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Case Report

Radiological findings of retroauricular dermoid with intra-calvarial extension

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ABSTRACT

Dermoid cysts are rare in head and neck region. A dermoid cyst arising in the retroauricular region over the mastoid can be easily misdiagnosed as a lymph node, thus posing significant diagnostic challenges. We present a case report of two year old boy with retroauricular dermoid showing intracalvarial extension. The child presented with the swelling in the right retro-auricular region with no associated hearing loss, tinnitus or vertigo. Provisional diagnosis clinically made was that of a large necrotic post auricular lymph node. On ultrasonography, well defined heterogeneous hypoechoic lesion showing few hyperechoic areas with scalloping and extension into the underlying temporal bone. On computed tomographic scan, heterogeneous non enhancing lesion showing areas of negative attenuation value seen in retroauricular region with extension into the temporal bone. Final diagnosis was confirmed by histopathological examination of the surgically resected specimen.

Keywords: Retroauricular dermoid, Dermoid cyst, Intra-calvarial extension, Post auricular lymphadenopathy

INTRODUCTION

Dermoid cyst is a benign cystic lesion, primarily situated close to the midline. Dermoid cyst comprises of both ectoderm and mesoderm, thus containing, mature squamous epithelium, keratin debris, hair follicles, sebaceous, sweat glands, smooth muscle and fibro-adipose tissue. Dermoid cysts are thought to arise from inclusion of ectodermal elements at the time of neural tube closure. Occurrence of dermoid cyst is rare, with only 7% of cases presenting in head and neck region; frequently encountered at lateral canthus, medial canthus and bridge of nose.[1]

CASE REPORT

A 2 year old boy was brought to our department with complaints of retroauricular swelling on right side noticed since birth which had grown slowly over the duration. On clinical examination, right auricle was displaced antero-laterally. Left auricle was normal. There were no clinical signs of inflammation associated with lesion and tenderness was absent. No growth was visible in bilateral external auditory canals. No associated hearing loss, tinnitus, vertigo was present.

On gray scale ultrasonography, well defined heterogeneous hypoechoic lesion showing few hyperechoic areas seen in right post auricular region with scalloping and extension into the underlying temporal bone corresponding to Figure (1a, 1b).

On computed tomographic scan, heterogeneous non enhancing lesion showing areas of negative fatty attenuation value seen in right retroauricular region with extension into the temporal bone. Few areas of erosion of

Korner's septum seen. However, no extension into the middle ear cavity or internal ear seen (Figure 2).

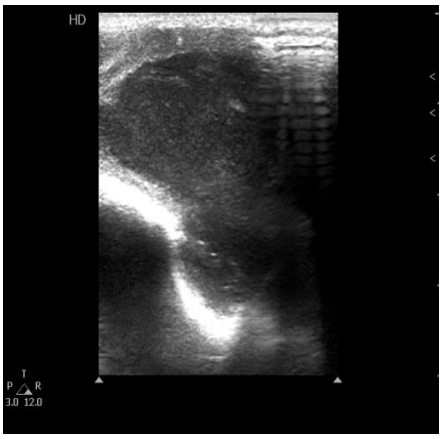

Figure 1a: Gray scale images reveal a heterogeneous hypoechoic mass with scalloping of underlying temporal bone.

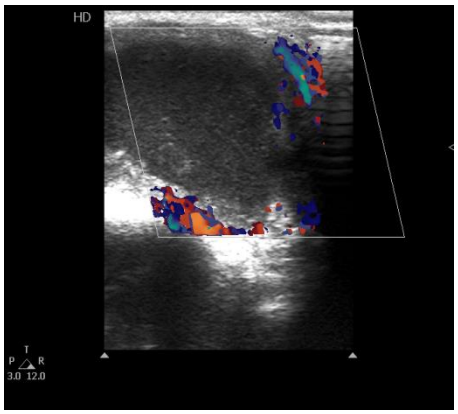

Figure 1b: Colour flow images reveal a heterogeneous hypoechoic mass with scalloping of underlying temporal bone showing no significant internal vascularity.

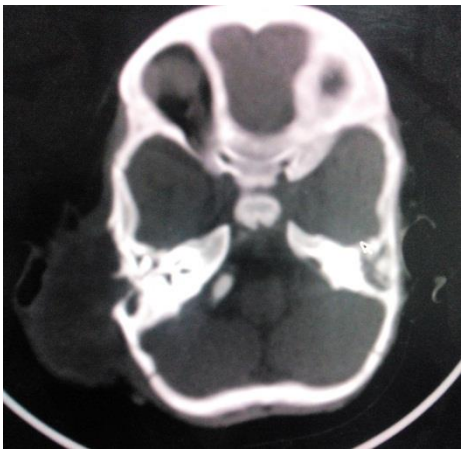

Figure 2: Axial CT section shows a large retroauricular mass with extension into adjoining mastoid part of temporal bone.

Axial Computed tomography bone window images reveal calvarial scalloping and intracalvarial extension. However, middle ear cavity and internal ear are intact (Figure 3a, 3b).

The patient underwent wide local excision. The mass was histopathologically proven to be dermoid cyst.

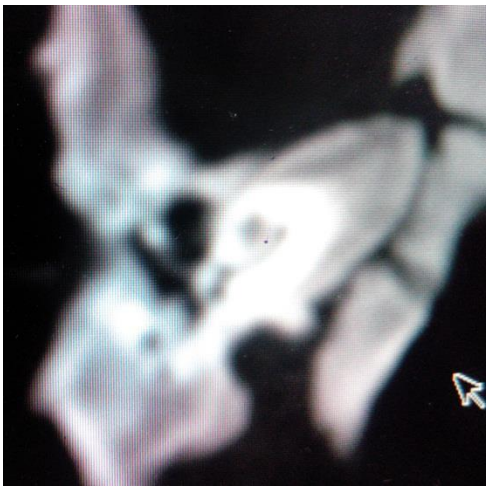

Figure 3a: Axial computed tomography bone window images reveal calvarial scalloping and intracalvarial extension. However, middle ear cavity and internal ear are intact.

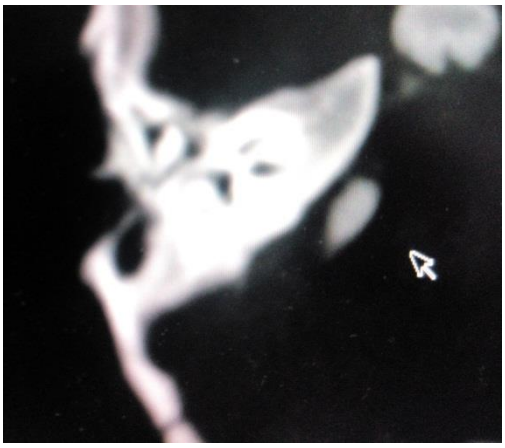

Figure 3b: Axial computed tomography bone window images reveal calvarial scalloping and intracalvarial extension. However, middle ear cavity and internal ear are intact.

DISCUSSION

Congenital dermoids represent benign developmental anomalies. They are derived from both ectodermal and mesodermal elements and originate during early embryogenesis. The exact aetio-pathogenesis remains a point of debate, though the most likely theories are traumatic implantation of skin elements or incomplete closure at fusion lines.[2]

Dermoid cysts are rare in head and neck region. They represent about 7% of all dermoids. As reported by New and Erich, about 49.5% of head and neck dermoids are located in the periorbital region, 25% are located in the oral cavity, and 13% occur in the nasal cavity.[1]

Twenty-four cases of dermoids of the temporal bone were reported in the English literature. Multiple sites of involvement within the temporal bone have been described.

Grossly, dermoid cyst is a thick walled unilocular cyst usually polypoid, pedunculated and rarely sessile. They appear grayish white or pink in color, covered by skin often containing thick greasy sebaceous material, keratin debris and skin appendages such as hair follicles. Microscopically, it is lined by stratified squamous epithelium that contains epidermal appendages. The stroma contains fibro-fatty tissue and may show cartilage, smooth or striated muscle and other mesodermal or ectodermal derivatives.

Clinical presentation varies according to the location of the dermoid cyst. Extra-cranial dermoids are usually not symptomatic unless infected. They are mostly of cosmetic concern only.

Toynbee reported the first case of a dermoid cyst of the mastoid in 1866.[3] Steel identified four cases in the literature from 1866 to 1976 that referred to non-hair-bearing cysts in the mastoid. Steel reported a 67-year-old male, with a dermoid cyst in the middle ear who had been treated for intermittently-active chronic otitis media for years.[4] Fried and Vernick reported a patient who had a dermoid cyst of the middle ear and mastoid aged 22 months.[5] Howie identified a dermoid cyst in the middle ear of a 29-year-old female who presenting with hearing loss and vertigo.[6] Minatogawa et al. reported a 6 year old girl with unilateral conductive hearing loss due to a dermoid cyst in the middle ear.[7] Farris et al. reported the youngest patient, an 8 month old infant, with a congenital dermoid cyst of the middle ear with a moderate conductive hearing loss of the ear in 1998.[8] Scolozzi et al. reported an intracranial dermoid cyst in a 1-year-old presenting with a cutaneous fistula in the frontotemporal region.[9] No gender predilection has been identified.

The present case is unique because it represents an extracranial dermoid in retroauricular location with intracalvarial extension. However, there is no associated hearing loss, vertigo, tinnitus.

Dermoid cysts due to their high lipid content are hypodense on Computed tomography scan. On MRI imaging dermoids appear hyperintense on T1-weighted sequences and are hypo- to hyperintense on T2-weighted sequences.

The treatment is complete surgical excision. These tumors show low post surgical recurrence rates due to their limited growth potential.

The prognosis of dermoid in the head and neck region is favorable. A rare complication in a long-standing dermoid cyst is malignant transformation. Tsugu et al. reported a case of squamous cell carcinoma (SCC) arising in an intracranial dermoid cyst.[10]

Devine and Jones reported a case of malignant transformation to squamous cell carcinoma of a long-standing sublingual dermoid cyst.[11]

Differential diagnosis of these tumors includes lymphadenopathy, epidermoid cyst, sebaceous cyst, subperiosteal abscess, hematoma.

In conclusion dermoid cysts must be considered in the differential diagnosis of lumps in the mastoid/retroauricular region, however uncommon. Imaging plays an important role in pre-operative diagnosis of lesion and providing the extent of lesion.

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