

Unusual Cases of Epidermoid cyst: Case Series

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Abstract

In the oro-facial region cystic lesions of different etiologies are encountered owing to the presence of the teeth in the jaw bones. A bewildering variety of developmental, odontogenic and non-odontogenic cysts are seen. Epidermoid cyst is a rare developmental cyst of the oro-facial region which results from entrapped epidermal elements without adnexal appendages. Dermoid and epidermoid cysts occur in oro-facial region with an incidence of 6.9-7% and represents less than 0.01% of all oral cavity cysts. Here we report two cases of epidermoid cysts occurring at unusual locations involving upper left maxillary region lateral to the nose and pinna of the ear.

Keywords: Cyst, Epidermoid Cyst, Dermoid Cyst, Pinna of the Ear, Maxillary Region

Introduction:

Epidermoid cysts are non-odontogenic inclusion cyst lined by ectoderm.¹ These are rare lesions derived from germinal epithelium and are encountered throughout the body, in areas where embryonic elements fuse together.^{2, 3, 4, 5} Most cases have been reported in ovaries and the testicles, with 7% occurring in the oro-facial area and 1.6% in the oral cavity, representing 0.01% of all oral cavity cysts.^{4, 5}

Epidermoid cysts are indolent in nature, slow to progress and remain asymptomatic unless secondarily infected. Larger cyst can cause obstructive signs and symptoms like dyspnoea and dysphagia.^{3, 6, 7} In the current report, we describe two cases of epidermoid cyst in the upper left maxillary region and pinna of the ear, which are unusual sites of epidermoid cysts.

Case Reports

Case Report 1:

A 30 yrs old male patient presented with a swelling on the left malar region since 2 months. History revealed that the swelling was asymptomatic

and gradually increasing in size. On examination, a single well circumscribed swelling was seen in the left malar region, ovoid in shape, 1.5x1.5 cm in greatest diameter, skin over the swelling was smooth and of the normal color without any secondary changes, non-tender on palpation, soft and fluctuant in consistency. No dental abnormality was detected. Fine needle aspiration cytology (FNAC) revealed a creamy white fluid. A provisional diagnosis of Sebaceous Cyst was made and following complete excision of the swelling, the specimen submitted for histopathologic investigation. Gross examination shows a soft tissue specimen of size 2 x 2cm, oval in shape, yellowish white in color and cystic in consistency. Cystic lumen was filled with thick creamy, cheesy like material (Fig1). The entire tissue was kept for processing. Histopathological examination revealed cystic lining of keratinized stratified squamous epithelium which was 4-5 cell layers thick with numerous keratin flakes in the lumen. The surrounding connective tissue capsule was made up of collagen fibres, blood vessels, chronic inflammatory cell infiltrate and there was absence of dermal appendages (Fig 2, 3).

Case report 2:

A 27 yrs old male patient presented with swelling on the right ear lobe since 15 yrs. History revealed that the swelling has remained constant without any increase in size and is asymptomatic since the past 15 years. On examination, a single, well circumscribed swelling was seen in the right pinna, ovoid in shape, 1x1 cm in greatest diameter, skin over the swelling was smooth and of the normal color without any secondary changes, non-tender on palpation, firm in consistency. FNAC revealed no aspirate. A provisional diagnosis of benign soft

tissue neoplasm was made. The swelling surgically excised and the specimen submitted for histopathological examination. On grossing, the soft tissue specimen was approximately 1 x 2 cm in size, ovoid in shape, yellowish white to brown in color, firm in consistency (Fig 4). The entire tissue was kept for processing. Histopathological examination shows cystic lumen lined by keratinized stratified squamous epithelium and the surrounding connective tissue. Capsule lumen was filled with keratin flakes and there was no evidence of dermal appendages in the connective tissue wall (Fig 5, 6).



Fig 1- Gross examination shows a soft tissue specimen oval in shape, yellowish white in color and cystic in consistency.

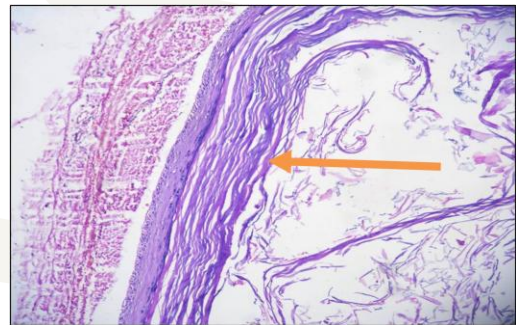


Fig 3- Under low power, histopathological examination shows numerous keratin flakes in the lumen (orange arrows) - 10X.

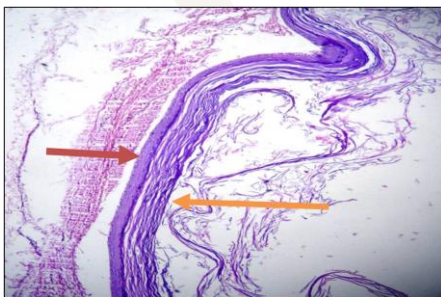


Fig 2- Under scanner view, section shows cystic lining of keratinized stratified squamous epithelium (red arrows) with numerous keratin flakes in the lumen (orange arrows) - 4X.



Fig 4- Gross examination shows a soft tissue specimen showing numerous keratin flakes.

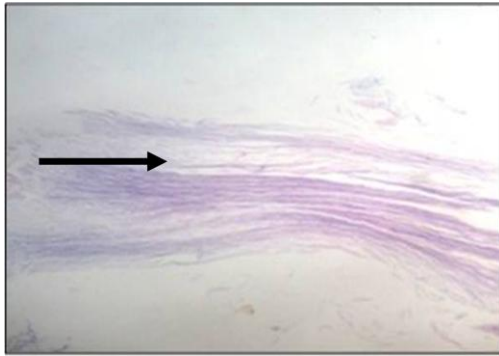


Fig 5- Under low power, section shows numerous keratin flakes in the cyst lumen (10X).

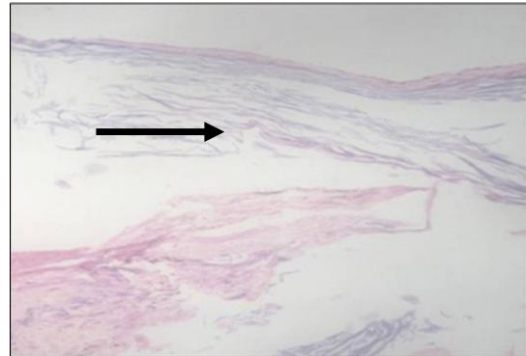


Fig 6- Under low power, section shows absence of dermal appendages in the connective tissue wall (10X).

Discussion:

Roser, in 1859 first described epidermoid cyst.⁸ These are rare benign conditions in the oro-facial region derived from abnormally situated ectodermal tissue. They can develop anywhere in the body, with the incidence ranging from 1.6% to 6.9% occurring in the oro-facial area and 1.6% within the oral cavity and represent less than 0.01% of all oral cavity cysts.⁹

Depending on the pathogenesis, Epidermoid cyst can be divided into:

- 1) Congenital
- 2) Acquired

Congenital cysts are dysembryogenic lesions that arise from ectodermal elements entrapped during midline fusion of the first and second branchial arches between the third and fourth week of the intrauterine life. Alternatively, they may also arise from tuberculum impar of His.^{10, 11, 12}

Acquired cyst are derived from traumatic or iatrogenic inclusion of epithelial cells or from occlusion of sebaceous gland duct, it was first recognized by Werhner in 1855 and originally referred to as "Implantation cyst" by Sutton in 1895.^{11, 12, 13}

There are two theories for epidermoid cyst formation: Firstly, Epidermoid cyst may occur when two epidermal surfaces fuse together during early intrauterine life and an ectodermal implant is

retained deep to the surface. Secondly, due to traumatic entrapment of surface epithelium in the connective tissue; later these cells may differentiate to form cyst.^{13, 14, 15}

In 1955, Meyer updated the concept of epidermoid cyst to describe three historical variants:

Dermoid cyst: Epithelium lined cystic cavity encloses skin appendages such as hair, hair follicles, sebaceous and sweats glands.

Epidermoid cyst: Epithelium lined cystic cavity without skin appendages.

Teratoid: The cyst cavity encloses mesodermal derivatives such as bone, muscle along with skin appendages.^{4, 5, 16, 17}

Epidermoid cyst is mainly reported from face, trunk, neck, extremities and scalp. In the oral cavity, floor of the mouth is the most common location, and occasional occurrence have been reported involving buccal mucosa, tongue, lips, uvula and intraosseous location within the mandible and maxilla.^{3, 6, 7} This is consistent with our case that occurred at unusual location involving upper left maxillary region lateral to the nose and pinna of the ear.

Epidermoid cysts are generally diagnosed in young adults in the second and third decades of life. It is twice as common in men as in women with a male to female ratio of 3:1.^{3, 6, 7} The clinical findings in our case were consistent with previous cases except for site of occurrence.

The epidermoid cyst rarely discloses malignancy.⁵ The occurrence of Basal cell carcinoma, Bowen disease, and Squamous cell carcinoma has been reported in the literature that had evolved from epidermoid cyst.⁷ Dini et al¹⁸ described a patient with basal cell carcinoma arising in the wall of an epidermoid cyst. Ikeda et al¹⁸ presented a case stating that basal cell carcinoma originates from an epidermoid cyst in which nests of basal cell carcinoma connected with the epidermoid cyst and partially replaced the cyst wall. Lopez-Rios et al¹⁹ described a case in which squamous cell carcinoma had evolved in the wall of conventional epidermoid cyst. An incorrect diagnosis could result in inappropriate therapy and if the lesion is completely excised, the treatment is definitive.⁵

Conclusion:

The cases presented show no variation from the normal histopathology, but, they prove to be significant, because of the variation in their anatomical presentation. Epidermoid cysts of an oral cavity are an uncommon entity. Ample understanding and vigilance about this slow growing mass is essential not only because of the symptoms it produces but also due to the malignant potential.

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