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ABSTRACT

Introduction: Steatocystoma, an uncommon cutaneous disorder, initially thought to be a sebaceous or retention cyst is recognized as a hamartomatous malformation of the pilosebaceous duct junction characterized by the development of numerous sebum containing dermal cysts with sebaceous glands in the cyst walls.

Case Report: A 70-year-old male patient presented with asymptomatic dark patches, bilaterally on malar region. Surgical excision was done under local anesthesia. Histopathology confirmed steatocystoma.

Conclusion: Steatocystoma is a cosmetic problem and a lifelong condition. Steatocystoma multiplex should be considered as a spectrum with different variations in anatomical distribution which can be treated successfully.
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Keywords: Steatocystoma Multiplex, Face, Malar region, Yellow Cutaneous cyst, Hereditary

INTRODUCTION

Steatocystoma multiplex was first described by Jamieson in 1873 and the name was coined by Pringle in 1899 [1]. Brownstein described regarding steatocystoma simplex in 1982. Since then a variety of names such as steatocystomatosis, sebocystomatosis and epidermal polycystic disease have been given to steatocystoma multiplex [2].

Steatocystoma, an uncommon cutaneous disorder, initially thought to be a sebaceous or retention cyst is recognized as a hamartomatous malformation of the pilosebaceous duct junction characterized by the development of numerous sebum containing dermal cysts with sebaceous glands in the cyst walls [3]. It, generally, can be classified into steatocystoma simplex when it is a solitary lesion and steatocystoma multiplex when there is multiple, small, soft, movable, yellowish to skin colored dermal cystic papules.

CASE REPORT

A 70-year-old male was presented to our outpatient clinic with asymptomatic dark patches bilaterally on malar region (Figure 1). The lesion had slowly enlarged over two years and physical examination revealed yellow to skin colored, soft movable cystic masses, ranging from 10 mm to 20 mm in size with no punctum. There was no remarkable family history of similar lesions and...
the patient had no other cutaneous diseases. The nails, teeth and hair were all normal. Routine laboratory investigations were within normal limits.

Surgical excision was done under local anesthesia (Figure 2). The lesion was sent for histopathological examination (Figure 3A–B). The specimen was stained in Hematoxylin and Eosin and was seen under 100x, the histopathology slide showed mild hyperkeratosis. Multiple cysts were lined by stratified squamous epithelium and granular layers were seen. The lumen of the cysts was filled with laminated keratin and a few of the cysts showed focal ulceration. Adjacent tissue exhibited skin appendages, dense acute and chronic inflammatory cells and numerous foreign body giant cells. A few melanophages were also seen in dermis. There was no evidence of granuloma or malignancy (Figure 4).

Figure 1: Dark patches seen unilaterally on malar region.

Figure 2: Post surgery of the patient.

Figure 3: (A) Surgical excision of the lesion done, (B) Excised specimen.

Figure 4: Histopathological examination showing stratified squamous surface epithelium exhibiting melanin pigmentation on the basal layer. Connective tissue shows multiple cysts filled with keratin along with few sebaceous gland and chronic inflammatory cell infiltrate, predominantly lymphocytes and few giant cells (H&E stain, x100).
DISCUSSION

Steatocystoma multiplex is an uncommon, inherited disorder that is characterized by multiple, asymptomatic, variably-sized dermal cysts. It is an uncommon disorder of the pilosebaceous unit characterized by the development of numerous sebum-containing dermal cysts [2]. Occurrence of large sized variant of these lesion clustered in large number in a localized region of body is rare. In our case steatocystoma multiplex was seen bilaterally on malar region, though it is the steatocystoma simplex variant that is more commonly seen on the face.

Steatocystoma simplex is the non-hereditary counterpart to the hereditary steatocystoma multiplex, clinically both are almost identical. Steatocystoma simplex is characterized by solitary, non-heritable growth mostly seen on face while multiplex is commonly seen on the axillae, groin, trunk and proximal extremities. It is rarely seen on face and scalp and is inherited, so is termed steatocystoma multiplex congenita. But some cases of steatocystoma multiplex seen on face and scalp can arise sporadically without any family history, as was the case seen in our patient [4]. It usually begins in late childhood and persists indefinitely, but in some patients the lesions have developed late in life [5]. Steatocystoma multiplex localized in the face and scalp has been divided into the following three types:

(i) Facial papular variant type
(ii) Sebocystomatosis and
(iii) Cysts located exclusively on scalp [5].

The etiological factors of steatocystoma multiplex are still unclear but trauma infection and immunological events have been proposed as some reasons [6]. Steatocystoma multiplex, familial cases have been linked to pachyonychia congenita and ectodermal dysplasia through a mutation in keratin 17.8. Pachyonychia congenita is a rare hereditary disorder characterized mainly by nail hypertrophy and dyskeratosis of skin and mucous membrane PC-2 (Jackson–Lawler form) and is accompanied by steatocystoma multiplex [7]. Steatocystoma is considered as forme fruste of pachyonychia congenita II.

Steatocystoma multiplex has also been associated with ichthyosis, koilonychia, acrokeratosis, Verruciformis of Hopf, hypertrophic lichen planus, rheumatoid arthritis, hypohidrosis, hypothyroidism and hypotrichosis. In our case, there were no such associated findings. Eruptive vellus hair cysts can mimic steatocystoma multiplex clinically and epidermal inclusion cysts have to be excluded. Comparison studies of keratin expression showed that epidermoid cyst expressed K10, EVHC expressed K17, trichilemmal cyst and steatocystoma multiplex expressed both K10 and K17 [8]. Kligman and Kirchbaum postulated that pluripotential ectodermal cells retain the embryonic capacity to form appendages or naevi rather than retention or inclusion cysts [9, 10]. Both steatocystoma multiplex and steatocystoma simplex arise from ducts of sebaceous glands.

Clinically, lesion is characterized by numerous small skin colored or yellowish cutaneous cysts. Colour of the lesion varies from yellowish to skin color depending upon the depth of the lesion, the superficial lesion being yellow and deeper lesions being skin colored [4]. It appears as multiple subcutaneous nodules, occurring anywhere on the body, even intraorally, but trunk and proximal extremities are the more common sites. The size of the nodules range from 0.2–2 cm and has no punctum. It remains asymptomatic but may get inflamed. When an inflamed cyst ruptures it produces steatocystoma multiplex suppurativa. The cyst may contain oily yellow fluid and vellus hairs. Steatoma is due to over production of sebum by one or more sebaceous glands while the outlet is closed. The sebum collected inside may undergo calcareous degeneration after remaining in situ for years destroying the epithelial lining.

The differential diagnosis should include other inherited syndromes of multiple benign adnexal tumors such as cylindromas, trichoepitheliomas, trichilemmomas who also have their counterpart in non-heritable solitary tumors.

The condition is histologically characterized by cystic structure with sebaceous glands within the cyst wall and epithelium that displays an eosinophilic cuticle. Inflammatory cells of macrophage monocyte lineage in connective tissue are also seen [6]. Keratin, oil and hairs in the lumen are also associated findings. Hyperkeratosis, multiple cyst lined by stratified squamous epithelium, lumen filled with lamellated keratin, foreign body giant cells in the lining cells, hair follicle, focal ulceration of cyst and more importantly a hyaline cuticle, all these features correlated well with our histological study too.

Various treatment modalities have been described each with its own merits and demerits. Oral isotretinoin, a retinoid, tetracycline along with incision and drainage, intralateral steroids, extirpating the contents with aspiration without removing the cyst wall or excocchleation of cyst wall with a curette are all methods often tried but with a high rate of recurrence.

Radiofrequency, cryosurgery, carbon dioxide laser, YAG laser are better options but residual scarring, blister formation, pain, postinflammatory hyperpigmentation are problems that have to be dealt with. Wide excision with its removal in toto is still the preferred choice because of the less postoperative complications and its cost effectiveness.

CONCLUSION

Steatocystoma is a cosmetic problem and a lifelong condition. There are no reports of malignant transformation from these benign adnexal tumors and we were able to confirm the unique nature of this lesion and rule out malignancy. Steatocystoma multiplex should be considered as a spectrum with different variations in
anatomical distribution which when properly diagnosed and timely intervened can be treated successfully.

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Author Contributions
Surej Kumar LK – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Nikhil Mathew Kurien – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Varun Menon P – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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