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Steatocystoma multiplex: A rare skin lesion

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ABSTRACT

Steatocystoma multiplex, a rare genetic condition causes the development of several hamartomatous abnormalities at the pilosebaceous duct junction (the unit that holds the hair follicles). This condition usually arises during the adolescent period. Offspring will surely develop the propensity to form cysts as this is an autosomal dominant disorder and can affect both sexes.

1. INTRODUCTION

Familial steatocystoma multiplex can occur due to mutations in genes encoding a protein named keratin 17 (K17) found in nails, hair follicles and sebaceous glands. Mutation in the same K17 gene can also lead to a variant of pachyonychia congenita, a disorder characterized by the presence of cysts, plantar keratoderma, and natal teeth. The sebaceous duct has an irregular lining leading up to the oil glands which Is believed to be the source of steatocystomas. Steatocystoma multiplex arises during adolescence as the child reaches puberty around this time and the increase in hormones acts upon the pilosebaceous unit. These cysts are mostly commonly seen over the chest, belly, upper arms, armpits, and face although they can arise anywhere on the body. In rare instances, these cysts are seen to arise throughout the body. While these cysts typically range in size from 2 to 20 mm, they can reach much higher diameters containing a yellowish-oily fluid, ranging from soft to firm in consistency with a semitranslucent gross appearance. Sometimes, these cysts show a central opening containing eruptive vellus hairs. Many different forms of steatocystoma multiplex have been documented, including localized, generalized, facial, acral, and suppurative forms. An uncommon, hereditary disorder known as isolated steatocystoma of the vulva or scrotum can also manifest and is seen typically in old age.

Case report

A 23-year old man was seen in the dermatology OPD with complaints of skin coloured raised lesions that began to appear 2 years ago on his chest. Gradually, these lesions spread to involve his torso, face, lower limbs and upper limbs. These skin-coloured lesions were raised, firm to hard to touch and were movable nodules. The clinical differential diagnosis included-lupus miliaris disseminates faciei, histioid hansens, subcutaneous xanthoma, steatocystoma multiplex or vellous hair cyst. Skin biopsies from the left shoulder, back and the left temporal region of the scalp were sent to the pathology department for histopathological examination. Microscopically, the epidermis demonstrated orthokeratosis and a dermal cyst lined with stratified squamous epithelium on one specimen from the back while the specimen from the scalp revealed a large cyst lacking the granular layer containing multiple sebaceous glands along the cyst wall. This led to a diagnosis of steatocystoma multiplex. The specimen from the shoulder revealed skin with dermis, periadnexal region and the subcutis displayed well-defined granulomas with numerous langhans type giant cells and rapid exophytic activity leading to the diagnosis of lupus miliaris disseminates faciei.

Discussion:

This condition was first characterized by Jamieson in 1873, however, it was named as ‘ steatocystoma multiplex’ by Pringle in 1899 [5]. This condition affects the pilosebaceous duct junction leading to hamartomatous abnormalities [2], with no specific sex predilection, typically arising in adolescents [6]. Although researchers have yet to pinpoint where these cysts originate from, several hypotheses show that they are either hamartomas, a type of dermoid cyst, or the product of naevoid sebaceous retention cysts [7]. Few other researchers link Hypohidrosis, hypothyroidism, hidradenitis suppurativa, hypotrichosis, ichthyosis, koilonychias, congenital pachyonychia, acrokeratosis verruciformis of Hopf, hypertrophic lichen planus, and a few other conditions to steatocystoma multiplex [2].

There have been some cases of familial steatocystoma multiplex studied linking it to mutations in the keratin 17 gene, namely in regions that are identical to those seen in PC-2 patients [8]. The 14 mutations found in patients with steatocystoma multiplex or pachyonychia congenita type 2 so far have all been localized to the K17 gene's helix initiation region (1A domain). Genomic mutations disrupt keratin intermediate filament network formation; the KR T17 gene lies on chromosome 17's long(q) arm, between locations 12 and 21 [1,9]. mutations in the keratin 17 gene, namely in regions that are identical to those seen in PC-2 patients [8]. The 14 mutations found in patients with steatocystoma multiplex or pachyonychia congenita type 2 so far have all been localized to the K17 gene's helix initiation region (1A domain). Genomic mutations disrupt keratin intermediate filament network formation; the KR T17 gene lies on chromosome 17's long(q) arm, between locations 12 and 21 [1,9].

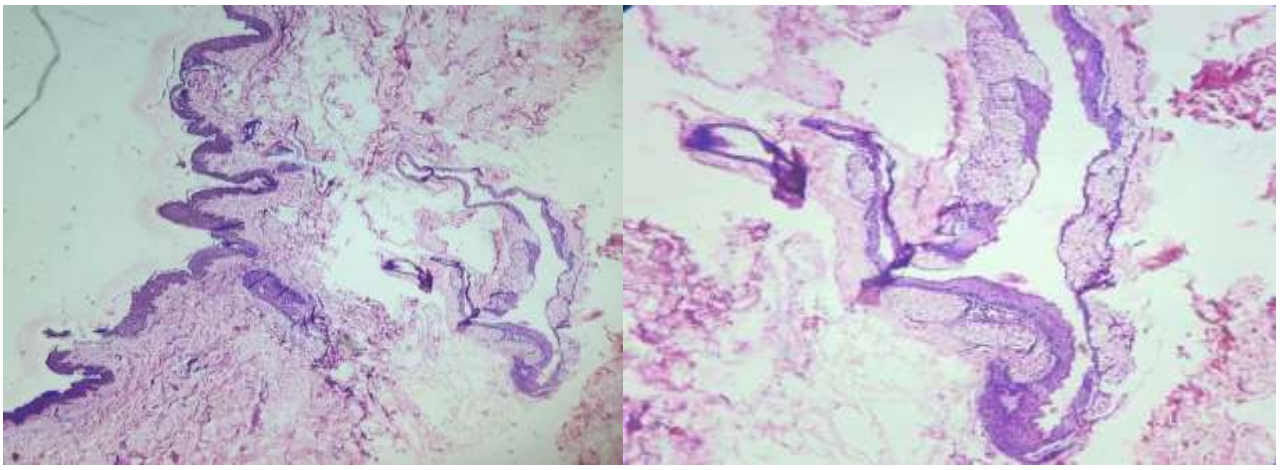

Image 1 – shows low power view showing skin lined by stratified squamous epitheium with underlying cyst.

Histopathology should always rule out eruptive vellus hair cysts, milia, trichilemmal cysts, tumors of the follicular infundibulum, and other important differential diagnoses. Vellus hair cysts and steatocystomas have a lot of commonalities. Cystic changes in the same pilosebaceous duct have been described in cases where the cysts exhibited characteristics of both steatocystoma multiplex and eruptive vellus hair cysts [13]. Histopathology of eruptive vellus hair cysts (EVHCs) reveals dermal cysts walled by stratified squamous epithelium with a granular layer. Inside the cysts are numerous vellus hair shafts that have been cut transversely and obliquely. Numerous eruptive vellus hair cysts are revealed by microscopic analysis of the expressed contents in a potassium hydroxide preparation. In most cases, the cyst wall will not include any sebaceous glands [13]. The stratified squamous epithelium lining the epidermoid cysts has a granular layer, and laminated keratin is visible inside the cyst lumen.

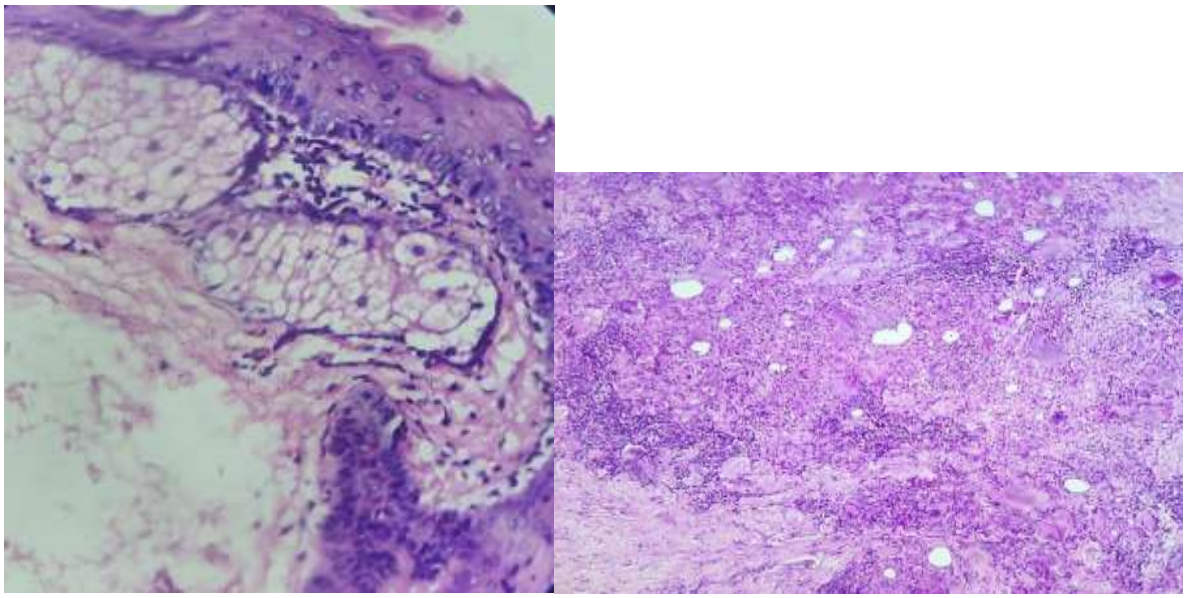

Image 2 & 3– show a high power view of cyst with sebaceous galnds.

Image 4 – asscociated lesion showing lupus miliaris disseminates faciei

As for mili, they reveal tiny cysts. A granular cell layer encases the milium, with an epithelial lining typically found in the dermal surface. Lamellated keratin is also revealed". Squamous epithelium devoid of a granular layer lines the inside of trichilemmal cysts, and cells around the cyst cavity enlarge to accommodate the homogeneous keratin that fills it. Although there may be a few vellus hairs in the cystic cavity, the hallmark histologic feature of steatocystomas is the presence of sebaceous lobules near the cystic wall. This wall is bordered with stratified squamous epithelium without a granular layer.

The expression patterns of keratin 10 (K10) and keratin 17 (K17) can vary in many cyst types, including epidermoid, trichilemmal, eruptive vellus hair, and Steatocystoma multiplex cysts. In contrast to the trichilemmal cysts and Steatocystoma multiplex, which exhibit the expression of both K10 and K17, the epidermoid cysts express K10 and the eruptive vellus hair cysts express only K17.

Surgical excision, carbon dioxide laser treatment, oral isotretinoin, and cryotherapy are all options for treating steatocystoma multiplex. However, a radiofrequency incision probe has recently been added to the arsenal, allowing for the creation of tiny incisions and the expression of cyst contents and even the cyst wall. Treatment with the Er:yag laser, followed by the application of topical tetracycline ointment, has demonstrated good cosmetic outcomes [6], and it has the advantage of creating a bloodless field that cannot be achieved with surgical blades [15].

Conclusion:

The final diagnosis of steatocystoma multiplex is always done after histopathological examination. The cysts appear histologically as well-encapsulated masses with multiple layers of elaborately folded epithelial cell lining along the cyst walls with flattened sebaceous gland globules either within or near the cyst walls. When a patient presents with more than one asymptomatic intradermal cyst, skin biopsies should be examined for steatocystoma multiplex as a possible diagnosis. It is possible that the psychological effects of a sickness can be lessened with early diagnosis and suitable counselling, however, poor treatment outcomes characterize this uncommon clinical condition.

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