Case Report

Multiple generalize epidermoid cysts with giant epidermoid cyst: An uncommon and rare case report

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ABSTRACT

An epidermoid cyst (EC) is a fairly common lesion observed in young and middle-aged populations. A giant EC is described as having a size of more than 5 cm. A generalized EC involving multiple sites in the body is quite rare. Here, we present a case of a 72-year-old male presenting with a generalized EC with a giant cyst measuring 6 cm over the left knee. There was no other systemic illness. The aim of this paper was to report such a rare clinical presentation of ECs.

Key words: Giant epidermoid cyst, Multiple cyst, Inclusion cyst

Epidermal inclusion cysts are extremely common benign cystic lesions of the skin that present as fluctuant skin-colored nodules with a punctum and soft whitish keratin as their cyst contents. They are commonly known as epidermoid cysts (ECs), epidermal cysts, epidermal inclusion cysts, infundibular cysts, or keratin cysts [1]. These are slow-growing cysts with sizes ranging from a few millimeters to a few centimeters. Giant epidermal cysts have a diameter of over 5 cm and are of rare clinical occurrence, with only a few case reports. Epidermal cysts may be multiple but uncommon, with few cases reported on the face, scalp, and gluteal region. However, generalized multiple ECs are very rare.

To the best of our knowledge, only two such cases have been reported till date [2,3]. We present the third case of multiple generalized ECs with a giant EC in an elderly male.

CASE REPORT

A 72-year-old male presented with complaints of multiple nodular swellings all over the body since childhood. There was no history of any systemic complaints. Physical and mental development was normal. There was no history of trauma, surgery, or drug injection. A family history of similar swelling was present. His sons and grandchildren also had similar swellings. These swellings gradually increased in number and size since childhood over time.

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The swellings were multiple, ranging in size from 1 cm to 6 cm in diameter, and were present over the face, body, back, knee, legs, forearms, and finger (Fig. 1a). The largest nodule measuring 6 cm was present over the left knee (Fig. 1b). The nodules were well-circumscribed, painless, and slowly progressive in nature without any surface ulceration or signs of inflammation. On palpation, the nodules were non-tender and firm to cystic in nature.

Routine laboratory tests, including complete blood count, renal function tests, liver function tests, other biochemical parameters, and urinalysis, were within normal limits. Ultrasonography revealed multiple subcutaneous hypoechoic cystic lesions.

Fine needle aspiration cytology was done from multiple sites, including nodules over the proximal and distal phalanx of the right ring finger, left forearm, and left knee. The nature of the aspirate was dirty and foul smelling from all the sites. Smears from all the sites showed similar features. They showed abundant anucleate squames with few keratinizing squamous cells over a dirty background. There was no evidence of fungal elements, inflammation, necrosis, dysplastic cells, or any evidence of malignancy (Fig. 2a). The largest cyst was excised. However, the patient was reluctant to go ahead with further surgical resection. We received the gross as a solid cystic mass measuring 6 × 3 × 2 cm. The cut section showed multiple cysts containing dirty and flaky material (Fig. 3). Histopathological photomicrograph showed a well-circumscribed lesion with an intact cyst wall, lined by stratified squamous epithelium with epidermal keratinization, keratohyaline granular layer and abundant keratin flakes as its contents. There was no evidence of any fungal elements, infection, giant cell reaction, or any evidence of malignancy (Fig. 2b).

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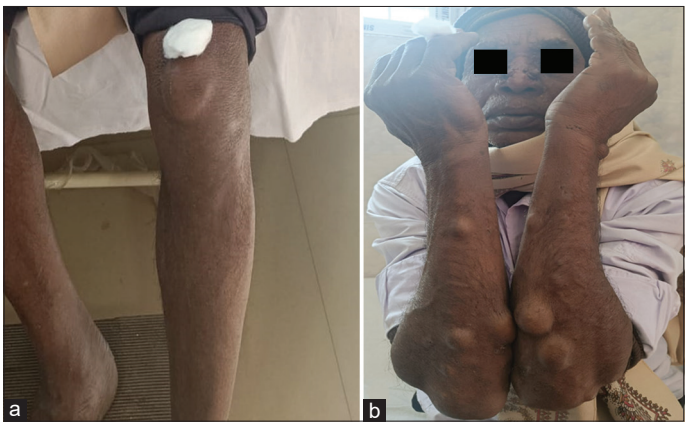

Figure 1: (a) Giant subcutaneous swelling over left knee; (b) Multiple subcutaneous swellings over both forearms and face

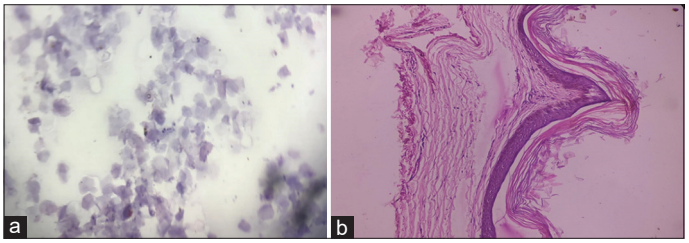

Figure 2: (a) Plenty of anucleate squames without any inflammatory cells (Diff-quick stain, ×400); (b) Skin epithelium shows epidermal keratinization, granular layer, and keratin flakes (H and E stain, ×400)

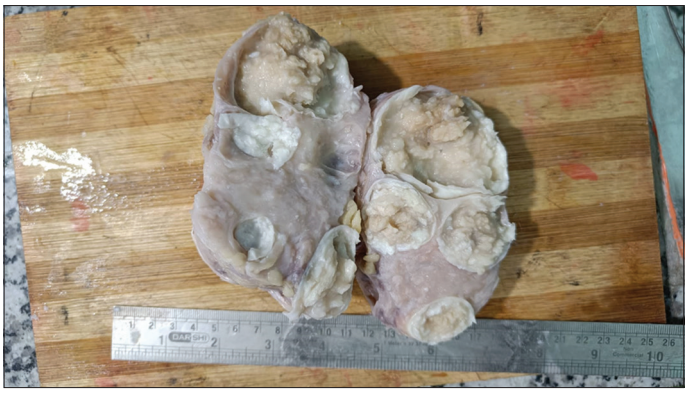

Figure 3: Cut section of solid and cystic grayish white mass measuring 6×3×2 cm shows cyst cavities filled with cheesy flaky material

Hence, the diagnosis of recurrent generalized and multiple epidermal inclusion cysts was rendered.

DISCUSSION

EC are common, slow-growing, smooth, dome-shaped swellings mimicking skin tumors in young and middle-aged adults, affecting both males and females. These are the cysts filled with keratin and a connecting duct to the skin surface. The immunohistochemical profile of ECs is similar to the infundibulum of hair follicles [4].

The pathogenesis starts from the blocking of comedones due to dense keratinocytes filling the duct [5]. The plugging of hair follicular orifices with bacteria or keratin can lead to cystic dilation and keratin debris entrapment. Penetrating skin trauma may also complicate the implantation of the squamous epithelium into the dermis [1,6]. They are slow-growing, with a growth rate of <0.5 cm/year [2]. The diameter of ECs usually ranges from a few millimeters to 5 cm. The preferential location of EC in the face, chest, and back may be explained because of the abundance of the pilosebaceous units. ECs are usually solitary; however, cases of localized multiple ECs on the face, scalp, and gluteal regions have been reported with a past history of acne vulgaris during adolescence [7]. The role of human papillomavirus in EC formation due to metaplasia of eccrine duct epithelium has been proposed [8].

Giant ECs are commonly found in the gluteal region, scrotum, extremities, and scalp [3,9]. It usually results from a long duration of EC, leading to gradual accumulation of keratin and neglected asymptomatic swelling. They are of clinical importance as there is a risk of malignant transformation such as squamous cell carcinoma, basal cell carcinoma, melanoma, Merkel cell carcinoma, and mycosis fungoides, as reported in the literature [10-12]. Generalized multiple ECs are extremely rare, with just two cases reported till date [2,10,13].

Commonly, these lesions are sporadic; however, familial inheritance is possible, especially in individuals with multiple ECs, which can be irregularly distributed or localized in the retro auricular area. Atypical localization and multiple ECs may be associated with autosomal dominant Gardner syndrome, X-linked Lowe syndrome, or Gorlin syndrome. ECs may also result from chronic sun damage and long-standing comedones [5,14]. Epidermal cysts can also occur with the use of immunosuppressants such as cyclosporine or tacrolimus in post-transplantation settings [15]. The present case was not associated with any syndromes.

Small, uncomplicated cysts usually do not require treatment. Larger cysts can be removed through complete surgical excision of cysts or with punch biopsy and expulsion of intact cysts through small defects [13]. Larger cysts require cold steel surgery, whereas smaller cysts can be treated using CO_2- or erbium-YAG-laser. Draining epidermal cysts using erbium: Yttrium aluminum garnet laser has also been reported as an effective treatment option with good cosmetic results [16]. Surgery should be avoided during active inflammation of the cyst due to increased risk of infection, wound dehiscence, and cyst recurrence.

CONCLUSION

ECs are common skin lesions. It is alarming when the lesions are multiple and generalized and larger than 5 cm. To ensure accurate diagnosis, surgical excision through histopathological examination is essential, though malignant transformation is rare.

We are presenting this rare case to the best of our knowledge; few cases have been reported as multiple EC with giant cysts to date.

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