

Letter to Editor

Unilateral Multi-segmental Type 2 Pilar Leiomyomatosis: Uncommon Entity

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Dear Editor,

Cutaneous leiomyoma (CL) is a benign tumour of smooth muscle cells. CL comprises approximately 5% of all leiomyomas. CL can be classified based on the number of lesions as solitary, multiple and segmental. Segmental leiomyomatosis (SL) can be of two types, namely type I and type II. Herein, we report a rare case of type II SL.

A 45-year-old male presented with multiple painful swellings and nodules over the right side of the back for 10–12 years. It initially started as a small papule on the right side of the back, which gradually increased in size and number over time. However, the patient noticed a recent increase in number of lesions for 2 years. These lesions were associated with moderate-to-severe pain with exacerbation on exposure to cold, emotional stress and during winter season. The patient had neither systemic symptoms nor a family history of a similar illness. On examination, there were multiple indurated skin-coloured to slightly hyperpigmented fleshy papules and nodules involving multiple thoracic and lumbar dermatomes at the right side of the back [Figure 1]. The lesions in the right lumbar segments were coalescing to form plaques, while few discrete nodules were noted on the left side of the back. Histopathology examination of a nodule from the back (Haematoxylin and Eosin $\times 100$) [Figure 2a] revealed epidermal atrophy with Grenz zone, bundles of spindle-shaped smooth muscle cells with an eosinophilic cytoplasm [Figure 2b and c] and a cigar-shaped nucleus with a perinuclear halo. Immunohistochemistry was strongly positive for smooth muscle actin, suggesting the presence of smooth muscles ([Figure 2d], $\times 100$). The renal imaging studies, including ultrasonography and magnetic resonance imaging (MRI), did not reveal any abnormality. Based on clinical and histopathological features, a diagnosis of type II SL was made. The patient was advised to avoid cold exposure and started on tablet nifedipine 10 mg twice daily. The patient had partial relief in pain.

CL is a rare benign painful smooth muscle tumour. They can be single or multiple. The multiple CL can be either generalised or segmental. The segmental distribution has been further subclassified as type I and type II by Happle. Most cases present between the ages of 21 and 51 years, typically in the 3–5th decade of life, with a slight male predominance. The lesions can be papulonodular, firm and reddish-brown, usually unilateral and scattered across various segments of the body, including the back, chest and limbs. A left-sided predilection has been observed in some cases, though both sides may be involved. Systemic associations such as uterine leiomyomas are common in females, while genetic predisposition is noted in a minority of cases, suggesting that most instances are sporadic [Table 1].^[1-6] The pathophysiology behind this varied presentation

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Table 1: Unilateral type II segmental leiomyomatosis.

Author	Particulars of patients (age/sex)	Clinical presentation	Family history	Systemic associations	Diagnosis/comments (if any)
Ritzmann <i>et al.</i> , ^[2] (2006)	Four female patients aged between (43 and 51 years)	Single to multiple segmental involvements along with scattered lesions at opposite side	+++	Uterine leiomyoma in all four cases	Type II segmental leiomyomatosis with Reed syndrome/age of onset was between (20 and 30 years)
Vasani ^[3] (2012)	22 years/male	Papulonodular lesions on two segments at the left side of upper and lower back	+	Nil	Type II segmental leiomyomatosis
Kudligi <i>et al.</i> , ^[1] (2013)	30 years/female	Multi-segmental distribution along the 5 th cervical, 6 th dorsal and 1 st sacral segments of right half of the body	Nil	Nil	Type II segmental leiomyomatosis
Tsoitis <i>et al.</i> , ^[4] (2001)	21 years/male	Multiple leiomyomas preponderantly involving the right half of the chest and the back	Nil	Nil	Type II segmental leiomyomatosis
Das <i>et al.</i> , ^[5] (2015)	32 years/male	Left T9, T10 segment scattered lesion on the opposite side	Nil	Nil	Type II segmental leiomyomatosis
Paolino <i>et al.</i> , ^[6] (2022)	27 years/male	Multiple, cutaneous, papulo-erythematous lesions, distributed in the left mammal areas with other lesions in the upper left limb	Nil	Nil	Type II segmental leiomyomatosis



Figure 1: Multiple skin-coloured to slightly hyperpigmented fleshy papules and nodules involving several thoracic and lumbar segments on the right side of the back. The lesions in the right lumbar segments are coalescing to form plaques, while a few discrete nodules are noted on the left side of the back.

is not yet clear. However, it is postulated that type I SL results from heterozygous post-zygotic mutations, leading to

segmental skin lesions that are similar to those seen in a non-mosaic phenotype. Type II SL results from a post-zygotic mutational event in a heterozygous embryo, leading to loss of heterozygosity. This process produces a distinct pattern of segmental lesions that overlap with the typical phenotype of the underlying disease.^[1] Type I SL typically presents as a single unilateral segmental involvement, while type II involves multiple unilateral segments and may include non-segmental lesions on the contralateral side. In the present case, the lesions are predominantly distributed on the right side of the back with few scattered lesions on the contralateral side. The possibility of Reed syndrome (RS) must be ruled out in any case of multiple CL. The RS screening can be done in three ways: (1) Clinical screening (family history of CL and uterine fibroid), (2) radiological screening (ultrasound and MRI/computed tomography studies to look for renal cancer) and (3) genetic mutation test for fumarate hydratase gene or special stain for the semiquantitative assessment of the fumarate hydratase enzyme in the tumour tissue. There is an anecdotal report of RS associated with type 2 SL.^[2] In the current case, there was no family history of leiomyoma, and the renal MRI results were normal. The genetic screening could not be done due to non-availability of the genetic screening facility and economic constraints. Therefore, the patient is advised to have annual follow-ups for the early

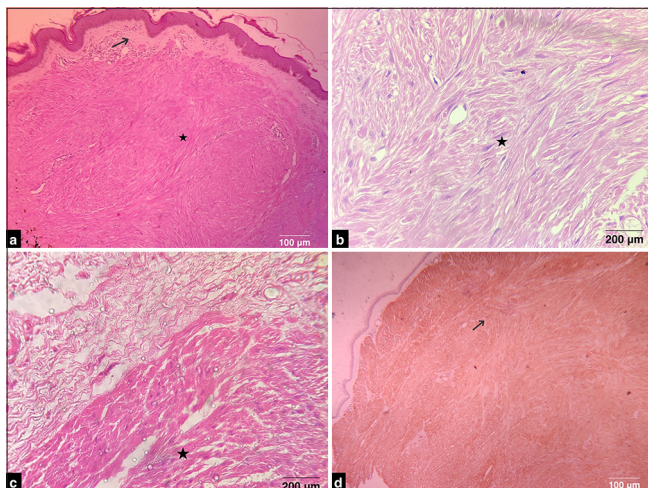


Figure 2: Histological section showing. (a) Epidermal atrophy with wide Grenz zone (black arrow), dermis showed circumscribed, non-encapsulated tumour with interlacing fascicles (black star) with wavy muscle fibre bundles (Haematoxylin and Eosin [H&E], $\times 100$). (b and c) (H&E, $\times 400$) dermis unveiled spindle-shaped cells with eosinophilic cytoplasm (black star), a cigar-like elongated nucleus with perinuclear halo. (d) On immunohistochemistry (smooth muscle actin, $\times 100$), cells were positive for smooth muscle actin (black arrow).

detection of renal or other extracutaneous complications. To conclude, clinicians should be aware of the varied presentation of CL. A detailed medical history and regular annual screenings are essential for the early detection of systemic involvement in CL, which can significantly reduce the risk of early mortality associated with this condition.

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Editor Query???

EQ1: Kindly check and confirm the edits