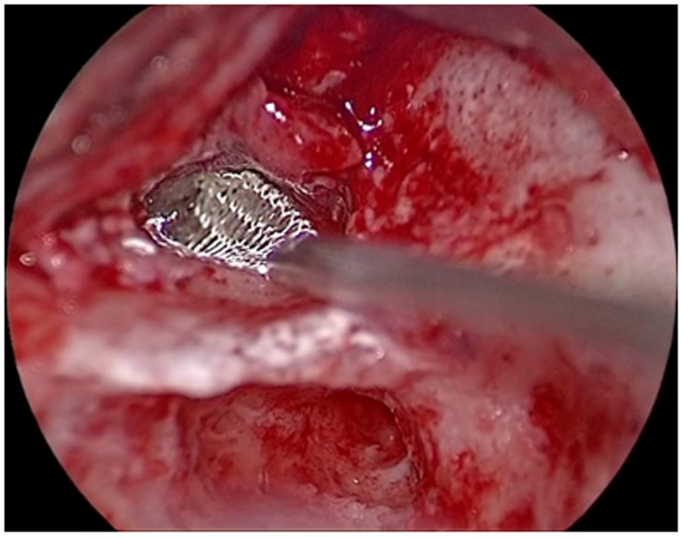

**Figure 10.** Intraoperative photograph showing sealing of the bony defect with surgicel.

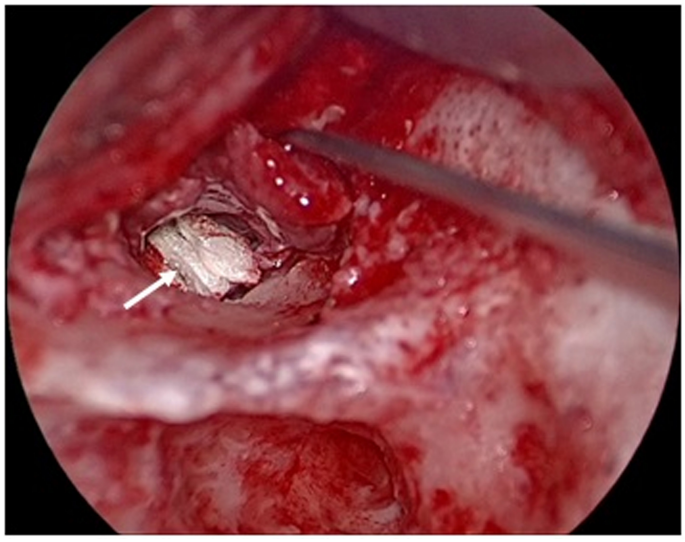

**Figure 11.** Intraoperative photograph showing gelfoam (arrow) packing.

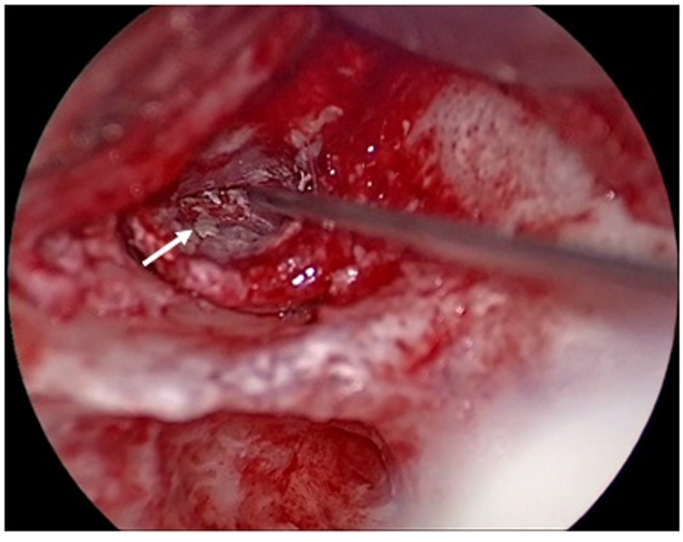

**Figure 12.** Intraoperative photograph showing replacement of the tympanomeatal flap.

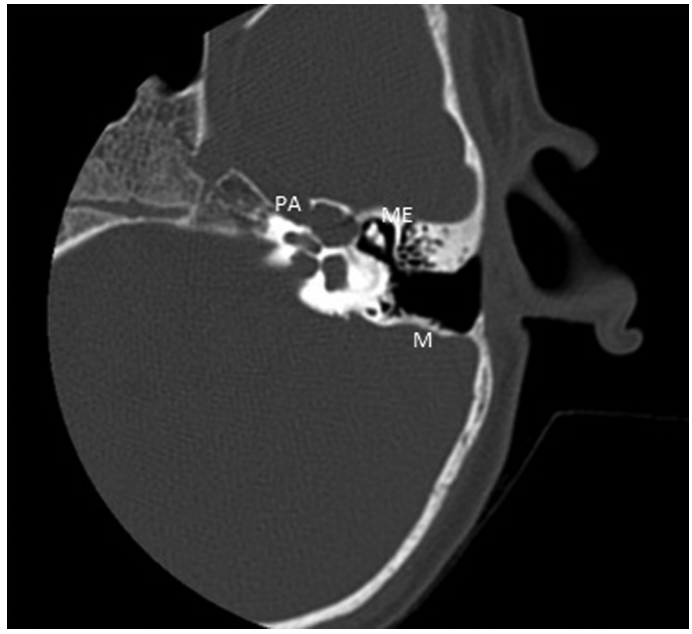


**Figure 13.** Postoperative high-resolution computer tomogram of the left temporal bone shows normal middle ear, soft tissue density in the petrous apex, with the mastoid cavity (arrow).

an intact tympanic membrane with a normal appearance. High-resolution computed tomography of the petrous temporal bone at the 6-month follow-up (Figure 13) showed a normal middle ear cavity. Pure tone and immittance audiometry at 6-month follow-up were normal.

Written informed consent was obtained from the parents of the child who participated in this study.

## DISCUSSION

The young child presented in this report had ABM with no previous history of OM or mastoiditis. Neuroimaging incidentally showed unilateral opacified mastoid, which did not corroborate with the otoscopic examination. The dilemma in the case management was due to the imaging showing a chronic disease process in the absence of ear findings. The subsequent HRCT temporal bone imaging showed rapid disease progression to coalescent mastoiditis and erosions in the tegmen tympani and PA despite antibiotic therapy.

Imaging plays a crucial role in PA lesions. Given the thinning and irregularity of the skull bone overlying the ACC in this child, the differential diagnosis included monostotic fibrous dysplasia (FD), Langerhans cell histiocytosis (LCH), and congenital cholesteatoma. The most common site of monostotic craniofacial FD is the zygomaticomaxillary complex.[5] Temporal bone involvement is seen in polyostotic FD.[4,5] The characteristic radiological appearance is an expansile ground-glass density due to the fibro-osseous tissue. The temporal bone is the site of pediatric LCH, with mastoid the most reported subsite and PA the least reported.[6,7] Ear discharge, ear pain, periauricular swelling, and scalp seborrhea prompt the diagnosis. Imaging shows a well-circumscribed soft tissue density with bony destruction. Rhabdomyosarcoma and Ewing sarcoma are other malignant neoplasms with soft tissue mass at the PA in the imaging.[4,7]

Congenital cholesteatoma usually presents with symptoms of hearing loss, facial paralysis, or tinnitus. Otoscopic examination shows a white mass behind an intact TM. The commonest site is the middle ear followed by PA and cerebellopontine angle. Computed tomogrpahy findings show subtle soft tissue densities in the middle ear with air-fluid level.[8,9] Magnetic resonance imaging helps to differentiate PA cholesteatoma from other lesions. Management of PA cholesteatoma is a surgical challenge as the PA is difficult to access and complications like CSF leak and facial nerve injury can occur during surgery.[10]

Intraoperatively, we observed a bony defect in the tegmen tympani and active CSF leak. The most common etiology of PA CSF leaks in pediatric population is a meningocele.[11] In this child, there were no signs of meningocele on imaging or during the surgery. Mastoid and petrous opacification were observed in the first brain imaging, and histopathology showed chronic inflammation suggestive of indolent petrous apicitis. Management includes transmastoid, middle cranial fossa approach (MFA), or combined approach (MFA and transmastoid) and closure.[11]

Aplasia cutis congenita is a rare congenital dermatologic condition predominantly affecting the scalp. Bessis et al[12] reviewed the neuroimaging of 56 patients with ACC. A cranial defect or hypoplasia was seen in 17 patients (30.4%), of which 16 had communication to the brain. Kuemmet et al[13] in a case series of 69 patients with scalp ACC demonstrated underlying skull abnormality in 10.1%. In this child, though there was thinning of the underlying skull bone, there was no defect. The acute presentation and progression to temporal bone erosion was likely caused by ABM. The ACC could have been an epiphenomenon.

## CONCLUSION

We conclude that PA lesions can present without cranial nerve palsy and ear findings. Temporal bone imaging is warranted in children presenting with ABM and incidental opacified mastoid with normal ear findings.

**Informed Consent:** Written informed consent for participation and publication of medical details was obtained from the patient's family.

**Peer-review:** Externally peer reviewed.

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**Declaration of Interests:** The authors declare that they have no competing interest.

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## REFERENCES

1. Lempinen L, Karppinen M, Pelkonen T, et al. Otitis media-associated bacterial meningitis in children in a low-income country. *Pediatr Infect Dis J*. 2019;38(8):791-797. [CrossRef]
2. Ren Y, Sethi RKV, Stankovic KM. Acute otitis media and associated complications in United States emergency departments. *Otol Neurotol*. 2018;39(8):1005-1011. [CrossRef]
3. Gadre AK, Chole RA. The changing face of petrous apicitis-a 40-year experience. *Laryngoscope*. 2018;128(1):195-201. [CrossRef]
4. Radhakrishnan R, Son HJ, Koch BL. Petrous apex lesions in the pediatric population. *Pediatr Radiol*. 2014;44(3):325-39; quiz 323. [CrossRef]
5. Valentini V, Cassoni A, Marianetti TM, Terenzi V, Fadda MT, Iannetti G. Craniomaxillofacial fibrous dysplasia: conservative treatment or radical surgery? A retrospective study on 68 patients. *Plast Reconstr Surg*. 2009;123(2):653-660. [CrossRef]
6. Zheng H, Xia Z, Cao W, et al. Pediatric Langerhans cell histiocytosis of the temporal bone: clinical and imaging studies of 27 cases. *World J Surg Oncol*. 2018;16(1):72. [CrossRef]
7. Majumder A, Wick CC, Collins R, Booth TN, Isaacson B, Kutz JW. Pediatric Langerhans cell histiocytosis of the lateral skull base. *Int J Pediatr Otorhinolaryngol*. 2017;99:135-140. [CrossRef]
8. Tada A, Inai R, Tanaka T, et al. The difference in congenital cholesteatoma CT findings based on the type of mass. *Diagn Interv Imaging*. 2016;97(1):65-69. [CrossRef]
9. Kim SH, Cho YS, Chu HS, Jang JY, Chung WH, Hong SH. Open-type congenital cholesteatoma: differential diagnosis for conductive hearing loss with a normal tympanic membrane. *Acta Otolaryngol*. 2012;132(6):618-623. [CrossRef]
10. Atlas MD, Moffat DA, Hardy DG. Petrous apex: diagnostic and treatment dilemmas. *Laryngoscope*. 1992;102(12 Pt 1):1363-1368. [CrossRef]
11. Ramadan O. Petrous Apex cerebrospinal fluid (CSF) leak: a review Article. *Otolaryngol Open J*. 2016;3(1):1-8. [CrossRef]
12. Bessis D, Bigorre M, Malissen N, et al. The scalp hair collar and tuft signs: A retrospective multicenter study of 78 patients with a systematic review of the literature. *J Am Acad Dermatol*. 2017;76(3):478-487. [CrossRef]
13. Kuemmet TJ, Miller JJ, Michalik D, Lew SM, Maheshwari M, Humphrey SR. Low risk of clinically important central nervous system dysraphism in a cohort study of 69 patients with isolated aplasia cutis congenita of the head. *Pediatr Dermatol*. 2020;37(3):455-460. [CrossRef]