

## CASE REPORT

# Zosteriform pilar leiomyoma associated with uterine leiomyoma: A case report

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Email: [elishasht1@gmail.com](mailto:elishasht1@gmail.com)**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.

**KEYWORDS**

leiomyoma cutis, segmental

## 1 | BACKGROUND

Leiomyomas were first termed by Virchow in 1854 as “*tuberculum dolorosum*.”<sup>1</sup> Cutaneous leiomyomas are rare, benign tumors that may be sporadic or inherited, originated from smooth muscle fibers, and can be associated with various disorders.<sup>2</sup> It can be categorized into three subtypes depending upon the origin, arrector pili muscles (pilar leiomyoma), smooth muscle of blood vessels (angioliomyoma), and tunica dartos or labia majora (dartoic or genital leiomyoma).<sup>3</sup> Cutaneous leiomyomas are painful tumors that are more common in adults than children.<sup>4</sup> In 1880, Besnier classified leiomyomas as solitary or multiple.<sup>5</sup> When multiple, these can be grouped, linear, segmental, zosteriform, or dermatomal pattern.<sup>6</sup> 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.<sup>7</sup> 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 fibers of the tumor.<sup>7,8</sup>

Pilar leiomyomas can be transmitted as an 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).<sup>8,9</sup>

## 2 | OBSERVATION

A 36-year-old, married woman 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 3 years. These lesions initially appeared over right forearm and gradually increased in number to involve right arm and right upper chest (Figures 1 and 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

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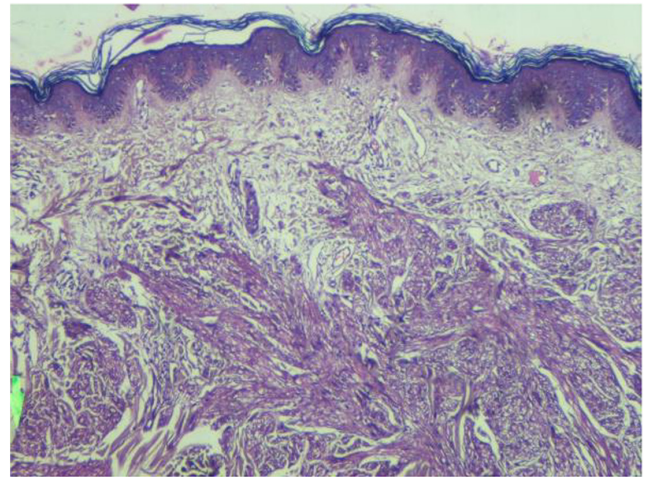


**FIGURE 1** Clinical pictures of cutaneous leiomyoma in zosteriform pattern involving right upper arm.

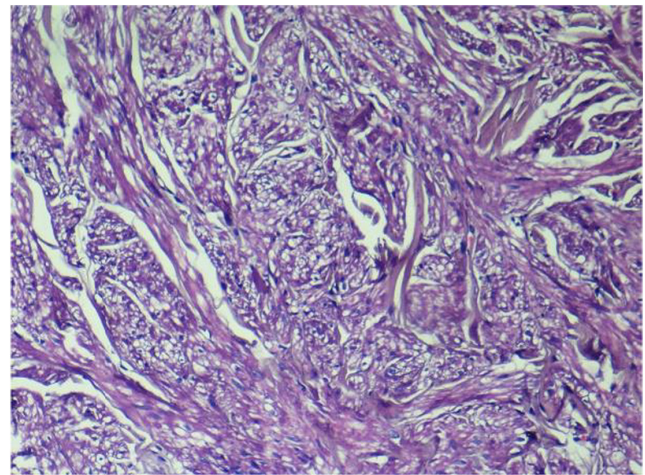


**FIGURE 2** Clinical pictures of cutaneous leiomyoma in right forearm.

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–C8) dermatome. Based upon history and clinical examination, we made differential diagnosis of leiomyomas, neurofibromas, and angioliomas. Routine hematological investigations including urea, creatinine, and urine examination did not reveal any abnormalities. However, ultrasound examination of abdomen and pelvis revealed a lesion measuring 19×18×13 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 (Figures 3 and 4).



**FIGURE 3** Low magnification view of cutaneous leiomyoma showing smooth muscles arranged in interlacing fascicles.



**FIGURE 4** Higher magnification view of cutaneous leiomyoma showing smooth muscles arranged in interlacing fascicles.

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

### 3 | DISCUSSION

Cutaneous leiomyomas are rare benign, painful tumors arising from smooth muscle fibers. 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.<sup>10</sup> 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.<sup>9,11</sup> 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, nonencapsulated 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 doxazosin and phenoxibenzamine, and gabapentin and topical 9% hyoscine hydrobromide. CO2 laser, cryotherapy, and electrocoagulation are alternative treatments.<sup>7</sup>

#### AUTHOR CONTRIBUTIONS

**Elisha Shrestha:** Data collection and management, preparing final version of manuscript. **Aditi Mishra:** Formal analysis; supervision; visualization.

#### ACKNOWLEDGMENTS

I would like to express my very great appreciation to Dr Aditi Mishra for her valuable and constructive suggestions during the planning and development of this research work. Her willingness to give time so generously has been very much appreciated. I would also like to thank the Department of Pathology of Dhulikhel Hospital for their constant support.

#### CONFLICT OF INTEREST STATEMENT

None.

#### DATA AVAILABILITY STATEMENT

The data that support the findings of this study are openly available in Clinical case reports.

#### ETHICAL APPROVAL

Hereby, I Dr Elisha Shrestha, MD consciously assure that for the manuscript Zosteriform pilar leiomyoma associated with uterine leiomyoma: A case report is fulfilled:

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**How to cite this article:** Shrestha E, Mishra A. Zosteriform pilar leiomyoma associated with uterine leiomyoma: A case report. *Clin Case Rep.* 2023;11:e6904. doi:[10.1002/ccr3.6904](https://doi.org/10.1002/ccr3.6904)