

Incidental Finding of Mastoid Dermoid Cyst During Cochlear Implantation: A Case Report

Abstract

Dermoid tumors are unusual pouch-like benign growth that may be apparent at birth. Described in this report, dermoid cysts that occurred in the middle ear and mastoid bone which is quite rare. A 3 years old male presented with severe to profound sensorineural hearing loss bilaterally since birth. During cochlear prosthetic device implantation surgery, an incidental intraoperative cyst was found in the postauricular area around 5 cm behind the bony external auditory canal. In this paper, we demonstrate our management for mastoid dermoid cyst with respect to the clinical, radiological, histopathological, and surgical aspects.

Introduction

Dermoid cysts are non-neoplastic abnormal growth also known as teratomas. They are rare benign tumors that have up to 0.2-2% chance of transforming into malignancy [1]. They are composed primarily of ectodermal tissue such as epidermis, hair follicles, sweat and sebaceous glands. Also, some mesodermal and rarely endodermal elements derived from residual embryonic cells. Dermoid cysts lie on or deep to the skin but not attached to it and is lined by stratified squamous epithelium. They are either congenital during antenatal development or acquired following an injury (implantation). They are result of incorporation of the ectoderm during closure of embryonic fissures. Therefore, they are usually found in the midline [2]. Even though they present at birth, they become apparent when they begin to enlarge. 7% of dermoid cysts are localized in the head and neck region [3].

Case Presentation

4 years old boy known case of hypertrophic cardiomyopathy and stable hydrocephalus. He is a preterm (36 weeks) who was admitted in the NICU and intubated for 15 days on ventilator due with intermittent preterm hypoxia. Also, he was diagnosed with left multicystic dysplastic kidney disease, for which he was on prophylactic antibiotic. The patient was diagnosed with bilateral profound sensory neural hearing loss since birth and he was included in a national cochlear implant program. The patient was fit with hearing aids at the age of 2 years and inconsistently worn them for 4 months. There was no history of ear pain, ear discharge or any other otolaryngology related symptoms. Additionally, no history of focal neurologic deficit has been reported. The patient has no facial dysmorphic features when compared to other siblings. Family along with child, underwent genetic screening with negative result. Brain CPA MRI under general anesthesia was done which revealed right hypoplastic cochlear nerve, and non-visualized left cochlear nerve. Also, bilateral cystic and hypoplastic cochlea with one-and-a-half turn, suggestive of Mondini malformation. CT was recommended for further assessment, which showed cochlea globular rounded appearance with no malleolus seen. Wide cochlear duct was noted, with normal

size internal auditory canal. Hypoplastic semicircular canal noted with residual posterior semicircular canal. Large left vestibule seen, with left normal size vestibular aqueduct. Normal facial nerve noted. The patient underwent cochlear prosthetic device implantation in the right ear, multiple channel surgery. Right postauricular incision around 6 cm was performed, deepened down till the temporal fascia and mastoid periosteum. A U-shaped superiorly based flap was elevated, and the mastoid cortex was exposed. An incidental finding of a cyst was seen in the post-auricular area around 5 cm behind the bony external auditory canal. The cyst was containing some keratinous material and hair. It was excised completely and sent for pathological examination, there was no intracranial extension of that cyst. Mastoidectomy was carried out. Facial recess approach was performed, and the cochlea was identified. Rest of the surgery went smoothly with no complication. Post-operatively, previous MRI was reviewed by a pediatric radiologist consultant showing a small cyst with restricted diffusion suggestive of dermoid-epidermoid origin cyst (Figures 1-4). Histopathological examination (Figure 5) shows dermoid cyst. Grossly, the specimen is brown soft tissue with tan-brown heterogeneous cut surface. Microscopically, sections show

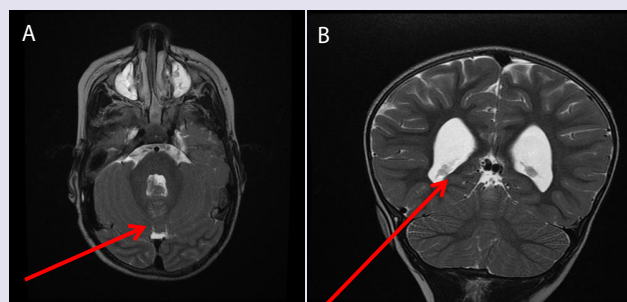


Figure 1A,B: Axial and coronal T2 FSE images show small cyst with high signal intensity in the right post-auricular mastoid space suggestive of fluid content, red arrow.



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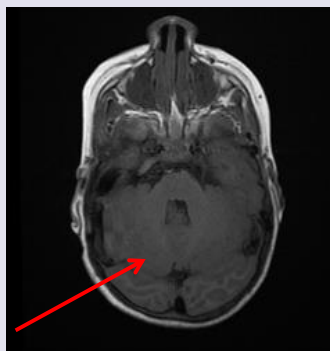


Figure 2: Axial T1 FSE image show small cyst with relatively high signal intensity in the right post-auricular mastoid space suggestive of proteinaceous material, red arrow.

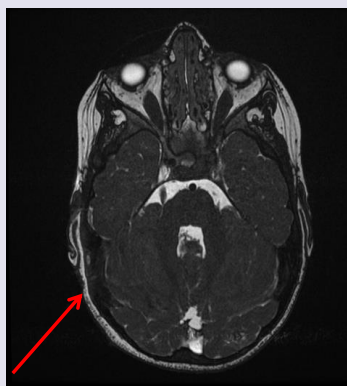


Figure 3: Axial T2 FIESTA sequence image show small cyst with relatively high signal intensity in the right post-auricular mastoid space, red arrow.

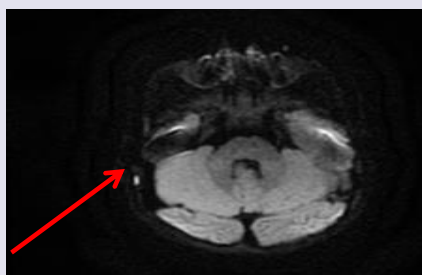


Figure 4: Axial DWI image show small cyst with restricted diffusion in the right post-auricular mastoid space suggestive of dermoid/epidermoid origin cyst, red arrow.

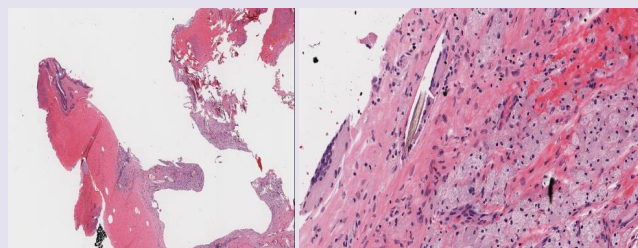


Figure 5: The cyst was lined focally by stratified squamous epithelium. Most of the lining, however, is obliterated by sheets of foamy histiocytes and multinucleated giant cells admixed with hair shafts.

skin, dense connective tissue and a cyst lined focally by stratified squamous epithelium. Most of the lining, however, is obliterated by sheets of foamy histiocytes and multinucleated giant cells admixed with hair shafts.

Discussion

Dermoid cysts are true benign ectodermal inclusion cysts. Although the majority of the cellular tissues are derived from ectoderm, they can contain tissue from all three germ layers. They are may be congenital which arise when epidermal cells are separated from the skin during fusion or acquired when the epithelial cells are implanted into the subcutaneous tissue. They tend to manifest in the 2nd-3rd decade due to the fact that they are fast growing [2]. The exact etiology of these lesion is still unknown, however most probable theories are deficient closure of fusion lines or traumatic implantation of skin elements [4]. The incidence of dermoid cysts is thought to be approximately 3 for every 10,000 pediatric patients [5]. The majority of dermoid cysts are frequently encountered in the gonads. Although they also occur somewhat less commonly as external angular or midline angular. It is estimated that 7% of the cysts develop in the head and neck. To elaborate, 50% of them develop in the orbital region, 25% in the oral cavity, and 13% in the nasal cavity. Dermoid cysts in the middle ear or mastoid bone have been reported a couple of times [2]. Dermoid cysts tend to differ in presentation depending on the site and the size of. In our case report, the patient presented with bilateral sensorineural hearing loss since birth. No ear discharge or pain was associated with it. It has also been reported to present solely as unilateral conductive hearing loss [6]. Recurrent otitis media or unremitting serious otitis media could also occur [1]. Lastly, only two cases reported vestibular symptoms, dizziness, and unsteady walking [4].

On CT imaging, dermoid cysts appear rounded, well-circumscribed lesion. The cysts are extremely hypodense due to their high fat content. The capsule can be classified. However, they don't cause vasogenic edema and only hardly associated with hydrocephalus. On MRI, dermoid cysts are hyperintense on T1-weighted sequences. On the contrary, they are variable from hypointense to hyperintense on T2-weighted images as a result of their high lipid content [4]. In our case, the lesion was not appreciated in the preoperative imaging as it was missed by the radiologist. Grossly, dermoid cysts are usually unilocular, polypoid, pedunculated with thick wall. The color ranges from grayish white to pink. Microscopically, the surface is lined by stratified squamous epithelium which contains epidermal appendages. The stroma of the cyst contains fibrous and fatty tissue. Also, it has mesodermal and ectodermal derivatives like cartilage, smooth or striated muscle, bone, salivary glands, nerves, and lymph nodes [1,4]. The definitive treatment for dermoid cysts is through complete surgical enucleation. The excision should be done as soon as possible to avoid un-necessary expansion of the cyst which could lead to destruction of the surroundings. It is the surgical cornerstone of any multidisciplinary approach to accurately prevent any chance of recurrence [1,4]. In head and neck region, dermoid cysts usually have a favorable prognosis. Although very uncommon, dermoid cyst can undergo malignant transformation as a complication of long-standing retained remnants [7-10].

Conclusion

The report describes a rare case of a dermoid cyst occurring in the mastoid bone that was incidentally found intraoperatively during cochlear implantation surgery. Complete surgical excision of the tumor has been done successfully.

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