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CASE SERIES

Clinical and Diagnostic Features of Steatocystoma Multiplex: A Case Series of a Rare Condition

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Abstract

Steatocystoma Multiplex (SC) is a rare genetic disorder characterized by cystic lesions within the dermis, originating from sebaceous glands. Despite established clinical and morphological criteria, diagnosis can be challenging due to the condition's rarity. This study reviews six cases diagnosed in a medical institution in Uttarakhand. Most patients presented with symptom onset in adolescence. Although family history was generally absent, associations with pachyonychia congenita (PC) suggest shared genetic factors. Patients presented with painful and itchy cysts. Histopathological findings consistently revealed multiple thin-walled cysts with sebaceous glands. Immunohistochemistry (IHC) for cytokeratin and EMA confirmed the diagnosis. The study highlights the significance of early recognition, thorough clinical assessment, and a multidisciplinary approach to management, emphasizing their crucial role in ensuring effective care and achieving improved outcomes in SC.

Key Words

Steotocystoma, Sebaceous, Cysts, Dermis

Introduction

Steatocystoma multiplex (SC) is a rare genetic disorder characterized by hamartomatous malformations at the junction of the pilosebaceous duct. It presents as encapsulated cystic lesions within the dermis, often accompanied by adjacent sebaceous glands.[1] This condition represents a rare, benign adnexal tumor marked by the formation of multiple cysts derived from sebaceous glands. First described by Pringle in 1899, the clinical profile and morphological criteria for SC are well defined, but diagnosis can still be challenging due to its rarity and varied presentation. SC typically arises in adolescence or early adulthood. It may be associated with other conditions such as pachyonychia congenita.[2, 3] When inflammation is present, mimicking hidradenitis, the condition is termed steatocystoma multiplex suppurativa, a rarely reported variant.[4] Here, we present the clinical, morphological, cytochemical, and immunocytochemical features of four cases of SC and two cases of steatocystoma multiplex suppurativa, emphasizing the diagnostic challenges encountered.

Materials and Methods

All cases of steatocystoma complex diagnosed in the Department of Pathology, Veer Chandra Singh Garhwali Government Medical Science and Research Institute, Srinagar, Uttarakhand, from 2016 to 2023 were included in the study. A total of six cases were retrieved. The clinical history and other details were taken from the case files. (Table No. 1). A clinical image is presented in Figure 2.

A histopathological examination of excised cysts was performed. Hematoxylin and eosin (H&E) staining was available for all cases. (Table No. 2)(Figure 1)

Results

Clinical and histopathological profiles of our 6 cases are summarized in Tables 1 and 2.

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Table 1: Clinical Features and Demographic Data

Features	Case I	Case II	Case III	Case IV	Case V	Case VI
Age/Sex	21 years/M	30 years/F	18 years/M	20 years/F	19 years/M	19 years/M
Onset	Adolescence	Early adulthood	Adolescence	Adolescence	Adolescence	Adolescence
Family History	-	-	-	-	-	+
Associated Conditions	-	Pachyonychia	-	-	Pachyonychia	Pachyonychia
Distribution	Trunk, arms	Face, neck	Trunk, legs	Chest, back	Abdomen, arms	Abdomen, arms
Symptoms	Asymptomatic	Painful cysts	Asymptomatic	Itchy cysts	Asymptomatic	Asymptomatic

Table 2 : Histopathological and Immunohistochemical Features.

Case	Histopathology Summary	Cytokeratin	EMA
Case I	Multiple cysts with sebaceous material, granulomatous inflammation	Positive	Positive
Case II	Cystic structures, sebaceous glands, keratin debris, mixed inflammatory infiltrate	Not done	Not done
Case III	Numerous cysts, sebaceous glands, hair follicle remnants, oily substance, pericystic fibrosis	Positive	Positive
Case IV	Multiple cystic lesions, sebaceous glands, lipid-rich material, calcification, mild chronic inflammation	Not done	Not done
Case V	Several cysts, sebaceous material, early rupture, granulomatous inflammatory response, fibrosis	Not done	Not done
Case VI	Cysts with sebaceous material, partial rupture, histiocytic reaction, giant cell formation	Not done	Not done

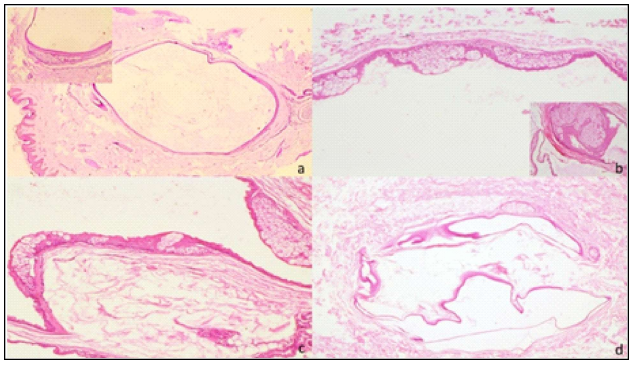

Figure 1: (a) Tissue lined by stratified squamous epithelium with the dermis showing a complete (H&E, 100X)). The inset highlights a part of a cyst with a sebaceous gland. (H&E, 400X). (b) A part of the cyst revealing multiple sebaceous glands within the cyst lining (H&E, 400X). The inset shows a complete cyst with a large sebaceous gland within. (H&E, 400X). (c) Cyst and its infoldings with sebaceous glands present in the lining (H&E, 100X). (d) Irregular cyst with two sebaceous glands in proximity (H&E, 100X).

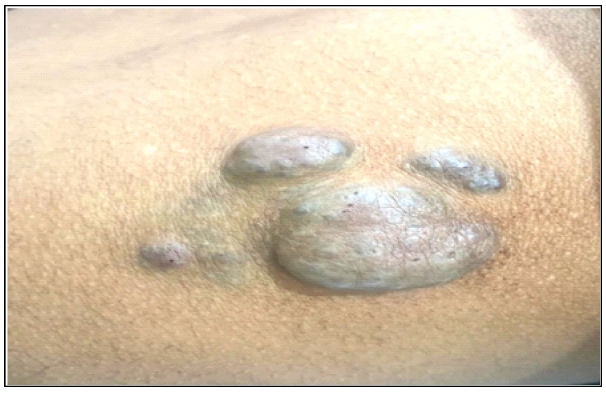

Figure 2: Shows Multiple Cutaneous Papulo-nodular Lesions.

The patients in the six cases are predominantly young, ranging from 18 to 30 years. The majority of cases (five out of six) experienced the onset of symptoms during adolescence, accounting for approximately 83% of the cases, while one case (17%) had an onset in early adulthood. There is a notable absence of significant family

history in five cases (83%), with only one case (17%) reporting a positive family history.

Pachyonychia is an associated condition in three cases (50%), highlighting its relevance in the presentation of symptoms. The distribution of lesions varies: two cases (33%) have lesions on the abdomen and arms, one case (17%) on the trunk and arms, one case (17%) on the face and neck, one case (17%) on the trunk and legs, and one case (17%) on the chest and back.

Regarding symptoms, the majority of cases (four out of six, 67%) are asymptomatic, with one case (17%) experiencing painful cysts and another case (17%) reporting itchy cysts. Histopathology reveals multiple thin-walled cysts lined by stratified squamous epithelium and sebaceous glands, containing sebaceous material, with occasional ruptures, granulomatous inflammation, fibrosis, calcification, and secondary infections. Two out of the five cases (40%) were diagnosed as Steatocystoma Complex Suppurativa, characterized by multiple inflamed cysts diagnosed as Steatocystoma Complex Suppurativa.

Discussion

Steatocystoma complex is a benign adnexal tumor arising from the sebaceous glands, typically presenting as multiple cysts.[5,6] Histopathological examination revealed consistent features across the cases, including numerous thin-walled cysts lined by stratified squamous epithelium and sebaceous glands. The cysts contained sebaceous material and showed evidence of occasional rupture, granulomatous inflammation, fibrosis, and, in some cases, calcification and secondary infections. These findings are typical of SC and help differentiate it from other skin conditions with similar clinical presentations.[7,8] The diagnosis is primarily based on clinical and histopathological findings. IHC for cytokeratin and EMA can confirm the diagnosis. While SC is usually asymptomatic, cysts can become painful or itchy, and associations with genetic conditions like pachyonychia congenita (PC) may be present.In the present study, PC was found to be associated in 50% of the cases.The association between SC and PC suggests shared genetic mutations affecting keratin expression and epithelial tissue homeostasis. PC arises from mutations in keratin genes, causing thickened nails and skin changes, while SC results from sebaceous gland dysfunction, leading to cyst formation. Genetic overlap may influence both conditions. [9] Earlydiagnosis and appropriate management are essential to prevent complications and improve patient quality of life.[10]

Conclusion

Steatocystoma complex, though rare, should be considered in patients presenting with multiple cysts, especially with a family history or associated conditions like pachyonychia congenita. A multidisciplinary approach involving dermatology, pathology, and genetics ensures comprehensive care for individuals affected by SC. Awareness of this condition can aid in early detection and management, providing better patient outcomes.

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