Zosteriform Pilar leiomyoma associated with uterine leiomyoma: A case report

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Abstract

Cutaneous leiomyomas are benign tumors arising from smooth muscles of the skin. Multiple lesions may be arranged in segmental, zosteriform, disseminated patterns. Multiple pilar leiomyomas may be inherited in an autosomal dominant pattern and may be associated with uterine fibroids and renal cell carcinoma, also known as Reed Syndrome.

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Key clinical message:

Cutaneous leiomyomas are rare benign tumors originating from smooth muscles of skin. Zosteriform leiomyoma is a type of cutaneous leiomyoma. Its association with uterine leiomyoma is one of the rarest findings.

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CASE REPORT

BACKGROUND

Leiomyomas were first termed by Virchow in 1854 as "tuberculum dolorosum".[1] Cutaneous leiomyomas are rare, benign tumors that may be sporadic or inherited, originated from smooth muscle fibres and can be associated with various disorders.[2]It can be categorized into three subtypes depending upon the origin, arrector pili mucles (pilar leiomyoma), smooth muscle of blood vessels (angioleiomyoma) and tunica dartos or labia majora (dartoic or genital leiomyoma).[3] cutaneous leiomyomas are painful tumors that are more common in adults than children.[4] In 1880, Besnier classified leiomyomas as solitary or multiple.[5] When multiple, these can be grouped, linear, segmental, zosteriform or dermatomal pattern.[6]Generally it affects extremities followed by trunk, face and neck and presents as firm skin colored to reddish brown papules or nodules. It is often associated with pain, spontaneous or induced by cold or pressure.[7] The cause of pain is unknown but some suggest it to be due to cutaneous nerve compression, or by local ischemia caused by contraction of smooth muscle fibres of the tumor.[7,8]

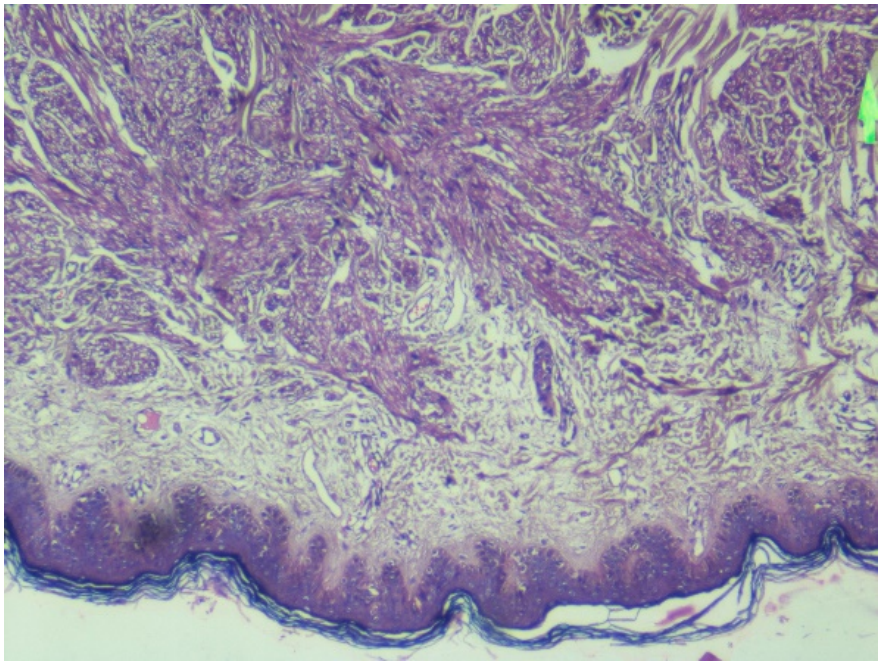
Pilar leiomyomas can be transmitted as autosomal dominant trait and may be associated with uterine leiomyomatosis or renal cell cancer, also known as Reed syndrome or Multiple cutaneous and uterine leiomyomatosis (MCUL) or hereditary leiomyomatosis and renal cell cancer (HLRCC).[8,9]

OBSERVATION

A 36 year old, married female presented to us with multiple painful skin colored to brownish papules and nodules involving right upper extremity and right side of upper chest for the duration of three years. These lesions initially appeared over right forearm and gradually increased in number to involve right arm and right upper chest.(Figure 1,2) The lesions started as skin colored papules and few of them progressed in size to form a nodule. It was associated with pain, during touch and exposure to cold. Menstrual and bladder history did not show any abnormalities. General physical examination was unremarkable. Cutaneous examination revealed multiple tender skin colored to hyperpigmented firm papules and nodules varying in size from 0.5 cm to 2 cm, distributed in segmental pattern involving right upper extremity and right upper chest (C5 to C8) dermatome. Based upon history and clinical examination we made differential diagnosis of leiomyomas, neurofibromas, and angiolipomas. Routine hematological investigations including urea, creatinine and urine examination did not reveal any abnormalities. However, ultrasound examination of abdomen and pelvis revelead a lesion measuring 19x18x13 mm arising from posterior wall of uterine body suggestive of intramural leiomyoma. Serological tests were negative. Histopathological tests taken from right upper chest and right forearm showed well circumscribed tumor composed of smooth muscles arranged in interlacing fascicles. The tumor cells were spindle shaped with elongated nuclei with blunt ends with abundant amount of fibrillary eosinophilic cytoplasm confirming the diagnosis of pilar leiomyoma.(Figure 3, 4)

Patient was managed symptomatically for pain with Tab Nifedipine 10 mg twice daily and is kept on routine follow up for uterine leiomyoma accordingly.

Figure 1 and 2: Clinical pictures of cutaneous leiomyoma in zosteriform pattern involving right upper limb.



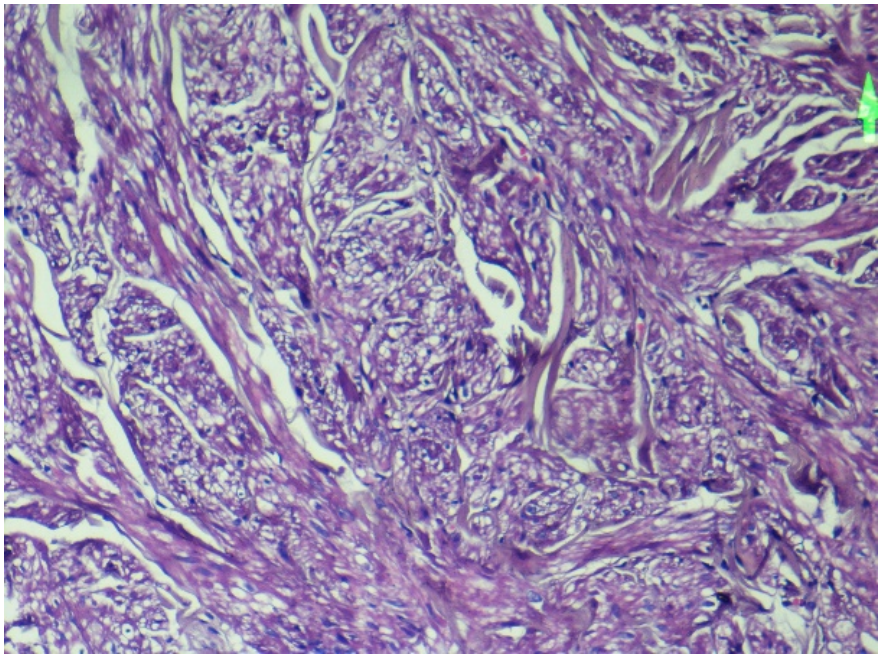


Figure 3 and 4: HIstopathological pictures of cutaneous leiomyoma showing smooth muscles arranged in interlacing fascicles.

DISCUSSION

Cutaneous leiomyomas are rare benign, painful tumors arising from smooth muscle fibres. According to site of origin, the types are pilar leiomyoma, angioleiomyoma and dartoic leiomyoma, pilar leiomyoma being the most common type. Pilar leiomyomas often occur as multiple and disseminated or segmental pattern in

80% of the cases.[10] Multiple lesions are inherited as autosomal dominant trait and may be associated with a condition termed Reed syndrome. The predisposing gene for Reed syndrome is localized in chromosome 1q42.3-43 which encodes Fumarate Hydratase (FH) gene. FH is involved in Krebs cycle for conversion of fumarate to malate. Along with this, FH is also believed to be a tumor suppressor gene and mutation of FH leads to increased risk for developing renal tumors.[9,11] The lesions in multiple pilar leiomyomas can be arranged in disseminated , blaschkoid or segmental(zosteriform) patterns. Segmental pattern is further categorized into type 1 and type 2 segmental.

Histopathologically, pilar leiomyomas are circumscribed, non-encapsulated tumors comprised of smooth muscles bundles arranged in interlacing patterns, and sometimes in whorled pattern. The cells have abundant eosinophilic cytoplasm and elongated nuclei with blunt ends.

Treatment of pilar leiomyomas remains frustrating. If the lesion is solitary or few in number, excision can be considered. However, multiple lesions are often treated symptomatically for pain. The treatment options for pain in leiomyomas are calcium channel blockers like nifedipine which blocks influx of calcium into smooth muscle, alpha adrenergic blocking agents such as doxazocine and phenoxibenzamine, and gabapentin and topical 9% hyoscine hydrobromide. CO2 laser, cryotherapy and electrocoagulation are alternative treatments.[7]

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Data availability statement

The data that support the findings of this study are openly available in Clinical Case Reports

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Detailed author's contribution :

ES contributed to the collection of data and the management of the patient. ES wrote the initial draft of manuscript. ES and AM revised and prepared the final version of the manuscript. All authors have read and approved the final manuscript and agree to take full responsibility for the integrity and accuracy of the work.

References

1. Garman ME, Blumberg MA, Ernst R, Raimer SS. Familial leiomyomatosis: a review and discussion of pathogenesis. Dermatology. 2003;207(2):210-3.
2. Malik K, Patel P, Chen J, Khachemoune A. Leiomyoma cutis: a focused review on presentation, management, and association with malignancy. Am J Clin Dermatol. 2015 Feb;16(1):35-46.
3. Bandyopadhyay D, Saha A, Bhattacharya S. Bilateral Multisegmental Zosteriform Leiomyoma Cutis: A Rare Entity. Indian J Dermatol. 2015 May-Jun;60(3):293-5.
4. Malhotra P, Walia H, Singh A, Ramesh V. Leiomyoma cutis: a clinicopathological series of 37 cases. Indian J Dermatol. 2010 Oct;55(4):337-41.
5. Kudur MH. A generalized multiple cutaneous piloleiomyomatosis in a young male: rare case report. Indian J Dermatol. 2013 May;58(3):245.
6. Hoyt BS, Tschen JA, Cohen PR. Solitary pilar leiomyoma of the nasal dorsum: case report and literature review. Indian J Dermatol. 2015 Jan-Feb;60(1):82-4.
7. Albuquerque MM, Rocha CF, Costa IS, Maia Rda R, Branco FJ, Gonçalves Hde S. Piloleiomyoma with segmental distribution–Case report. An Bras Dermatol. 2015 May-Jun;90(3 Suppl 1):178-80.
8. Basendwh MA, Fatani M, Baltow B. Reed's Syndrome: A Case of Multiple Cutaneous Leiomyomas Treated with Liquid Nitrogen Cryotherapy. Case Rep Dermatol. 2016 Mar 18;8(1):65-70.
9. Kudligi C, Khaitan BK, Bhagwat PV, Asati DP. Unilateral multi-segmental leiomyomas: a report of rare case. Indian J Dermatol. 2013 Mar;58(2):160.
10. Ritzmann S, Hanneken S, Neumann NJ, Ruzicka T, Kruse R. Type 2 segmental manifestation of cutaneous leiomyomatosis in four unrelated women with additional uterine leiomyomas (Reed's Syndrome). Dermatology. 2006;212(1):84-7.
11. Emer JJ, Solomon S, Mercer SE. Reed's Syndrome: A Case of Multiple Cutaneous and Uterine Leiomyomas. J Clin Aesthet Dermatol. 2011 Dec;4(12):37-42.

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