



MULTIPLE CUTANEOUS PILOLEIOMYOMA WITH UTERINE FIBROIDS (REED'S SYNDROME): A RARE CASE REPORT

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ABSTRACT

Cutaneous leiomyomas comprise approximately 5% of all leiomyomas. Solitary occurrence is more common than generalized. Multiple cutaneous piloleiomyomas are rare, painful and difficult-to-treat benign tumors originating in the arrectores pilorum muscles of the hair follicles. Multiple Cutaneous piloleiomyomas are associated with uterine fibroids in women (Reed's syndrome) and it is a marker of a condition called hereditary leiomyomatosis and renal cell cancer (HLRCC). We report a case of multiple cutaneous piloleiomyoma with uterine fibroid in an adult female. Because of its rarity and association with renal cell cancer, it deserves a mention in literature.

KEYWORDS: Cutaneous leiomyoma, Uterine leiomyoma, painful nodules.

INTRODUCTION

Cutaneous leiomyomas are of three types, namely piloleiomyoma arising from arrector pili muscle of hair follicle, genital leiomyoma from scrotal and labial dartos muscle as well as erectile smooth muscle of nipple, and angioleiomyoma from smooth muscle of vessels^[1]. Piloleiomyomas are uncommon benign smooth muscle tumors of skin. It usually presents as multiple reddish brown painful nodules which are sensitive to touch and cold. Multiple cutaneous leiomyomas may sometimes be associated with uterine fibroids (Reed's syndrome). It can also present without any risk of renal cell cancer. Smaller lesions can be treated conservatively by using calcium channel blockers, α blockers, nitrates etc and topical injections of triamcinolone acetonide. While larger lesions require surgical excision for pain relief and improve quality of life^[2].

CASE REPORT

A 45 years old Indian female presented to out-patient department of skin of our institute with complaints of multiple painful, itchy, skin colored nodules on back, inframammary area since 10 years. She gradually developed similar type of nodules

on left arm 2 years back. Nodules were not increasing in size but they gradually increased in number. Nodules were painful and sensitive to touch and cold. Pain typically disturbed the patient's daily activities. No history of discharge from the lesions, or systemic complaints. She had past history of hysterectomy for uterine leiomyoma. Both her sisters had similar history of skin lesions and hysterectomy done for uterine leiomyomas. Patient had no any associated co-morbidity and denied using any medication. On physical examination, multiple painful, dusky red-colored firm nodules measuring 5-10 mm in diameter were found on back (Figure 1), inframammary area (Figure 2) and left arm. On ice application over the skin nodules, the patient reported severe pain. Rest of the physical examination and systemic examination revealed no abnormality. Clinically, leiomyoma and neurofibroma were the differential diagnoses. On laboratory investigations, complete blood count, biochemical parameters, urine examination and chest x-ray were found to be normal. Skin biopsy was done from the nodule over back and on histopathological examination it revealed normal epidermis with underlying dermis showing well circumscribed

mass comprising of interlacing bundles of eosinophilic spindle cells without cytological atypia and/or mitotic figures, features suggestive of cutaneous pilo leiomyoma (Figure 3A, 3B). On Masson trichrome staining it showed interlacing bundles of red colored spindle cells within dermis confirming it to be of smooth muscle origin and differentiating from fibrous tissue (Figure 4). Abdominal ultrasonography and CT scan were advised to screen the patient for renal malignancy which revealed no abnormality.



Figure 1. Multiple dusky red nodules of varying size present on back



Figure 2. Multiple dusky red nodules on inframammary region

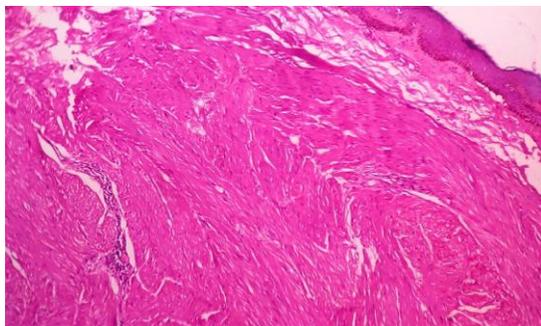


Figure 3A. Normal epidermis and dermis showing interlacing bundles of eosinophilic spindle cells (HE 10X)

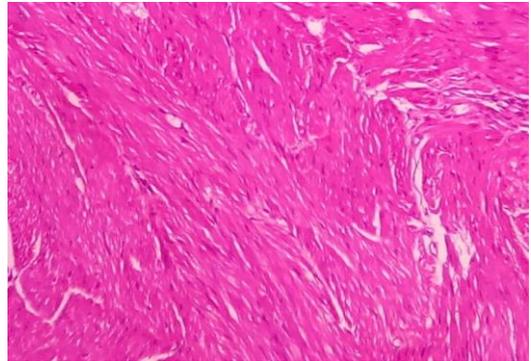


Figure 3B. High power view of Figure 3A showing interlacing bundles of spindle cells without atypia and/or mitotic figures (HE 40X)

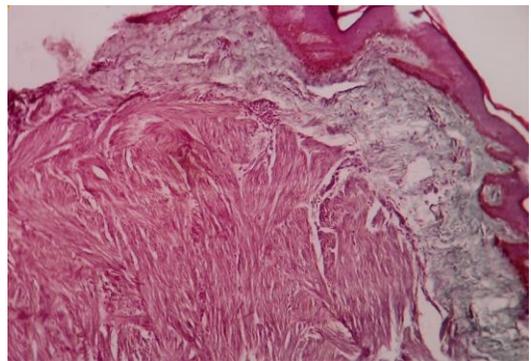


Figure 4. Masson trichrome staining showing bundles of red colored spindle cells in dermis confirming smooth muscle origin and differentiating it from collagen

DISCUSSION

The pilo leiomyoma is a benign tumor of the skin described originally by Virchow (1854)^[3]. Besnier, in 1880, classified leiomyomas as solitary and multiple^[4]. Cutaneous leiomyomas are of three distinct types as pilo leiomyoma, angioleiomyoma, and genital leiomyoma^[5]. Pilo leiomyomas are common in the age group of 10-30 years^[6]. Solitary lesions have male predominance while multiple lesions are more common in females^[1]. They may be disseminated, segmental, or zosteriform presentations over trunk. The arrector pili muscle, from which the pilo leiomyomas originate, attaches proximally to the hair follicle and distally to the multiple attachment points within the papillary and reticular dermis and basement membrane zone. Hence, pilo leiomyomas can originate from all these attachment points^[2].

Pilo leiomyomas can be developed sporadically or transmitted genetically^[7]. Pilo leiomyomas can be associated with several

conditions like uterine leiomyoma (leiomyomatosis cutis et uteri or Reed's syndrome)^[8], erythrocytosis/polycythemia^[9], and visceral involvement (gastrointestinal tract and retroperitoneal area). The gene responsible for Reed's syndrome has been localized to chromosome 1q42.3-43, encoding for fumarate hydratase enzyme^[2].

The classical clinical presentation in patients of piloileiomyomas is multiple painful skin nodules with diameter usually less than 2 cm^[1]. Pain may be induced by cold, tactile, or emotional stimuli, but it can also be spontaneous^[2]. The pain can be so intense that it can provoke nausea, vomiting, hypotension, micturition or pallor^[10,11]. In rare instances of severe impact on quality of life, the patients may attempt suicide^[12]. Pain may be caused by local pressure exerted by the tumor on cutaneous nerves or by infiltrating mast cells. The contraction of smooth muscle in the leiomyoma or the presence of increased numbers of nerve elements within the tumors are other proposed mechanisms for leiomyoma-associated pain^[13,14]. Immunohistochemical staining for the S-100 protein and neurofilaments has revealed more myelinated nerves and nerve axons in the lesions than in the normal skin of the same patient^[15].

The important differential diagnoses include other painful skin tumors such as blue naevus, glomus tumor, neurofibroma, eccrine spiradenoma, dermatofibroma, and cutaneous endometriosis^[1]. Special stains like Masson trichrome stain, Von Gieson stain and phosphotungstic acid haematoxylin stain can be used to distinguish smooth muscle cells from surrounding collagen. Piloileiomyomas stain positively for desmin and actin which are markers of smooth muscle differentiation.

Treatment of piloileiomyomas is surgical excision. In case of multiple lesions, larger and painful lesions can be excised with high chances of recurrence (around 50%)^[2]. The recurrences may be due to new lesions or they may result from the growth of partially excised lesions^[12]. Medical line of treatment aims at relieving pain in case of smaller lesions and various drugs used are calcium channel blockers, α blockers, nitrates, analgesics, gabapentin^[2]. Local injection of triamcinolone

acetone may be a promising new treatment option for multiple painful cutaneous piloileiomyomas^[16].

CONCLUSION

Multiple piloileiomyomas can be treated by medical therapy and so they should be distinguished from other surgically treatable causes of painful skin nodules like neurofibroma, angioliopoma, glomus tumor, eccrine spiradenoma as surgery can be avoided in such patients. Secondly, multiple piloileiomyomas can be associated with uterine leiomyomas and renal cell cancer. So patients with piloileiomyomas should be thoroughly screened for such visceral tumors.

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