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# Zosteriform Leiomyoma Cutis: A Rare Entity

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## Abstract

Leiomyoma is a well known benign tumour formed of hyperplastic smooth muscle elements, often encountered in uterus. Here we describe its rare skin equivalent leiomyoma cutis sub type -pilo leiomyoma in a young man. Presenting as papules and nodules over the back spanning over T7-T10 dermatomes in a zosteriform pattern for the past one year. The biopsy of which stood the histopathological test for leiomyoma demonstrating circumscribed non-encapsulated tumors, in the dermis, composed of poorly demarcated bundles of smooth muscle fibres arranged in a interlacing and whorled pattern. Immune histochemistry also confirmed smooth muscle actin positivity.

**Keyword:** Leiomyoma cutis, Zosteriform, pilo leiomyoma

### INTRODUCTION

Leiomyoma cutis is a rare painful skin tumour characterized by hyperplasia of smooth muscle elements found in the skin. There are three types of cutaneous leiomyomas: piloleiomyoma, angioleiomyoma and genital leiomyoma. Besnier classified these tumors as solitary and multiple. Multiple lesions occur commonly in men, seen in the first and third decades of life. The sites involved are the extensor aspect of the extremities, trunk, face and neck. Rarely, tumors are zosteriform or symmetrically distributed, suggesting a nevoid condition, and have been designated 'nevus leiomyomatosus systematicus'. We present here an unusual case of zosteriform leiomyoma cutis.

## Case Report

A 23 year old male presented with multiple painful skin coloured papules and nodules over the back for the past one year. The lesions had been gradually increasing in size and number in the last one year. The patient had started experiencing intermittent pain over the lesions, especially on exposure to cold. The patient's father had a history of similar lesions, with pain, restricted to his back. The patient had no other significant co-morbidity.

Examination revealed skin coloured, rounded, smooth, firm, tender papules and nodules ranging from 2 mm to 2 cm, arranged in clusters over the back.(Fig 1) The papules and nodules spread over T7-T10 dermatomes with a sharp demarcation over midline representing a zosteriform pattern. (Fig 2) On cold stimulation using ice, the patient reported severe pain over the lesions. There were no other mucocutaneous findings. Systemic examination was normal.

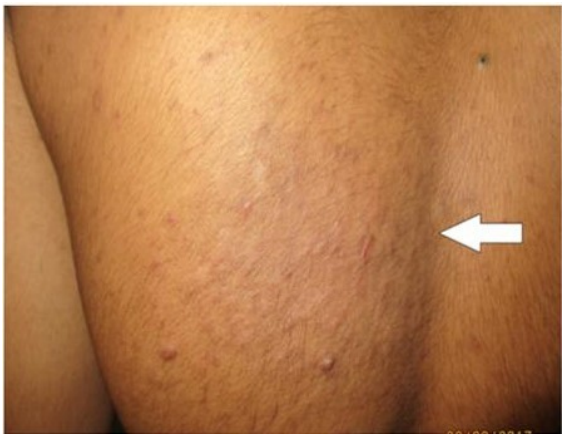

FIGURE-1

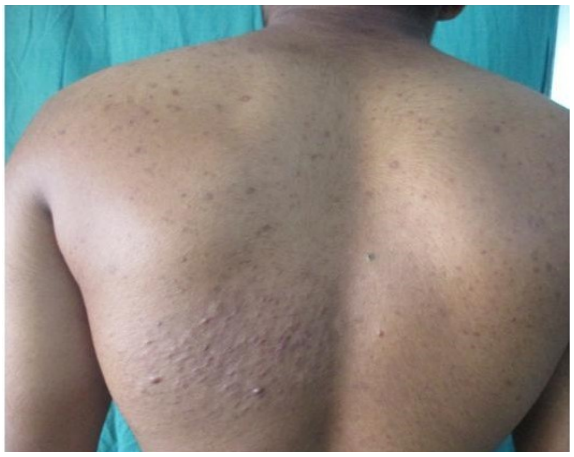

FIGURE-2

Clusters of papules and nodules in zosteriform pattern over back

Clinical differential diagnosis included dermatofibroma, angiolipoma, glomus tumor, eccrine spiradenoma, neurofibroma and lipoma.

Routine haematological, biochemical investigations and ultrasonography of abdomen were normal. Histopathological examination with hematoxylin and eosin staining showed circumscribed non-encapsulated tumors, centered on the dermis, composed of poorly demarcated bundles of smooth muscle fibres arranged in a interlacing and whorled pattern which suggested piloleiomyoma( Fig 3). On higher magnification the muscle fibers are straight, with little or no waviness. They had abundant eosinophilic cytoplasm with centrally located, thin, very long, blunt edged "eel-like" nuclei. (Fig 4) IHC for smooth muscle actin was positive confirming the diagnosis of- piloleiomyoma( Fig 5)

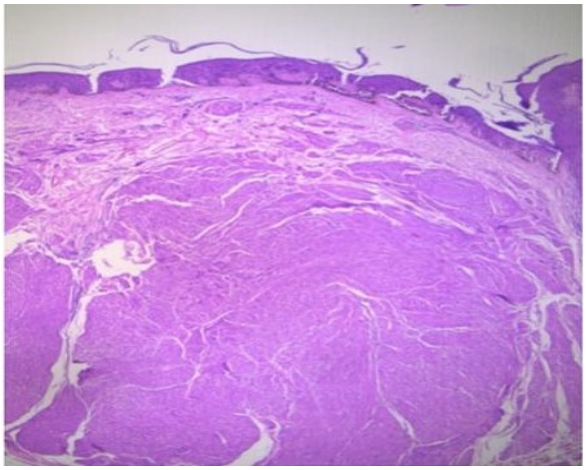

FIGURE-3 Histopathology showing intertwined fascicles of smooth muscles occupying the mid-dermis (H and E)

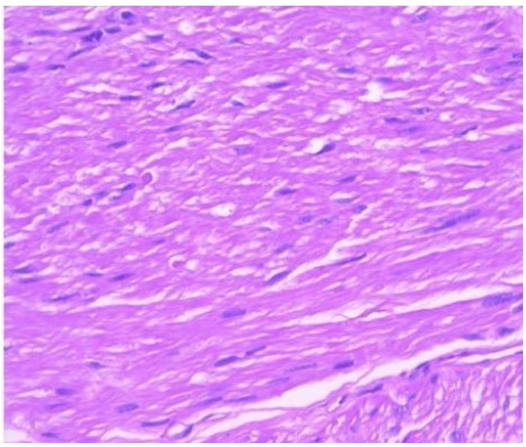

FIGURE-4 Elongated nuclei of smooth muscles with rounded edges. (H and E)

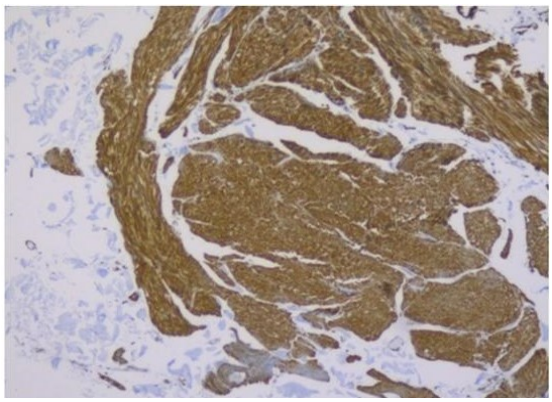

FIGURE-5 IHC stain –smooth muscle actin(SMA) positive

## Discussion

Cutaneous leiomyoma comprises approximately 5% of all leiomyomas. Three distinct types of cutaneous leiomyomas such as piloleiomyomas, angioleiomyomas, and genital leiomyomas are described. This classification hinges on the origin of the smooth muscle tumor and corresponds to the histologic or anatomic site where the leiomyomas are found. Thus tumors in each classification have distinct clinical and histologic characteristics.

Piloleiomyomas arises from the erector pili muscle of the pilosebaceous unit, which attaches proximally to the hair follicle and distally to multiple attachment points within the papillary and reticular dermis, as well as to the basement membrane. Piloleiomyomas can emerge from each of these various points of insertion and occur as multiple tumors Angioleiomyomas originate from smooth muscle within the walls of arteries and veins.

Genital leiomyomas originate from the dartos muscle of the scrotum and the labia majora, as well as those from the erectile muscle of the nipple.

Multiple piloleiomyomas may be inherited in an autosomal-dominant fashion and may be associated with uterine leiomyomas and aggressive renal carcinoma, also known as multiple cutaneous and uterine leiomyomatosis (MCUL) or Reed's syndrome and hereditary leiomyomatosis and renal cell carcinoma (HLRCC), respectively.[1, 2] The predisposition gene for Reed's syndrome has been localized to chromosome 1q42.3-43 and the gene encoding fumarate hydratase.[2] Appropriate screening measures for associated disorders are mandatory. In our patient, screening was done.

Two types of segmental manifestation of the autosomal dominantly inherited disease are postulated. The type 1 occurs due to heterozygosity for the underlying mutation with a clinical picture similar to that in a non-mosaic phenotype. In type 2, loss of heterozygosity causes homo- or hemizygosity, with a pronounced segmental manifestation of lesions[3,4] There was family history cutaneous leiomyoma in our patient , the clinical profile of our patient was suggestive of familial type 1 segmental leiomyoma.

Patients with piloleiomyoma often have pain that may be spontaneous or secondary to cold, pressure, or emotion.[5] The pressure on nerve fibers and abnormal muscle contraction have been suggested as possible explanations for pain in cutaneous leiomyoma which was present in our patient.

Surgical excision with skin grafting may be indicated for a small group of lesions. The condition may however recur, particularly in patients with multiple lesions. A number of pharmacologic treatment modalities have been employed for patients with painful multiple lesions involving large areas of the body. These include nifedipine, oral nitroglycerine, and oral alpha-1 adrenoceptor antagonist doxazosin. Cryotherapy and electrocoagulation have been attempted.[6] However; satisfactory results have been reported only with CO2 laser ablation therapy.[7]

## Conclusion

Based on the distinct clinical and histopathologic findings, a diagnosis of zosteriform leiomyoma cutis probably type 1 segmental type was made. This case is presented here for its rarity.

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