Segmental Leiomyoma: A Report of Two Cases

Introduction

Leiomyomas are a rare form of benign smooth muscle tumors. The most common site of occurrence is the uterus (95%), followed by the skin. Cutaneous leiomyoma accounts for 75% of the extrauterine leiomyomas.[1] The pathogenesis of cutaneous leiomyoma is unknown. Based on their site of origin, they are classified as piloleiomyoma, angioleiomyoma, and genital leiomyoma.[2] Piloleiomyoma is the most common variant arising from the arrector pili muscle.[3] Segmental leiomyomas represent the mosaic manifestation of cutaneous leiomyomas.[2] We present two cases of Type 1 segmental leiomyoma, which are rare and unique.

Case Report 1

A 27-year-old man presented with a history of multiple intermittently painful skin-colored raised lesions limited to the right side of the back region for 3 years. The patient experienced increased pain whenever lesions came in contact with cold water. Cutaneous examination revealed multiple, firm, tender, skin-colored to pale red papules and nodules. The lesions are grouped over the right side of the posterior trunk in a segmental distribution [Figure 1a]. Incisional skin biopsy revealed a well-circumscribed lesion composed of spindle cells arranged in a whorled pattern in the deep dermis [Figure 2a]. Masson trichrome stained the smooth muscle bundles in the dermis as red color [Figure 2b]. He was started on oral nifedipine and advised regular follow-up.

Case Report 2

A 26-year-old man presented with painful raised lesions over his left upper arm since 1 year. He experienced an increased intensity of pain upon friction. Cutaneous examination showed three well-defined, firm, tender, skin-colored to erythematous nodules of varying sizes. These nodules were present over the antero-medial aspect of the upper left arm [Figure 1b]. Based on the distinct clinical and pathological findings, he was diagnosed to have segmental leiomyoma and the lesions were excised.

Discussion

Piloleiomyomas commonly manifest between the first and third decades of life with no gender predilection.[4] It clinically presents as skin-colored to red-brown dermal papules or nodules, mainly distributed over the proximal extremities, followed by trunk, face, and neck.[3] The pain in leiomyoma may either be spontaneous or induced, secondary to cold, pressure, emotional stress, and friction.[3] The pain may be of burning, stabbing, or pinching in nature which could be attributed to pressure of the tumor on the underlying nerves or smooth muscle fiber contraction and infiltration of mast cells. Pain is more commonly observed in diffuse and segmental pattern.[5]

Piloleiomyomas can either be solitary or multiple. Multiple leiomyomas are more common and are termed as leiomyomatosis. When the leiomyoma lesions are more than 5000, it is known as myomatosis cutis miliaris.[6] Usually, multiple piloleiomyomas are commonly observed between 10 and 30 years of age, whereas solitary piloleiomyomas are seen with advancing age.[1] Multiple piloleimyomas are transmitted in an autosomal dominant fashion, which may be associated with uterine leiomyomas (Reed's syndrome/MCUL—multiple cutaneous and uterine leiomyomatosis) and hereditary leiomyomatosis and renal cell carcinoma (HLRCC). A predisposing factor to MCUL and HLRCC is the gene encoding fumarate hydratase mutation on chromosome 1q42.3–43.[7] Multiple lesions in the leiomyoma may present in different patterns, such as disseminated/diffuse, blaschkoid, and segmental/zosteriform.[1,6] The other reported forms are grouped, linear, and symmetrical pattern.[8]

Segmental leiomyomas are of two types: (i) Type 1 and (ii) Type 2. Both are inherited as autosomal dominant pattern of inheritance.[2] The salient features of the two types have been summarized in Table 1.[6] Cutaneous leiomyomas have to be differentiated from other painful cutaneous tumors such as the dermatofibroma, angiolipoma, eccrine spiradenoma, schwannoma, and neuromas.[6] Other painful conditions in segmental distribution are given in Table 2.[9-14]

The piloleiomyoma is histopathologically characterized by poorly demarcated interlacing bundles of smooth muscle cells with varying amounts of intermingling collagen fibers in a low-power field. The muscle fibers are usually straight, with centrally located long, thin, "eel-like nuclei."[8] The histopathological differential diagnosis includes other spindle tumors such as dermatofibroma, leiomyosarcoma, neurofibroma, and schwannoma.[8] Masson trichome stain and immunohistochemistry are helpful in differentiating piloleiomyomas from other spindle cell tumors.[1,8]

The treatment aspects for leiomyomas are not satisfactory. Avoiding trauma and exposure to cold can prevent the pain in leiomyoma.[3] Surgical excision can be done when

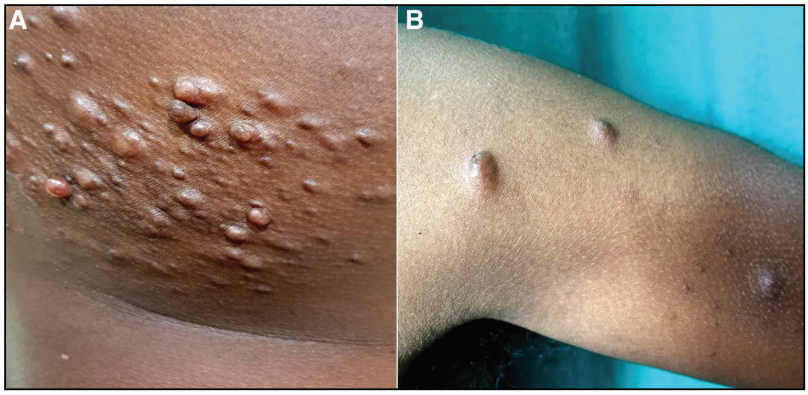

Figure 1: (a, b) Multiple skin-colored to red-colored papules and nodules in a segmental distribution

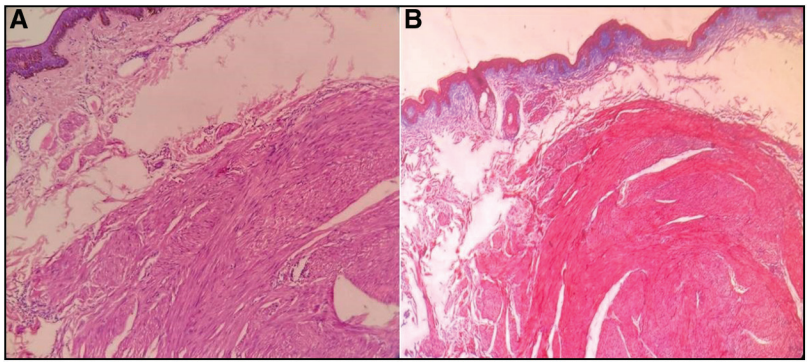

Figure 2: (a) HPE 10×—A circumscribed tumor with fascicles of smooth muscle bundles. (b) Masson trichrome stain (4×)—Red color highlights the smooth muscle bundles in the dermis

Table 1: Difference between Type 1 and Type 2 segmental leiomyomas

Type 1 segmental leiomyomas	Type 2 segmental leiomyomas
Due to the heterozygosity of the postzygotic mutation	Loss of heterozygosity causes homo/hemizygosity of the underlying mutation
Clinical features resemble non-mosaic phenotype	Causes pronounced segmental lesion superimposed on non-segmental phenotype
More common	Less common
Less severe	More severe

lesions are few; however, recurrence is common.[4] In case of multiple lesions, where excision is not possible, liquid nitrogen cryotherapy, electrocoagulation, and CO_2 laser ablation can be considered. Drugs such as nifedipine, doxazosin, gabapentin, antidepressants, and topical 9% hyoscine hydrobromide are helpful in relieving the pain.[5] Recently, botulinum toxin has been used for pain management.[15]

To conclude, leiomyoma cutis Type I is a rare but painful tumor that can affect the quality of life. The cornerstone of management is planning the treatment appropriate to the patient.

Acknowledgments

Nil.

Table 2: Segmental painful cutaneous tumor

- Eccrine spiradenoma
- Blue rubber bleb nevus
- Dermatofibroma
- Eccrine angiomatous hamartoma
- Glomangioma
- Schwannoma

Author Contribution

Arumugam Iswarya has contributed to the content, literature search, manuscript preparation, and submitted the article.

Palaniappan Vijayasankar has contributed to the design and literature search of the article.

Kaliaperumal Karthikeyan has planned the concept, manuscript preparation, manuscript editing, manuscript review, and is the guarantor of this article.

Vijayaraghavan Sriram has contributed to the histopathology part.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will

not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Arumugam Iswarya[1], Palaniappan Vijayasankar[1], Kaliaperumal Karthikeyan[1], Vijayaraghavan Sriram[2]

[1]Departments of Dermatology, Venereology, and Leprosy, [2]Pathology, Sri Manakula Vinayagar Medical College and Hospital, Puducherry, India

Address for correspondence: Dr. Kaliaperumal Karthikeyan, Department of Dermatology, Venereology, and Leprosy, Sri Manakula Vinayagar Medical College and Hospital, Puducherry 605107, India.
E-mail: karthikderm@gmail.com

References

1. Malhotra P, Walia H, Singh A, Ramesh V. Leiomyoma cutis: A clinicopathological series of 37 cases. Indian J Dermatol 2010;55:337-41.
2. Tsoitis G, Kanitakis J, Papadimitriou C, Hatzibougias Y, Asvesti K, Happle R. Cutaneous leiomyomatosis with type 2 segmental involvement. J Dermatol 2001;28:251-5.
3. Albuquerque MM, Rocha CF, Costa IS, Maia Rda R, Branco FJ, Gonçalves Hde S. Piloleiomyoma with segmental distribution—Case report. An Bras Dermatol 2015;90:178-80.
4. Kura MM, Parsewar S. Skindia quiz 19: Soft tumors in segmental fashion. Indian Dermatol Online J 2015;6:302-3.
5. Vasani RJ. Unilateral segmental type II leiomyomatosis: A rare occurrence. Indian J Dermatol Venereol Leprol 2012;78:521.
6. Lang K, Reifenberger J, Ruzicka T, Megahed M. Type 1 segmental cutaneous leiomyomatosis. Clin Exp Dermatol 2002;27:649-50.
7. Choudhary S, McLeod M, Torchia D, Romanelli P. Multiple cutaneous and uterine leiomyomatosis syndrome: A review. J Clin Aesthet Dermatol 2013;6:16-21.
8. Suwattee P, Dakin C. Bilateral segmental leiomyomas: A case report and review of the literature. Cutis 2008;82:33-6.
9. Englander L, Emer BJ, McClain D, Amin B, Turner RB. Segmental eccrine spiradenomas. J Clin Aesthet Dermatol 2011;4:38-44.
10. Happle R. Type 2 segmental manifestation of "blue rubber bleb nevus syndrome": A reappraisal of 5 case reports. Eur J Dermatol 2010;20:266-8.
11. Bhabha FK, Magee J, Ng SY, Grills CE, Su J, Orchard D. Multiple clustered dermatofibroma presenting in a segmental distribution. Australas J Dermatol 2016;57:e20-2.
12. Patterson AT, Kumar MG, Bayliss SJ, Witman PM, Dehner LP, Gru AA. Eccrine angiomatous hamartoma: A clinicopathologic review of 18 cases. Am J Dermatopathol 2016;38:413-7.
13. Cabral CR, Oliveira Filho Jd, Matsumoto JL, Cignachi S, Tebet AC, Nasser Kda R. Type 2 segmental glomangioma—Case report. An Bras Dermatol 2015;90:97-100.
14. Cherian A, Baheti NN, Sarada C, Iype T, Mathew M. Segmental schwannomatosis: An uncommon cause of crural monoparesis. J Neurosci Rural Pract 2012;3:420-1.
15. Onder M, Adişen E. A new indication of botulinum toxin: Leiomyoma-related pain. J Am Acad Dermatol 2009;60:325-8.

Submission: 15-06-2021 **Revision:** 05-08-2021
Acceptance: 09-08-2021 **Web Publication:** 29-09-2021

Access this article online

Quick Response Code:

Website: www.tjdonline.org

DOI: 10.4103/tjd.tjd_50_21

How to cite this article: Iswarya A, Vijayasankar P, Karthikeyan K, Sriram V. Segmental leiomyoma: A report of two cases. Turk J Dermatol 2021;15:80-2.