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UNILATERAL LOCALIZED STEATOCYSTOMA MULTIPLEX- A CASE REPORT

Dermatology

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ABSTRACT

Steatocystoma multiplex is a rare inherited condition of the pilosebaceous unit characterized by the appearance of multiple cystic masses over the chest, arms and sometimes over the face at the time of puberty. We report a case of steatocystoma multiplex in an adult male patient localized to the abdomen and back in a unilateral distribution

KEYWORDS

steatocystoma multiplex, pilosebaceous unit, Keratin 17

INTRODUCTION:

Steatocystoma multiplex as a term was first coined by Pringle in 1899 though it was described earlier by Jamieson in 1873[1]. It is an autosomal dominantly inherited disorder though several sporadic cases have been reported, wherein there occurs hamartomatous malformations of the pilosebaceous duct junction. Clinically this is characterized by appearance of numerous small (2-20 mm) semi translucent skin coloured to yellowish compressible cysts over the chest and outer arms at puberty.

We report a case of unilateral localized steatocystoma multiplex as a sporadic occurrence in a 43 year old male patient.

CASE REPORT:

A 43 year old male patient came to the OPD with complaints of multiple small swellings over the right side of his abdomen and back for the past 25 years. It started appearing over the abdomen then slowly progressed to involve the other sites. They were otherwise asymptomatic. The patient did not give any history of itching, pain, fever, weight loss or any other constitutional symptoms. He had no other comorbidities. Patient does not give history of similar lesions in other family members. He did not seek any treatment for the lesions as they did not bother him.

General and systemic examination of the patient revealed no abnormalities. Dermatological examination revealed multiple well circumscribed yellowish- brown papules and nodules (2-6 mm) over the right anterior and lateral aspects of the abdomen and a few similar lesions were noted over the back. The lesions were soft, compressible, and non-tender and lacked a central punctum.

Biopsy from the lesion was taken for histopathological study and it revealed collections of sebaceous glands in the dermis surrounded by a layer of epithelial cells with an absence of granular layer in a wavy pattern. All these findings were suggestive of steatocystoma multiplex.

DISCUSSION

Steatocystoma multiplex is an uncommon skin condition of the pilosebaceous unit characterized clinically by the appearance of multiple sebum filled dermal cysts most often manifesting during adolescence; however, it can occur at any age with cases being reported since birth. It is inherited as an autosomal dominant condition but several sporadic cases have been reported.

Mutations to the keratin (K) 17 gene have been cited as the cause for the condition. Keratin 17 is a type 1 cytokeratin found in nail bed, outer root sheath of hair follicles and sebaceous glands. Keratin 17 is responsible for the strength and resilience of the skin, nails, and other tissues and when altered the defective keratin network leads to the disruption of growth and function of these cells. These abnormalities result in the formation of sebum-containing cysts in people with steatocystoma multiplex. The observation that it commonly appears during puberty suggests a possible hormonal trigger.

Clinically the patient presents with multiple soft skin coloured to yellowish compressible cysts of size ranging from 2mm – 20 mm in size; commonly over areas with high sebaceous gland density like chest, arms, axilla and neck[2]. The lesions are mobile and the surface of skin lacks any visible punctum. When punctured the cysts exude an oily fluid. The cysts are asymptomatic however when the contents are infected due to bacterial colonization the discharge becomes malodorous with significant scarring and sinus tract formation; this is then termed as steatocystoma multiplex suppurativa[3]. The lesions commonly occur in a generalized pattern however rarely lesions may be localized to certain sites like scalp, face, retroauricular area, groin etc.

Histopathological study of the lesions reveals multiple cysts lined by a wavy homogenous, eosinophilic cuticle. The walls are formed by several layers of squamous epithelia with embedded lobules of sebaceous glands amongst the epithelial cells. There is a characteristic lack of granular layer. An abortive epithelial tract may extend from the cyst to the epidermal surface. The cystic spaces are filled with keratin, vellus hairs and sebum esters. Electron microscopic examination shows cyst wall cells undergoing trichilemmal keratinization just as in inner hair root sheath of the hair follicle.

A close differential diagnosis for this condition include eruptive vellus hair cysts which has a strikingly similar clinical presentation but can be differentiated histologically by the absence of sebaceous glands within the lining epithelium and the presence of a granular layer, also eruptive vellus hair cysts have not been associated with K17 mutations [4]. Other differential diagnosis would include acne conglobata, sebaceous adenomas, cystic sebaceous hyperplasia, dermoid cysts and syringomas

The condition carries an excellent prognosis and there are no reports of malignant transformation of the tumors. The treatment of the condition is primarily surgical however infected lesions may have to be managed with appropriate antibiotics. Isotretinoin has been tried to varying degrees of success however flare up of lesions following its administration has been reported in a few cases. Cryosurgery is an option but carries the risk of residual scarring and pigment alterations. Simple incision and drainage of lesions with an 18 gauge needle has proven to be effective with much better cosmetic outcome. Surgical excision becomes impractical in situations with multiple lesions and

scar formation limits its use. Electrosurgery employing radiofrequency (RF) incision probes to make small incisions followed by manual extrusion of contents and cyst wall removal has shown good results in some reports with minimal or no scar formation or depigmentation[5]. CO2 lasers are increasingly being used as it carries the advantages of being able to treat multiple lesions without the need for anesthesia and provides good cosmetic outcome to the patient

CONCLUSION

Steatocystoma multiplex is an uncommon benign inherited disorder of the pilosebacous unit most often appearing during the time of adolescence. It is usually reported to occur in a generalized distribution in affected patients; however, the lesions may be confined to a few anatomic sites. Here we have reported one such case wherein lesions were confined to the abdomen and back in a unilateral distribution.

ACKNOWLGEMENT

None

CONFLICT OF INTEREST:

The authors declare they have no conflict of interest

Figure 1: Clinical picture showing multiple yellow- brown well circumscribed cysts over the abdomen

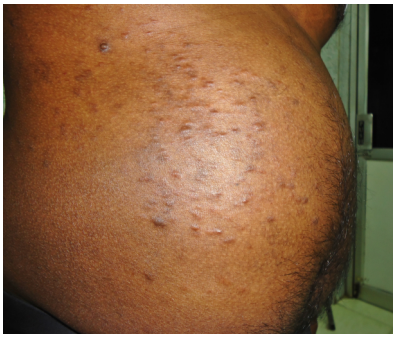


Figure 2: Histopathological picture on scanning view showing proliferation of sebaceous glands in the dermis surrounded by squamous epithelium

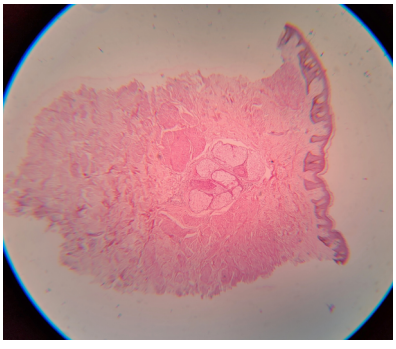
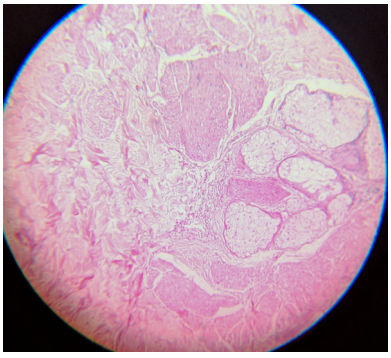


Figure 3: Histopathological picture on low power showing the squamous epithelium in more detail, there is a lack of granular layer



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