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Case Report

Localised Form of Steatocystoma Multiplex: A Case Report

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Abstract: Steatocystoma multiplex is a rare, benign, autosomal dominant disorder. These lesions have been described as hamartomatous malformation of pilosebeceous duct junction (1). We report, a case of 36-year-old man who presented with swelling over both the axilla since 10 years. There was a family history of similar lesion in mother. Clinical diagnosis was epidermal cyst. Histopathogical examination revealed Steatocystoma multiplex.
Keywords: Steatocystoma multiplex, Pachonychiacongenita, pilosebaceousduct.

INTRODUCTION

Steatocystoma multiplex is a rare, autosomally dominant, genetic disorder characterised by multiple asymptomatic, slow growing, cystic lesions 2-3 cm in diameter principally seen on the axilla, groin trunk, scrotum, proximal extremities and rarely located in face and scalp (Belinchon, I. *et al.,* 1995). In male common site affected is sternum.Steatocystoma multiplex subgrouped into localised, generalised,facial, acral and suppurative types.Steatocystoma multiplex tends to occur during adolescence or early adult life but it is also described in persons in sixth decade of life (Rongioletti, F. *et al.,* 2002). Overlying epidermis is normal without a punctum but comedons are associated features (Cho, S. *et al.,* 2002). Lesions present as dome shaped nodules,translucent to yellow in colour. The contents may be either clear, oily liquid or yellow cheesy or creamy material.

Clinical Synopsis

A 36-year-old man presented with multiple asymptomatic round to oval well defined yellow to skin coloured 0.5 to 1 cm diameter firm papules in both the axilla; which have been present for about 10 years. There were no similar lesions on the other parts of the body. His mother also had the similar history of swelling within the axilla. There was no history of trauma, erythema or infection. On examination right axilla had a single lesion measuring 1x 0.5 cm and left axilla had two well defined lesions measuring 0.5 x 0.3 cm and 0.3 x0.2 cm size respectively and it was nontender. Clinical diagnosis made was epidermal cyst for which excision biopsy of both lesions were done. Macroscopically both were skin covered soft tissue pieces with a yellowish lesion measuring 1x 1x 0.5 cm and 0.5 , 0.8x 0.5cm. Cut section showed cheesy material in both. Microscopy showed empty cysts containing serpiginous walls lined by thin squamous epithelium with minimal granular layer and sebaceous glands were seen adjacent to the cyst wall.

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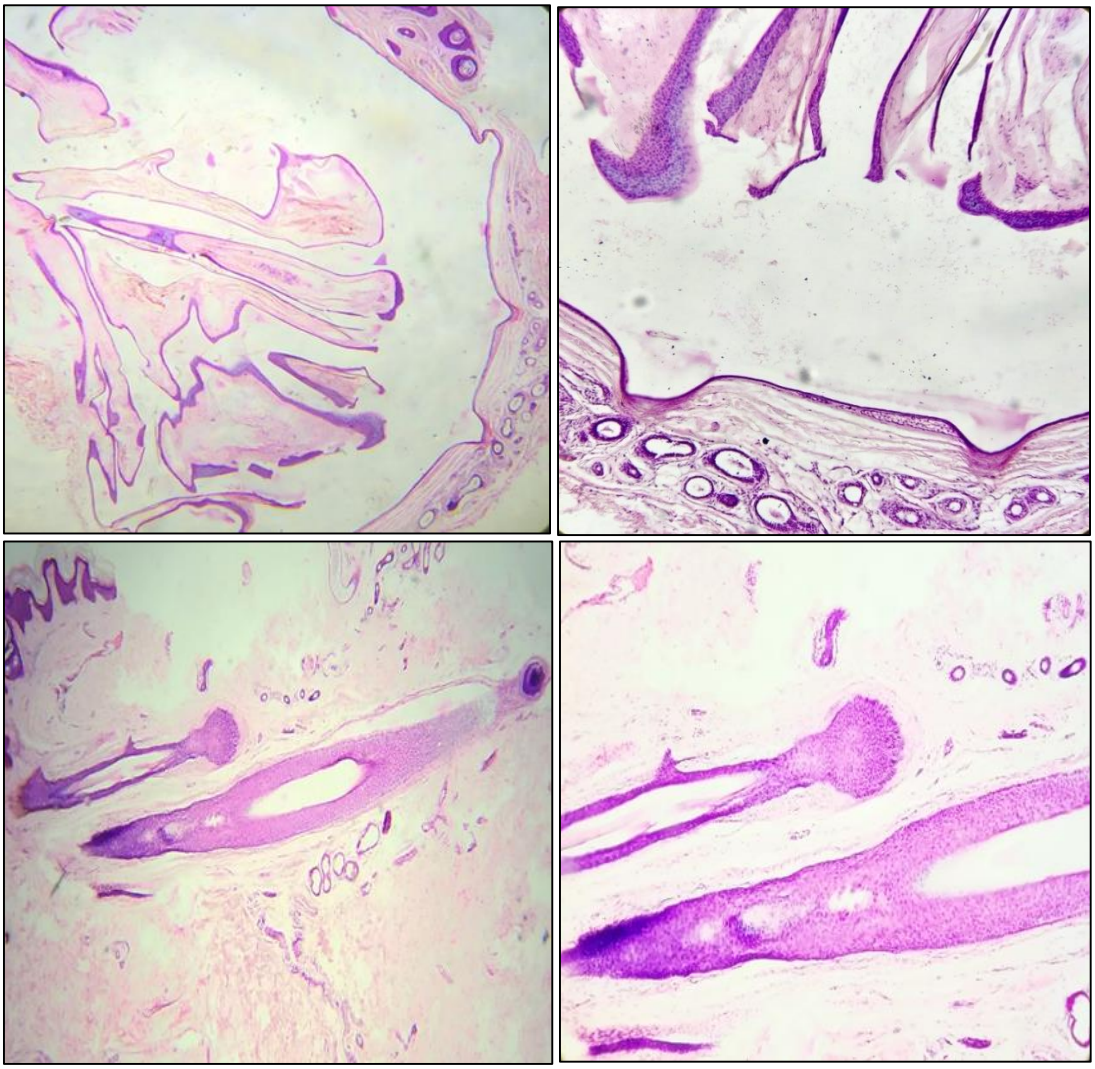

Fig: Cystwall lined by flattened stratified squamous epithelium with absence of granular layer and vellus hair.

DISCUSSION

Steatocystoma multiplex was first described by Jamieson in 1873 and the term was coined by Pringle in 1899. It is a hamartomatous malformation of the pilosebaceous duct junction. The exact origin of the cysts is still unknown but multiple theories suggest that they results from sebaceous retention cyst of nevoid nature or hamartomatous or they are a variety of dermoid cysts. It may be associated with Pachonychia congenita, ichthyosis, koilonychias, hypertrophic lichen planus, hypohydrosis, hypothyroidism and hidradenitis suppurativa (Jeong, S. *et al.,* 2009).

The familial steatocytoma multiplex is associated with mutation in keratin 17 gene which is identical to the mutation seen in patients with pachyonychia congenita type II. Steatocystoma multiplex and pachyonychia congenita type II are associated with 14 types of mutations and all are localised to the helix initiation domain (1A domain) of the k17 gene. The KR T17 gene is located on long arm of chromosome 17, between position 12 and 21 and the mutation in this gene interfere with the assembly of keratin intermediate network filament.

Xiuying W studied the keratin 17 mutation in the cDNA of the cystic tissue of 5 patients of Steatocystoma multiplex and in the peripheral blood of 25 patients with Steatocystoma multiplex from Steatocystoma multiplex family, by direct sequencing of RTPCR products, nested PCR and restricted fragment length polymorphism and he revealed the R94Cmutation the keratin 17 gene (LIU, Q. *et al.,* 2015).

The differential diagnosis includes epidermal inclusion cyst,milia,trichilemmalcysts and eruptive villous haircysts.Steatocystoma multiplex and villous hair cysts are closely related to each other because of their cause is related to cystic changes in pilosebaceous ducts.On microscopy eruptive villous hair cyst shows dermal cysts that are lined by stratified squamous epithelium with granular layer and also contain obliquely and transversely sectioned villous hair shaft. On microscopic examination of expressed contents in a potassium hydroxide preparation shows numerous

villous hair is mainly the eruptive villous hilar cyst. Milia shows very small cysts located in superficial dermis and it has epithelial lining with a granular layer. The trichilemmal cysts are lined by squamous epithelium without granular layer and the cells are swollen close to the cystic cavity, which is filled with homogenous keratin. But the characteristic finding in Steatocystoma multiplex is presence of sebaceous lobules close to the cyst wall.

On electronmicroscopy,Steatocystoma multiplex found to be a nevoid duct and a sebaceous gland tumour. Piliary units which continuously causing villous hair which are trapped in cystic cavity or in piliary canal. Steatocystoma multiplex may be connected to the epidermis by epithelial cord, the remnant of follicular infundibulum which is a solid strand which contain sebocytes or sebaceous lobule like structures. A lumen is present in the cord is filled with cellular debris of keratinocytes,corneocytes,sebocytes or trapped hair.

Various modalities of treatment includes surgery, Co_2 laser therapy, cryotherapy and isotretinoin. Recent treatment includes the use of radiofrequency incision probe to make mini incision for expressing the contents of the cyst. It has the advantage of bloodless field (Koley, S. *et al.,* 2010).

To conclude, present case highlights the importance of appropriate diagnosis of lesion by histopathology and its differentiation from clinical mimickers for optimal treatment of the patient.It also projects the familial nature of the lesion and the occurance of vellus hair in Steatocystoma multiplex.

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