Case Report

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Dermatology Section

A Sporadic Case of Late-onset Vulvar Steatocystoma Multiplex

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ABSTRACT

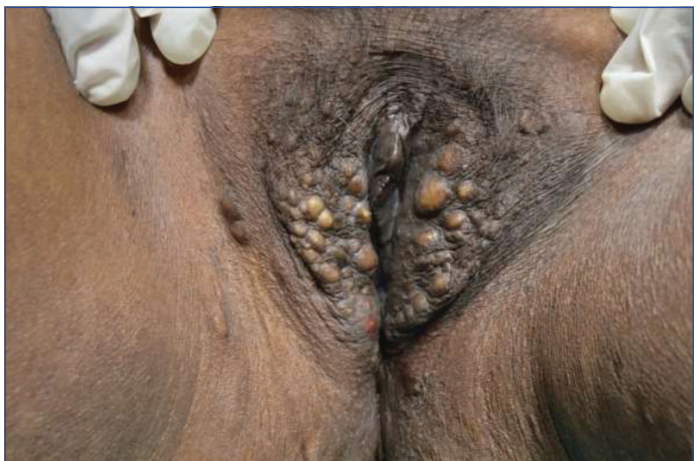
Steatocystoma multiplex is a benign disorder involving the pilosebaceous unit, characterised by the presence of numerous cutaneous cystic lesions in the dermis. While cases with random occurrence are commonly observed, the condition can also be transmitted genetically in an autosomal dominant fashion. The lesions are frequently seen in body parts with a high number of pilosebaceous units including the axillae, scalp, trunk, thighs, arms, and less frequently occurring on the genitals and breasts. Hereby, the authors present a case report of a 48-year-old female who presented to the Outpatient Department of Dermatology with multiple asymptomatic swellings on her vulva since nine years. On physical examination, the swellings were noted to be calcified nodules. These nodules were excised and sent for histopathological analysis that revealed the diagnosis of steatocystoma multiplex. Steatocystoma multiplex can occur as sporadic, familial, or as a part of a syndrome. Although the lesions themselves are symptomless, their presence in the genital region makes walking difficult, thereby hampering routine work and adversely affecting the quality of life of the patient. There is also a need to educate patients regarding its benign nature and possible treatment options. Hence, the present case report aimes to emphasise the importance of its early identification, diagnosis, and to raise awareness among general practitioners.

Keywords: Appendageal neoplasm, Cyst, Histopathology, Subcutaneous nodule

CASE REPORT

A 48-year-old female presented to the Dermatology Outpatient Department (OPD) with complaints of multiple raised lesions over her genitalia, since nine years. The patient had difficulty in walking, and these lesions were not associated with pain, discharge, itching, or burning sensation. The lesions started as small swellings and gradually increased in size over a nine-year duration to the current size. The patient received Ayurvedic treatment for the same with no improvement in the lesions. She did not provide any affirmative history of a family member having similar complaints.

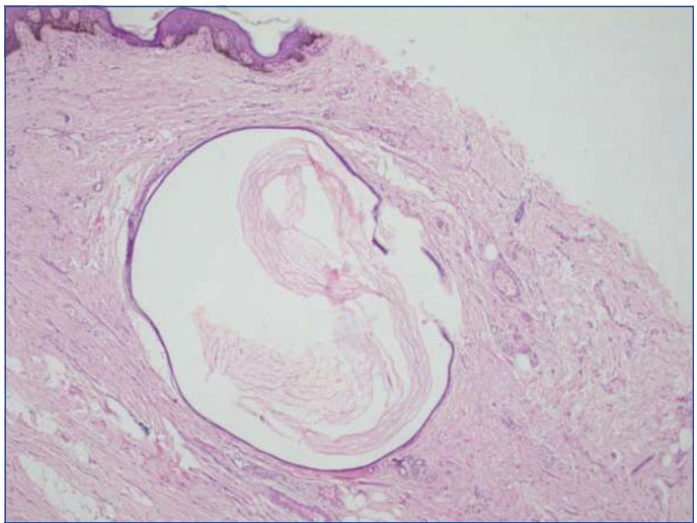
Physical examination revealed the presence of multiple, non tender, skin-coloured to whitish-yellow cystic nodules with sizes ranging from 0.1×0.1 cm to 0.5×0.5 cm over the inner aspect of labia majora [Table/Fig-1].



[Table/Fig-1]: Multiple skin-coloured and whitish-yellow calcified nodulocystic lesions on labia majora.

There were no other lesions present elsewhere on the body. The skin on her palms and soles appeared normal. Hair strands appeared normal. She had no nail dystrophy and had normal dentition. The routine laboratory examination results were within normal limits. The patient was explained and counselled about her condition.

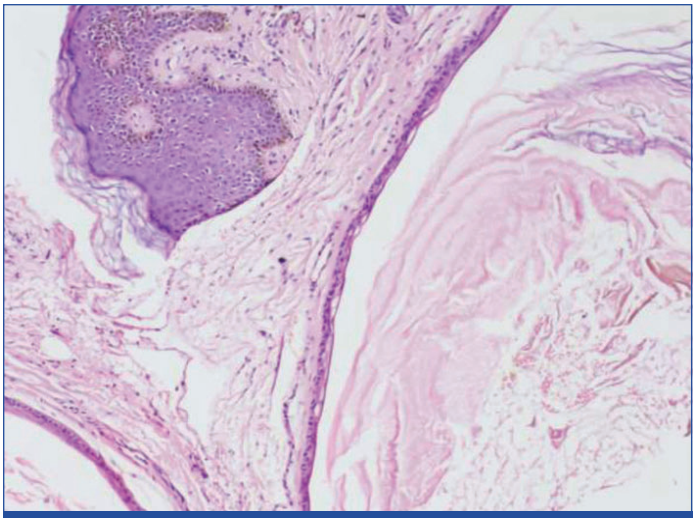
The patient refused conservative management because of her disappointing experience with Ayurvedic therapy in the past and she expected a rapid outcome. She opted for surgical excision of the lesions, for which she was referred to the Department of Surgery where the biopsy of nodules was planned. Skin biopsy and histopathological examination {Haematoxylin and Eosin (H&E, staining)} revealed a stratified squamous epithelium with an absent granular layer. Sebaceous glands were present in the cyst wall, and there was presence of a homogeneous, undulating eosinophilic cuticle on the luminal side of the cyst wall [Table/Fig-2,3]. Based-on the clinical and histopathological findings, a diagnosis of steatocystoma multiplex was made.



[Table/Fig-2]: A simple cystic cavity occupying the dermis lined by a squamous epithelium with focal areas of disruption and filled with lamellated keratin (H&E, 40×).

DISCUSSION

Steatocystoma is a variant of a tumour that develops from sebaceous follicles and sebaceous glands. It is characterised by the presence of nevoid sebaceous ducts [1] and can be classified as steatocystoma simplex and steatocystoma multiplex. Brownstein first reported



[Table/Fig-3]: The cystic epithelium composed of two layers of epithelial cells and an inner epithelial lining showing eosinophilic cuticle (H&E, 100×).

steatocystoma simplex in 1982 [2], and steatocystoma multiplex was first described by Jamieson WA [3]. Clinically, Steatocystoma Multiplex manifests as multiple, round, smooth, firm, mobile cystic papules and nodules that are typically asymptomatic. The fluid in lesions of steatocystoma multiplex is odourless, oily, clear or opaque, milky or yellow. The overlying epidermal skin appears normal and without a central punctum [4].

The occurrence of vulvar steatocystoma multiplex is uncommon. After a diligent search, the present case appears to be the first case report of vulvar steatocystoma from India [Table/Fig-4] [5-10].

follicular infundibulum tumours, milia, neurofibromatosis, lipoma, and xanthomatosis. Diagnosis is confirmed by histopathological features. Dermal cysts with vellus hair shafts in the lesion are typical features of EVHCs. Epidermoid cysts are lined by stratified squamous epithelium with a granular layer, and the lumen contains laminated keratin. The milium has a granular layer and is predominantly located in the superficial dermis. The follicular infundibulum tumours consist of dermal tumour islands that are connected to the epidermis. A distinctive histopathological feature of steatocystoma multiplex is the existence of sebaceous lobules near the cystic wall, which is lined by stratified squamous epithelium without a granular layer. The epidermis overlying the sebum-containing cysts is apparently normal and without a punctum. The histopathological findings of this case showed the characteristics of steatocystoma multiplex. Although any part of the body might be affected, reports of sebaceous cystic lesions of the vulva are very few [13,14]. In the present case, histological features and the lack of other associated symptoms aided in the diagnosis of sporadic vulvar steatocystoma multiplex.

Steatocystoma multiplex is usually asymptomatic unless it becomes infected; however, patients are affected psychologically. Despite its benign nature, the treatment of steatocystoma multiplex is a challenge. Management is often influenced by personal experience, nature of lesions, and literature. Literature assessing vulvar steatocystoma multiplex has documented management using isotretinoin and surgical excision. Prognosis was questionable as reported recurrence ranging from 0-25% was observed in the literature [15].

Name of the author	Place of the study	Year	Patient age	Disease duration	Histopathology	Family history	Treatment	Prognosis
Lewis B [5]	London	1948	37	Unknown	Non characteristic	Familial	Excision	Unknown
Puech I et al., [6]	France	2000	40	20 years	Unknown	Familial	Isotretinoin without success	Partial vulvectomy
Rongioletti F et al., [7]	Italy	2002	81	27 years	Steatocystoma	Sporadic	No treatment	Unknown
			60	3 months	Thin-walled epidermal cyst demonstrating features consistent with steatocystoma	Sporadic	Some lesions were excised	Unknown
Park J et al., [8]	South Korea	2014	82	6 months	Sebaceous gland on the cyst wall	Sporadic	No treatment	Unknown
Kartal SP et al., [9]	Turkey	2016	32	1 year	Sebaceous gland on the cyst wall	Sporadic	Surgical excision	Four years follow-up showed new lesions to be located on previously untreated areas
Kerrouch H et al., [10]	Morocco	2023	24	5 years	Stratified squamous epithelium without granular layer	Sporadic	Excision	No recurrence during eight months of follow-up
Present case	India	2024	48	9 years	Steatocystoma	Sporadic	Surgical excision	Unknown

[Table/Fig-4]: Reported cases of vulvar steatocystoma multiplex [5-10].

While sporadic cases of vulvar steatocystoma multiplex can occur, there is evidence in the literature highlighting its familial occurrence [11]. In the present case, the nature of lesions was sporadic. The age of occurrence varied from 32-82 years. The pathogenesis of steatocystoma multiplex is hypothesised to involve mutations in the keratin 17 (KRT17) gene, which is expressed in sebaceous glands and hair follicles. A similar mutation has been identified in pachyonychia congenita type 2 (Jackson-Lawler syndrome), a disorder that is transmitted in an autosomal dominant fashion and characterised by palmoplantar keratoderma, follicular keratoses, epidermal inclusion cysts, oral leukokeratoses, and nail dystrophy. Pili torti, vellus hair cysts, steatocystoma multiplex, and natal teeth are also sometimes found in pachyonychia congenita type 2 [12].

The differential diagnosis of the disease are epidermal inclusion cyst, Eruptive Vellus Hair Cysts (EVHC), trichilemmal cysts, and

CONCLUSION(S)

Being able to detect numerous asymptomatic dermal cysts and identify the distinct pattern of multiplication with gradual expansion observed in steatocystoma multiplex enables early education regarding the non malignant nature of this condition. A concrete understanding of the available treatment options based on the location of the lesions, along with their associated risks and advantages, is pivotal in effectively addressing the medical and psychosocial consequences associated with steatocystoma multiplex. Patients should be counselled that viable treatment choices require a substantial amount of time and frequently lead to scarring, with recurrence being common. Thoroughly addressing unrealistic expectations and the risks of unnecessary treatment is crucial when engaging with this patient group, given their inclination towards seeking unachievable results.

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