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Case Report

EPIDERMAL INCLUSION CYST - A REPORT & REVIEW ON ITS DIAGNOSTIC APPROACH

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Abstract

Epidermoid cysts are common benign intradermal or subcutaneous tumors. Their prevalence is 7% in head and neck patient and 1.6% within the oral cavity. We present the case of a 30 year old male with a solitary epidermoid cyst on the right supra-orbital region which was asymptomatic but aesthetics being his primary concern. Clinical examination was performed and decided upon initial investigation with FNAC followed by surgical enucleation of the lesion. Histopathological findings of the excised lesion revealed that it was an epidermoid cyst. From the surgical point of view, they have a very good prognosis and plus it is a non-aggressive lesion. Even Though this is the classical site for occurrence of epidermoid cysts the removal of these cysts is of great concern as it can cause social stigma, aesthetic and functional impairment.

Keywords : Cutaneous cyst, Epidermoid cyst, Diagnosis

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INTRODUCTION

A cyst is defined as "a pathological cavity having fluid, semi-fluid, or gaseous contents and which is not created by accumulation of pus" - Kramer 1974 which may or may not be lined by epithelium (1). Various cysts exhibit various clinicopathological and histopathological characteristics, of which one of the primordial features is the presence of epithelial lining. "True cysts" are the ones which are lined by either simple epithelium or stratified squamous epithelium and in contrast, the cysts which are not lined by epithelium are called the "Pseudocysts" (2,3).

Epidermoid cysts are the most common type of cutaneous cyst which are slow-growing, painless and fluctuant cyst; seen underneath the skin, with or without a prominent central punctum that represents the plugged orifice of the pilosebaceous follicle (2,3). Epidermoid cysts constitute <0.01% of all the oral cysts whereas considering the head & neck region, its occurrence has been increased to about 7% (4,5). Still its incidence in the head & neck region is relatively less, hence are more likely for misdiagnosis (6). They are mostly sporadic but familial inheritance is also possible in case of multiple lesions (2,3). Lesions typically are spontaneous, also injury-induced epithelial implantation, on the other hand, is regarded as an etiologic factor. They occur along the embryonic fusion lines which includes face, scalp, neck and chest. Also seen infrequently in the oral cavity. These cysts are seen prevalent at any age yet slight predilection for second and third decades of life. Small, uncomplicated epidermoid cysts are usually untreated. Removal can be achieved through a simple surgical excision of the cyst with no damage to the cyst wall (2,3).

CASE REPORT:

A 30-year-old male presented with a swelling on the right supraorbital region for the past 3 years which was asymptomatic slow growing mass with evident increase in size. The patient could not precisely narrate the incidence of the lesion unless it became evident. Also, no history pertaining to multiple similar lesions. Medical history was noncontributory to the lesion. On examination, the lesion was a solitary, well defined, smooth-surfaced, normal colored mass seen protruding beneath the skin in the right supraorbital region with no surface changes evident. Approximately, it measured about 3x2 cm in size [Figure 1a & 1b]. On palpation, the lesion was partially mobile, with slight compressibility and a positive slip sign noted. No tenderness or temperature alterations were noted. No radiographic history contributing to the diagnosis of the lesion.

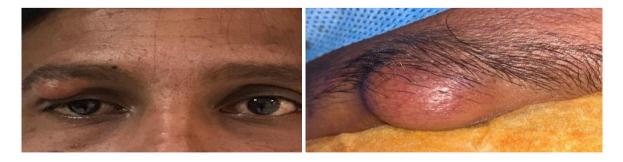


FIGURE 1a showing the anatomy - right lateral supraorbital region and 1b showing the clinical display of the cutaneous cyst.

Fine needle aspiration cytology (FNAC) was performed. The aspirate was received to the department of Oral pathology and Microbiology in a 5 ml syringe. The aspirate was cheese-like thick malodorous material. Conventional smears were made on a clean glass slide and fixed with 99.9% isopropanol for 30 minutes and H&E staining was performed. The H&E stained smear showed abundant anucleated squames and few flakes of keratin in an inflammatory background which pointed to a provisional diagnosis of epidermoid cyst on correlating with the clinical history. Yet, the entire excisional biopsy specimen was requested to be submitted for final diagnosis post fixation, tissue processing and sectioning. Surgical excision of the lesion was performed under local anesthesia [Figure 3].



FIGURE 3 showing the surgical excision of the cutaneous cyst.

Macroscopically, the lesion appeared encapsulated and was flocculent with a measurement about 1.1x0.9x0.7cm approximately. The specimen was cut into equal halves and it contained a creamy white cheesy material [Figure 4]. The specimen was fixed using 10% neutral buffered formalin for 24 hours and submitted to a series of reagents and finally sectioned and stained with H&E.



FIGURE 4 showing the gross image of the excised specimen. The specimen was brownin color and soft to cystic in consistency. The specimen was cut and a curdy white and disintegrated material was evident. The specimen along with the material was subjected to histopathological processing.

Microscopically, the lesion showed a cystic cavity lined by orthokeratinized stratified squamous epithelium with uniform 5-10 cell layer thickness. The epithelium is composed of stratum basale with a single layer of palisaded columnar to cuboidal cells; stratum spinosum with few layers of polygonal cells and the superficial layer is the stratum granulosum with 2-3 layers & most importantly the prominent keratohyalin granules within the cytoplasm of these cells to produce abundant keratin [Figure 5a]. There is evidence of abundant anucleate keratin within the cyst lumen suggestive of orthokeratin [Figure 5b]. The keratin present within the lumen are seen as flakes. The interface between the epithelium and connective tissue is almost flat with no rete pegs formation. The supporting mature cyst wall showed dense collagen with moderate chronic inflammatory cell infiltrate, predominantly the lymphocytes along with moderate vascularity and other connective tissue components. Final histopathological diagnosis was given as Epidermoid cyst with relevant clinical correlation.

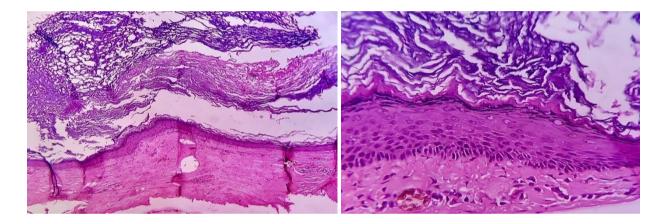


FIGURE 5a shows the epithelial lining with abundant keratin and the minimal connective tissue wall in 10X & 5b shows the stratifications of the epithelial lining with excessive flakes of produced orthokeratin.

DISCUSSION:

Multiple theories have been proposed to explain the development of epidermoid cysts. The benign cystic lesions known as epidermoid cysts are caused by the trapping of surface epithelium or, more typically, by the infundibular epithelium's aberrant repair during a follicular inflammatory episode (4,7). They are categorized as congenital and acquired typed based on their pathogenesis. Congenital type - Dysembryonic lesions that develop from entrapped ectodermic elements during the midline fusion of the first and second branchial arches between the third and fourth weeks of intrauterine life. Alternatively, they may arise from His tuberculum impar, which forms the floor of the mouth and the body of the tongue with each mandibular arch (8). Acquired type - The implantation of epithelium into deeper mesenchymal tissues via surgical or accidental trauma. Trauma and prior surgical history is known to be an important etiology of acquired type but due to a lack of adequate history, congenital type is believed to exist increasingly than the acquired one (9,10).

Epidermoid cysts have been referred to as epidermal cysts, epidermal inclusion cysts, follicular infundibular cysts and sebaceous cysts (11). The term epidermal inclusion cyst refers to an epidermoid cyst caused by the implantation of epidermal elements in the dermis. Because most lesions arise from the follicular infundibulum, the more general term epidermoid cyst is preferred. But the term sebaceous cyst was not accepted because of its absence to exhibit sebaceous glands within the lining (2,3).

The occurrence of epidermoid cysts ranges between 10-72 years of age, with frequency in 2nd - 3rd decades of life. The high incidence of young adults could be due to excessive secretion of fat affected by pubertal hormonal stimulation can stimulate cyst growth. Gender preferences have been reported to be inconsistent with themselves, and few reports have supported male dominance (7,12,13).

Previous literature identified with 103 epidermoid cyst diagnosed patients showed head & neck lesions with 46.6% of these were orbital, 23.3% buccal and submental, 12.3% nasal, 10.7% cervical and 2.9% labial (6). The floor of the mouth, tongue, lips, palate, and the sublingual region are some of the documented sites in the oral cavity for epidermoid cysts, which make up less than 0.01% of all oral cysts (4). Location of the occurrence of the epidermoid cyst plays a pivotal role in the diagnosis of the lesion and also to differentiate from other cutaneous tumors and cysts.

Radiographic imaging of extensive epidermoid lesions can contribute to the specific diagnosis. In ultrasound, they have a circular structure, with a well-circumscribed, avascular mass of subcutaneous tissue with the phenomenon of dorsal acoustics reinforcement and lateral shadowing. On MRI, a slightly hypointense signal strength weighted to T1 medium to high signal weighted by T2 will be seen. Limited diffusion is typical of epidermoid cysts. These characters help distinguish epidermal cysts from neoplastic lesions (2).

The diagnosis of epidermoid cyst is usually made clinically. This entity is frequently confused with other cutaneous tumors such as lipoma, cystic basal cell carcinoma, trichilemmal cyst and other variants of cutaneous cysts especially the 'Dermoid cyst' because of its diagnostic similarities but "Epidermoid cyst" and "Dermoid cyst" are two different entities. Both cysts contain abundant keratin produced by keratinizing squamous epithelium yet the histopathological difference between these two entities is that dermoid cysts have skin appendages on their lining while epidermal cysts do not have these appendages (14). A single lesion of an epidermoid cyst is seen to be more protruding than the dermoid cysts.

Lipomas are mature adipose tissues that are lobulated, sluggish to grow, and have little connective tissue stroma. They often are found in the subcutaneous tissues and are not connected to the overlying tissues. Hence clinically these lipomas are mobile to an extent and they are firm since their main component is adipose tissue rather than a liquid/ semi-solid/ gaseous content (15). Lipoma can be considered as clinical differential diagnosis rather than for histopathological analysis.

A basal cell carcinoma cystic lesion may have telangiectasia on the surface but does not have a distinct central punctum clinically (2,3). Histopathologically, they present with an epithelial lining and cystic spaces. The epithelium is stratified squamous epithelium with palisaded basaloid cells. Clefts between the stroma and tumor edge, which are often seen in typical basal cell carcinomas (16).

Histopathological picture of trichilemmal cyst shows an epithelial lined cyst with an attenuated granular layer and abrupt keratinisation which is adherent to the epithelium. A closer inspection of the cyst wall identifies trichilemmal differentiation as it occurs in the outer root sheath of the hair follicle. This is seen as maturation of squamous epithelium with lack of a granular layer (17).

Genetic disorders which may increase the risk of developing multiple epidermoid cysts include Gardner syndrome, Pachyonychia congenita type 2 and Basal cell nevus syndrome (11).

Another primary histopathological differential diagnosis is the orthokeratinized odontogenic cyst (OOC). Even Though it is a completely different entity the histological picture of dermoid/epidermoid cyst and OOC is very similar in terms of epithelial lining and type of keratin production. When compared to keratocystic odontogenic cysts, epidermoid cysts contain laminated keratin in the stratified squamous epithelium-lined cyst, whereas keratocysts have keratinizing lining epithelium with corrugated parakeratin layer and satellite cysts in cystic capsule (18).

Surgical excisions are based on the location of the dermoid cysts. The prognosis is excellent with previous reports having noticed no recurrence post-surgery. These cysts cause specific complications such as cyst rupture, abscesses, secondary and infectious diseases during treatment, making it a differential diagnosis for keratin cysts. Therefore, these cysts have the potential for malignant transformation and require thorough histopathological examination and close follow-up (19). Malignant transformations have also been noticed in head and neck cutaneous lesions but not commonly in the oral cavity (12). Approximately 1% of epidermoid cysts have been noted to have a malignant transformation to squamous cell carcinoma and basal cell carcinoma (11).

CONCLUSION:

Epidermoid cysts are benign, slow-growing, high, round, firm, subcutaneous, or intradermal cysts that typically grow 1–5cm in diameter but not > 5cm (very rare) and are asymptomatic with common occurrence in face, neck and upper trunk, still can be seen anywhere in the body. Epidermoid cyst can be successfully diagnosed and treated with a simple and effective procedure. An accurate diagnosis requires general knowledge of the etiology, epidemiology, pathophysiology, histopathology, clinics and radiology. Surgical resection is a cure that can be selected and performed under local anesthesia without anticipation of recurrence.

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