

Case Report

Zosteriform cutaneous leiomyoma: a case report

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ABSTRACT

Cutaneous leiomyomas are rare, benign soft tissue tumors arising from smooth muscles of the skin and comprise of three distinct subtypes, namely piloleiomyoma, angioleiomyoma and genital leiomyoma. Piloleiomyomas can present as solitary form, multiple disseminated and zosteriform or segmental forms. Cutaneous leiomyomas are rare, of which zosteriform leiomyoma is not commonly encountered. Here we report an uncommon case of Type I zosteriform cutaneous leiomyoma in a middle-aged individual which was confirmed on histopathology. Patient was further started on nifedipine with significant symptomatic improvement. The patient is planned for surgical excision and long-term follow in view of its association with aggressive renal malignancy.

Keywords: Cutaneous, Leiomyoma, Piloleiomyoma, Zosteriform

INTRODUCTION

Cutaneous leiomyomas are rare, benign painful tumours arising from smooth muscles. There are three different subtypes of cutaneous leiomyoma, according to their site of origin, namely; piloleiomyoma, angioleiomyoma and genital leiomyoma. Piloleiomyomas can have multiple disseminated (diffuse), segmental (zosteriform) or solitary presentation. Zosteriform leiomyoma can occur either alone (Type I), or with scattered non-segmental lesions elsewhere (Type II).¹ Here we report an uncommon presentation of Type I zosteriform cutaneous leiomyoma in a middle-aged individual with a brief review of literature on cutaneous leiomyomas.

CASE REPORT

A 35 years old male presented to the Dermatology Out-Patient Department with multiple, painful, skin coloured nodules of varying sizes ranging from 1 cm to 3 cm over right lower back in a grouped manner, distributed over

dermatomes T2-T5, for a duration of 6 years with history of associated paroxysmal burning pain, which exacerbated on exposure to cold, touch or physical stress (Figure 1). Figure 1 describes multiple tender skin coloured nodules over right lower back in a grouped manner (dermatomes T2-T5).

On systemic examination, no abnormality was detected. With this presentation and history, it was clinically diagnosed as zosteriform cutaneous leiomyoma, which is a rare entity and the biopsy was done to confirm the diagnosis. Histopathological examination revealed features of a benign dermal leiomyoma composed of bundles and fascicles of smooth muscle cells with abundant eosinophilic cytoplasm and cigar shaped nuclei and absence of nuclear pleomorphism and mitotic activity. Figure 2 shows photomicrograph of dermal leiomyoma composed of bundles and fascicles of smooth muscle cells. (Hematoxylin and Eosin 10X). Figure 3 shows photomicrograph of tumor composed of fascicles of smooth muscle cells with abundant eosinophilic cytoplasm

and elongated nuclei with blunt ends (Hematoxylin & Eosin, 40X).



Figure 1: multiple tender skin coloured nodules over right lower back in a grouped manner (dermatomes T2-T5).

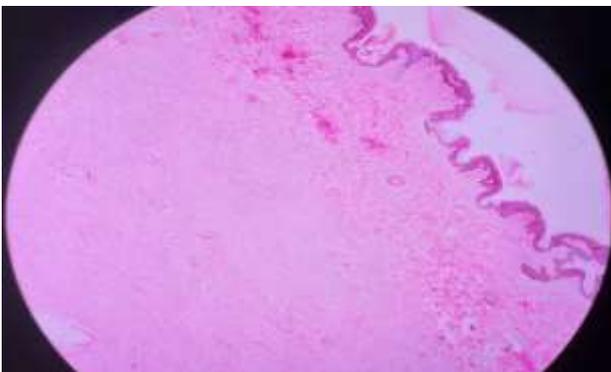


Figure 2: Photomicrograph of dermal leiomyoma composed of bundles and fascicles of smooth muscle cells. (Hematoxylin and Eosin, 10x).

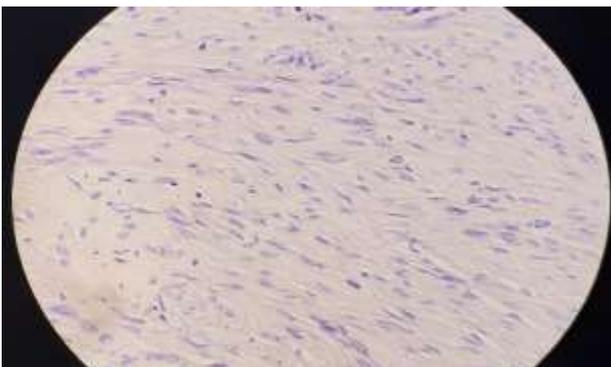


Figure 3: Photomicrograph of tumor composed of fascicles of smooth muscle cells with abundant eosinophilic cytoplasm and elongated nuclei with blunt ends (Hematoxylin and Eosin, 40x).

DISCUSSION

Leiomyomas are benign soft tissue neoplasms arising from smooth muscle cells. Skin is the second most common site for leiomyomas, next to uterus and comprise 75% of all extra- uterine leiomyomas.²

Literature on incidence and prevalence of cutaneous leiomyomas is lacking. They tend to occur more in adults than children. All races are equally affected and there is no sex predilection.³ Cutaneous leiomyomas are subcategorized based on the site of origin of tumor from the smooth muscle cells in the skin, namely, piloleiomyoma (arising from arrector pili muscle), angioleiomyoma (arising from tunica media of blood vessels) and genital leiomyoma (arising from dartos muscle, vulva, nipple and areola).⁴ Piloleiomyoma is the most common type. Cutaneous leiomyoma can present as multiple piloleiomyoma, solitary piloleiomyoma, solitary angioleiomyoma or solitary genital leiomyoma.

Piloleiomyomas present as solitary or multiple, firm, reddish-brown papulonodule with a predilection for the face, back, and extensor surfaces of the extremities.⁴ Solitary piloleiomyomas usually appear in the elderly. Multiple piloleiomyomas occur in younger patients in the age group of 10 -30 years sporadically or in the setting of a Multiple Cutaneous and Uterine Leiomyomatosis syndrome (MCUL), formerly known as Reed's syndrome. It is an autosomal dominant genetic disorder, resulting from a heterogenous germline mutation in the gene encoding the enzyme fumarate hydratase (FH gene) located on 1q42.3-q43 region.⁵ Patients with Reed's syndrome have increased susceptibility to develop smooth muscle tumors in skin and uterus. Almost all female patients (more than 90%) with this syndrome have uterine leiomyomas which are larger and numerous compared to non – hereditary leiomyomas and occur by the age of 30 years.^{4,5}

The most concerning feature of MCUL is its association with renal cell carcinoma in approximately 15 to 20% of the cases. MCUL associated with renal cell carcinoma is labelled as Hereditary Leiomyomatosis with Renal Cell Carcinoma (HLRCC). HLRCC is associated with type 2 papillary renal cell carcinoma which is an aggressive malignancy.^{4,5} The loss of tumor suppressor function of FH gene is believed to be the predisposing factor in the pathogenesis of MCUL and HLRCC. Molecular detection of FH gene mutation confirms the diagnosis and also helps in screening of family members for early surveillance and detection.

Clinically, multiple piloleiomyomas can have varied distribution. It can have multiple disseminated or grouped distribution. Grouped distribution is common and can present as linear or zosteriform variants. Zosteriform lesions occur along a dermatome while linear lesions follow Blaschko's lines of fetal development.⁶

Zosteriform leiomyoma can be categorized into two types. Type I zosteriform leiomyoma is characterized by presence of dermatomal distribution only, while in Type II there are segmental lesions superimposed on scattered, isolated, nonsegmental lesions.^{1,7} The present case had only segmental lesion in the right lower back along dermatomes T2 to T5 and is classified as Type I

Zosteriform leiomyoma. In literature, a few cases have been reported with this type of zosteriform distribution.^{1,8} A rare case of unilateral multi-segmental cutaneous leiomyoma of type I has also been reported.⁹

Pathogenesis of pain is hypothesized to be due to local pressure exerted by the tumor on the cutaneous nerves or due to local ischemia caused by muscle contraction.^{3,4} The differential diagnoses include dermatofibroma, neurofibroma, schwannoma, angioliipoma, eccrine spiradenoma, granular cell tumors and glomus tumor.

The histologic finding of pilar leiomyoma is an unencapsulated circumscribed tumor composed of interlacing bundles and fascicles of spindle shaped smooth muscle cells with abundant eosinophilic cytoplasm and blunt cigar-shaped nuclei. Atypical or symplastic leiomyoma, akin to symplastic leiomyoma of uterus, with atypia, pleomorphism and minimal mitosis have been described as very rare variant.³ Though haematoxylin and eosin stained sections are diagnostic, positivity for immunohistochemistry markers like smooth muscle actin and desmin is confirmatory and rules out other differential diagnoses like schwannoma, neuromas and fibrohistiocytomas.

The treatment of choice for painful solitary as well as multiple cutaneous leiomyomas is surgical excision. However, recurrences have been reported in 50% patients with multiple lesions.² Pharmacological treatment modalities for patients with painful multiple lesions include nifedipine, oral nitroglycerine and oral alpha-1 adrenoceptor antagonist doxazosin. Cryotherapy, electrocoagulation and CO₂ laser ablation have been attempted; however only limited studies have shown satisfactory results.²

In the present case, patient was diagnosed to have Type I zosteriform cutaneous leiomyoma and was treated with nifedipine and planned for surgical excision. Due to its association with renal carcinoma, patient is being followed up regularly.

CONCLUSION

Cutaneous leiomyoma is a rare entity, out of which zosteriform leiomyoma is not common. They are benign yet problematic due to pain and discomfort. Their early

detection, management and follow up are crucial due to the fact that piloleiomyomas may be associated with underlying genetic mutations which increase the susceptibility to develop renal cell carcinoma.

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