Somatic Soft Tissue Leiomyoma in an Elderly Patient: A Case Report and Review of Existing Literature

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Abstract

Leiomyomas of deep soft tissue have been traditionally divided into gynecologic and somatic leiomyomas. Somatic leiomyoma is exceedingly rare and usually has excellent outcome. We report a case of slow growing lower extremity somatic leiomyoma in a 63-year-old man. It has been predominantly reported to affect younger age group. Our patient is the oldest reported patient in the English language literature. Macroscopically, the specimen consisted of a nodular lesion with focal calcification, measuring 3.1 x 2.2 x 1.5 cm. Microscopically, this was a well-circumscribed, mature smooth muscle tumour with a fibrous pseudocapsule and dense calcification admixed with areas of hydropic and myxoid change, and mild parenchymal and perivascular hyalinization. The smooth muscle cells exhibited focal minimal atypia without increased mitotic activity or necrosis. All margins were clear of tumour. Immunohistochemically, the neoplastic cells were strongly and diffusely positive for smooth muscle markers. The case was reported as “somatic type smooth muscle tumour of soft tissue, probably benign”, and close clinical and radiological follow-up was recommended. The patient remains clinically recurrence-free four months after the diagnosis.

Introduction

Smooth muscle tumours have been traditionally divided into benign leiomyomas and malignant leiomyosarcomas. These tumours are relatively common in the genitourinary and gastrointestinal tracts and less frequent in the skin. Unequivocal leiomyomas of deep soft tissue are exceptionally rare compared to leiomyosarcomas and should be diagnosed using strict criteria derived empirically from the evaluation of soft tissue smooth muscle tumours. Two types of soft tissue leiomyomas have been described. The more common leiomyoma of gynecologic or uterine type occurs almost exclusively in women. The less common somatic leiomyoma arises in the deep somatic soft tissue of the extremities and shows no gender predilection. It usually measures several centimeters at presentation and contains calcifications in one-third of cases. The presence of calcifications has led to a spectrum of radiological diagnoses including “calcifying schwannoma”, “synovial sarcoma or “myositis ossificans”. Histologically, these lesions are composed of fascicles of well-differentiated smooth muscle cells with abundant eosinophilic cytoplasm. Rare cases show a predominantly clear cell appearance or psammoma bodies. By definition, somatic leiomyomas should not contain necrosis, significant atypia or mitotic figures (<1 mitosis/50 high-power fields [HPF]). These patients usually have excellent outcome.

We report a case of slow-growing somatic type smooth muscle tumour of soft tissue in the left calcaneus in a 63-year-old man.

Case Presentation

A 63-year-old man with a history of hyperlipidemia, hypertension, obstructive sleep apnea, type 2 diabetes
mellitus, chronic renal failure and valvular heart disease presented to orthopedic surgery clinic for assessment of a soft tissue mass on his left calcaneus. The mass had been growing over two decades. On clinical examination, the mass was soft, mobile and non-tender. X-ray and ultrasound imaging showed a soft tissue mass with calcification overlying the Achilles tendon insertion, closely approaching the distal tendon and measuring 2.3 cm in diameter (Figure. 1). The ultrasound features raised a differential diagnosis that included both benign and malignant soft tissue tumours such as ganglion, chronic bursitis and soft tissue sarcoma.

![Figure 1. X-ray (A) and ultrasound (B) imaging 2.3 cm soft tissue mass with calcification overlying the Achilles tendon insertion. Increase in vascularity in the soft tissue component of the lesion, and degenerative enthesopathy at the Achilles insertion in the calcaneus.](image)

The patient underwent excisional biopsy of the soft tissue mass. A longitudinal incision was made over the dorsal aspect of the ankle 2 cm above and below the lesion with a curvilinear incision to the lateral aspect of the mass. The lesion was resected en bloc leaving enough tissue to ensure competency of the skin. There were no intraoperative or postoperative complications. The patient remains well 4 months after the procedure. Macroscopic examination of the mass showed a nodular fragment of firm, yellow tissue encapsulated by a thin mobile membrane, with attached soft tan-grey tissue, measuring 3.1 x 2.2 x 1.5 cm. The cut surface of the specimen was focally calcified. The margins were inked. The specimen was fixed in 10% neutral buffered formalin, processed in the usual manner, and embedded in paraffin. Four micrometer thick sections were stained with hematoxylin-phloxine-saffron (HPS) stain and immunohistochemical stains were performed on a Leica Bond-III autostainer. Appropriate positive and negative controls were used.

Microscopy showed a well-circumscribed, mature smooth muscle tumour with a fibrous pseudocapsule, containing areas of hydropic and myxoid change (Figure. 2A), dense calcification (Figure. 2B), and mild parenchymal and perivascular hyalinization (Fig. 2C). There was no evidence of significant cytological atypia, increased mitotic activity or necrosis. All margins were clear of tumour. Immunohistochemically, the neoplastic cells were strongly and diffusely positive for desmin, smooth muscle actin (SMA) and muscle specific actin (MSA), further confirming the smooth muscle differentiation (Fig. 2D-F). Estrogen receptor, Bcl-2, S100, CD99 and CD34 were negative. The case was reported as “somatic type smooth muscle tumour of soft tissue, probably benign”. Although the tumour was completely excised, close clinical and radiological follow-up was recommended.
Discussion

Smooth muscle tumours are mainly classified into benign and malignant categories, with an additional borderline category of smooth muscle tumour of uncertain potential. Well-differentiated smooth muscle tumours are typically composed of spindle cells resembling non-neoplastic smooth muscle cells, containing ovoid or cigar-shaped nuclei with a blunt end, and variably eosinophilic cytoplasm. The cytoplasm sometimes exhibits longitudinal striations and rarely contraction bands. The distinction between leiomyoma and leiomyosarcoma is based on the presence of nuclear atypia, mitotic activity and coagulative type necrosis. However, hormone receptor positive uterine or non-uterine tumours in women can have substantially higher mitotic activity than peripheral soft tissue smooth muscle tumours and still follow a benign clinical course. The neoplastic cells are usually positive for SMA, MSA, desmin and heavy caldesmon in most cases. The MyoD1 family of myogenic regulatory proteins, such as MyoD1 and myogenin, are usually negative.

Two types of peripheral soft tissue leiomyomas have been described. Gynecologic leiomyoma is more common and typically affects women, whereas somatic leiomyoma is relatively rare and usually occurs in the deep somatic soft tissue of the extremities. Somatic leiomyomas have been the focus of case reports in the literature. Macroscopically, these tumours are fusiform to spherical, circumscribed masses, measuring 0.6-12.5 cm (mean 5.4 cm), tan-gray to white in colour, often with a rubbery consistency. Large smooth muscle tumours of deep peripheral soft tissues may be heterogeneous and thus should be sampled extensively to identify mitotically active or atypical foci. Complete excision and follow-up is recommended, and the designation of ‘smooth muscle tumour of uncertain biological potential, probably benign’ may be used if the biological potential cannot be determined with certainty. This problem occurs when only some criteria for malignancy have been met or when the tumours have developed in unusual settings. The location and size of the lesion in our patient was in keeping with the literature.

Somatic leiomyomas have a typical histologic appearance and immunophenotype of a non-uterine leiomy-
oma, often with stromal calcifications, hyalinization and virtually no mitotic figures. Unlike gynecological leiomyomas, somatic leiomyomas do not typically express hormone receptors. 2 Unusual and less common features include an epithelioid appearance; a cord-like arrangement of the smooth muscle cells similar to uterine leiomyomas; degenerated large, eosinophilic, rounded smooth muscle cells mimicking rhabdomyoblasts; degenerative mucoid-cystic changes; degenerative nuclear changes, similar to ancient schwannoma; and multiple necrobiotic granuloma-like nodules resembling giant rosettes 5. Rare cases contain numerous thin and thick-walled blood vessels, suggesting their origin from a pre-existing vascular malformation. 2 Our patient had a well-circumscribed bland-appearing spindle cell tumour with hydropic and myxoid change, calcification and stromal hyalinization with no evidence of atypical features. The diagnosis was supported by positive immunostaining for desmin, SMA and MSA.

Two case series have reported that somatic leiomyomas can occur from childhood to middle age (age range 14-62 years, mean 37 years, median 35 years 5; age range 6-55 years, mean 37 years, median 44 years 2), with similar occurrence rates in both sexes. 2, 5 Our patient is 63 years old and thus represents the oldest reported patient with this lesion. Four months after the initial diagnosis, he remains clinically recurrence free. This is consistent with the literature, although the number of somatic leiomyomas with extended follow-up is still quite small. In the largest case series all tumours showed benign behaviour (9 patients, median 4.5 years 5; 11 patients, mean 58.7 months 3).

In summary, the histological and immunophenotypic features of this left calcaneal spindle cell tumour in a 63 year old man were in keeping with a somatic leiomyoma. Given the rarity of these lesions and lack of large clinical studies, the lesion was reported as “probably benign”, even in the absence of significant atypia. Furthermore, close clinical and radiological follow-up would be prudent in such cases.

References


