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Steatocystoma multiplex –presenting as hyperpigmented nodules.

KEYWORDS: Steatocystoma multiplex, sebum

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ABSTRACT *Steatocystoma multiplex (SM) is a rare disorder of the pilosebaceous unit characterized by the eruption of numerous sebum-containing dermal cysts. These are usually confined to presternal area, trunk, neck and proximal upper extremities. Localised and segmental variants have been occasionally described. However, pigmentation or itching has not been commonly described. We present a case with regular distribution but associated with itching and pigmentation.*

Introduction:
Steatocystoma multiplex (SM) is a rare disorder of the pilosebaceous unit characterized by the eruption of numerous sebum-containing dermal cysts. Most cases are sporadic; however, familial cases with autosomal dominant inheritance have also been described.[1]

Usually begining in adolescence or early adult life,[2,4] due to the action of androgens. It may be inherited as an autosomal dominant trait, although most cases have no family history.[5] Both sexes are affected equally.

Histologically it shows a mixture of a keratinizing epithelium and sebaceous lobules attached to the epidermis by a thin epidermal strand.[5]

Case Report:
A 30 year male patient presented to our OPD with multiple, pigmented lesions of few months duration with occasional intense itching. On examination, nodular, elongated, multiple lesions lying singly and in clusters were found on the front of chest, upper abdomen, back, flanks and neck. There are no lesions elsewhere. Most lesions show a greenish black appearance. One lesion on the neck is grain sized and yellowish in color.

General health is well preserved and there are no other systemic or cutaneous associations.

An excisional biopsy of a truncal lesion revealed a cyst lined by stratified squamous epithelium with peripheral focally palisading basal layer. Cyst cavity showed amorphous keratinous material and multiple vellus hair shafts and perivascular lympho-histiocytic infiltrate. These features are consistent with a diagnosis of steatocystoma multiplex.

Figures:

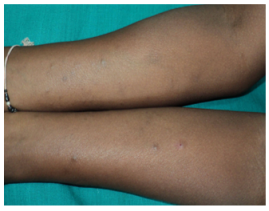

Fig1: Lesions on forearms

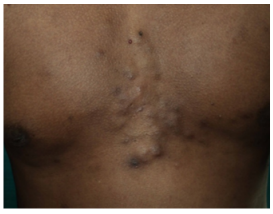

Fig 2: Multiple cystic lesions in the presternal area

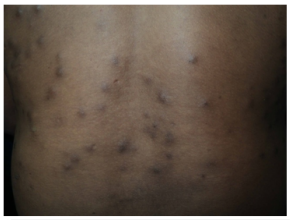

Fig 3: Intensely pigmented cystic nodules on the back

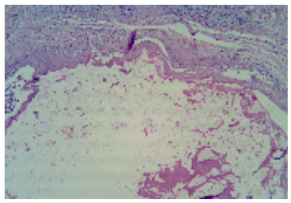

Fig 4: Histopathology showing cyst wall lined by stratified squamous epithelium

Discussion:
The aetiopathogenesis of SM remains elusive but there are several hypotheses to explain its cause, such as it originating from sebaceous retention cysts, or representing a naevoid malformation of the pilosebaceous duct. Surgical removal and decapitation of the cyst surface are the most frequently applied therapeutic options. Mutations in the keratin 17 (K17) gene (KRT17) have been repeatedly reported for familial cases of SM.

Kligman and Kirchbaum postulated that pluripotential ectodermal cells retain the embryonic capacity to form appendages or naevi rather than retention or inclusion cyst.[3,4]CK is a useful marker to evaluate the origin of epithelial tumours.[6]

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