Awrying Facial Steatocystoma Multiplex

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ABSTRACT

Steatocystoma multiplex is a hamartomatous condition of pilosebaceous apparatus characterized by the development of numerous benign sebum-containing dermal cysts. Facial steatocystoma multiplex is often confused with acne vulgaris because of the unfamiliarity with the former entity. In literature there are only about six cases reported in the past ten years and thereby we report such a case in a 35 year old male patient, attempting to throw light on this entity.

Facial steatocystoma multiplex is not an uncommon variant as it is thought to be, rather misdiagnosed and hence underreported. It should be considered in differential diagnosis of recalcitrant acne.

Key Words: *Steatocystoma multiplex, keratin 17, eruptive vellus hair cysts, recalcitrant acne*

INTRODUCTION

Steatocystoma multiplex (SM) is a hamartomatous malformation of the pilosebaceous unit characterized by the development of numerous sebum containing dermal cysts. We hereby report a case of facial steatocystoma multiplex as predominant or isolated involvement of face is rare.[1, 2, 3, 4] Also this case was misdiagnosed since 20 years and was treated for acne vulgaris with little or no response. In literature there are only about six cases reported in the past ten years, hence here we have made an attempt to write short review of isolated facial steatocystoma cases to help us in better understanding of this variant.

CASE REPORT

A 35-year old male patient came with complaints of asymptomatic numerous papules over the face since 20 years. He gave history of slow growth of few with pain resulting in spontaneous rupture and scar formation. For the above he has received multiple antibiotics as treatment for acne. No family history was present in this case.

On examination, multiple yellowish papules were present in both temple regions and also on right cheek. Multiple skin coloured, nontender, mobile, cystic nodules with no visible punctum were present on both cheeks (Fig. 1a and 1b). Excision biopsy of a nodule was carried out (Fig. 2). The reported finding was a cyst cavity lined by stratified squamous epithelium without granular layer and eosinophilic cuticle on the luminal side (Fig. 3). Hence diagnosis of facial steatocystoma multiplex was made and patient was started on tablet isotretion.

DISCUSSION

Jamieson in 1873 first described the entity and was later named by pringle in 1899.[2,3,5] It is also known as steatocystomatosis, sebocystomatosis and epidermal polycystic disease.[2,6] The etiology of this condition in uncertain. Autosomal dominant inheritance due to keratin 17 gene mutation is seen in steatocystoma multiplex cases, although sporadic occurrence is much more common.[3,7,8] Keratin 17 is an intermediate filament protein and is highly expressed in sebaceous glands and outer root sheath of hair follicles. Hence the mutation affects the sebaceous glands resulting in cysts. Many researchers believe that steatocystoma multiplex is a variant of pachonychia congenital type 2 caused by mutation of K6b and K17 genes.[3,7,8]

The steatocystoma simplex variant is non-hereditary and trauma or infection may play a role in pathogenesis of this condition.[6] Most cases have onset in adolescence because of peak in sebum secretion even though rare cases are described in newborns and old ages.[1] Both sexes are affected equally.[3]

Clinically it presents as solitary (simplex), or multiple (multiplex), yellowish or skin coloured cystic papules and nodules usually asymptomatic but inflammation does occur resulting in scar formation (steatocystoma multiplex suppurativa).[3] The later form resembles acne conglobata when present extensively.[6] It is reported to occur at any site including mucosa, palms and soles but predominant involvement of scalp, axilla, trunk and proximal extremities is seen.[5]

Many variants are described including, simplex; multiplex; suppurativa; acral; localized; generalized and facial.[3] Involvement of the scalp and face together is often seen and is divided into following types; facial papular variant, sebocystomatosis and cysts located exclusively on the scalp.[2] Isolated facial steatocystoma multiplex is rare as it is the simplex variant that is common on the face unlike in our case.[2] There are only 6 cases reported since 2005 as per our knowledge and shown in Table 1.

The reported associations of steatocystoma are eruptive vellus hair cysts, ectodermal dysplasia, icthyosis,

acrokeratosis verruciformis, rheumatoid arthritis, hypohidrosis and hypothyroidism.[2, 3]

The differential considered in our case was acne vulgaris. The differentiating points were absence of comedones, cystic nature and presence of true cyst on histopathology and its persistent nature. Varshney M, et al also reported a case of steatocystoma localized to head and neck, which was confused with acne.[9] Steatocystoma multiplex is not an uncommon disorder in the Indian subcontinent and hence should always be kept in mind when recalcitrant acne is encountered.[10]

Histologically the lesion is located in mid dermis. The cyst wall is wavy and is composed and of stratified squamous epithelium with sebaceous glands in its wall.[1,2] Eosinophilic cuticle on the luminal side of this wall is often present with keratin or hair or sebum in lumen. It may undergo calcification.[6,10]

Keratin expression can also be used to differentiate this condition from other mimicking ones. The epidermoid cysts express K10, the eruptive vellus hair cysts express K17, whereas the trichilemmal cysts and steatocystoma multiplex show the expression of both K10 and K17.[3]

The electron microscopy findings of steatocystoma multiplex are one pilary unit which continuously produces the vellus hairs trapped in the cystic cavity, connected to the epidermis by aepithelial cord (the remnant of the follicular infundibulum). A lumen which is partly present in a few areas of the cord is filled with the cellular debris. Hence it is found to be a nevoid sebaceous duct and a sebaceous gland tumour. [3]

Non-invasive treatments are only temporary such as oral isotretinoin, tetracyclines, intralesional steroids and aspiration. CO2 laser, ER: YAG laser are newer modalities but surgical excision with removal in toto is still the better choice.[2,3,5,10,11]

Table 1: Isolated facial steatocystoma cases over the last 10 years

S. No	Age	Sex	Part of face	Duration	Clinical features	Other feature	Treatment
1.	31	M	Forehead, temple, cheeks, jaw	Puberty	well defined smooth surfaced round, skin coloured, firm papules	Family history	Surgical excision
2.	78	F	Face	Few month	Multiple yellowish nodules	-	Curettage and CO2 laser therapy
3.	40	F	Eyelids, forehead and neck	Puberty	Skin colored cystic nodules	-	Curettage and CO2 laser therapy
4.	47	M	Forehead and cheeks	1 year	Soft, skin colored papules and nodules with a smooth surface and explicit borders	-	Er: YAG laser
5.	70	M	B/L malar region of face	2 years	Yellow to skin colored, soft movable cystic masses, ranging from 10 mm to 20 mm in size with no punctum	-	Surgical excision
6.	30	M	Forehead, temple, post auricular neck	Puberty	20 well defined smooth surfaced round, skin coloured, firm papules	Pilar cyst and preauricular sinus	

CONCLUSION

Steatocystoma multiplex is not uncommon as it is thought to be, rather misdiagnosed and hence underreported. As it is a major cosmetic problem it causes psychosocial morbidity. Hence we recommend proper screening of patients with recalcitrant facial nodules.

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