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CASE REPORT

Fernando López-Barea · José L. Rodríguez-Peralto
Emilio Burgos · José González-López

Calcified leiomyoma of deep soft tissue. Report of a case in childhood

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Abstract This report illustrates a calcified leiomyoma of deep soft tissue in the left leg of a 6-year-old boy. The tumour was composed of spindle cells arranged in interlacing bundles, between which were multiple small and large areas of calcification. Tumour cells were positive for vimentin, desmin and smooth muscle actin. Ultrastructurally, the cells showed numerous pinocytotic vesicles and bundles of intracytoplasmic filaments with smooth muscle dense bodies. Only four calcified leiomyomas have been previously reported in the deep soft tissues of limbs. Here we report a new case and suggest a new pathogenetic scheme involving alkaline phosphatase in the origin of these calcifications.

Key words Soft tissue tumour · Leiomyoma
Calcification

Introduction

Leiomyomas represent only 4.4% of benign tumours in soft tissues [6] and excluding those arising in cutaneous and subcutaneous tissues, leiomyomas in the extremities are very unusual [5]; seven such cases have been reported in the deep tissues of limbs [1, 2, 5, 7, 10], and only four of these lesions were calcified. Here we report an unusual calcified deep leiomyoma in a leg, and discuss the diagnostic and histogenetic aspects of this case.

F. López-Barea (✉) · E. Burgos
Department of Anatomical Pathology La Paz Hospital,
Paseo de la Castellana 261, E-28046 Madrid, Spain

J. L. Rodríguez-Peralto
Department of Anatomical Pathology, 12 de Octubre Hospital,
Madrid, Spain

J. González-López
Department of Traumatology, La Paz Hospital, Madrid, Spain

Case report

A 6-year-old boy was referred to La Paz Hospital for evaluation of a swelling in his left leg causing continuous pain. He had had slight traumatic antecedent in this area 6 months before. Physical examination revealed partial limitation of motion of the left leg, attributable to his pain. On palpation, a deep-seated 6×4 cm, fusiform mass was noted. The lesion was fixed to the underlying skeletal muscles without inflammatory signs. The pain was continuous, unrelated to activity and exacerbated under palpation. Routine laboratory data including the serum calcium, phosphorus and alkaline phosphatase levels were within normal limits.

Roentgenograms showed an intramuscular mass located in the posteroexternal area of the upper medium third of the left leg. The lesion was sharply circumscribed, stippled with multiple areas of calcification and not connected with the nearby bone as demon-

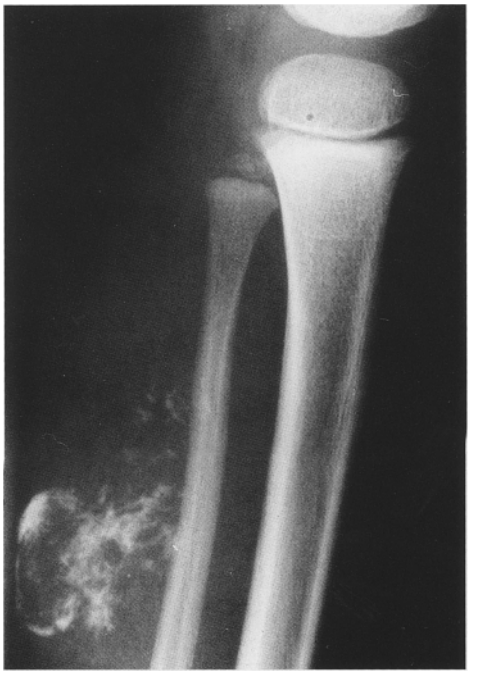

Fig. 1 Lateral radiograph of the left leg discloses a deep soft tissue calcified tumour clearly separated from the bony shaft

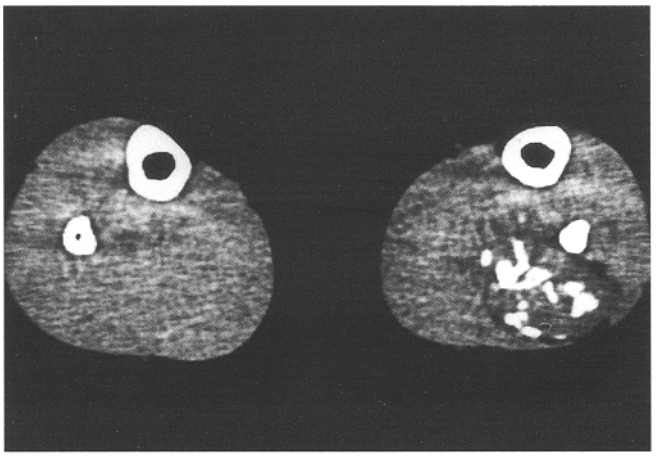

Fig. 2 Computed tomogram shows a deep soft tissue tumour sharply circumscribed and separated from the nearby bone. The mass displays multiple and scattered calcified areas

strated in the lateral radiograph (Fig. 1). The radiological diagnosis was calcified haematoma or myositis ossificans.

Computer tomography scan revealed a posteroexternal left leg mass adjacent to the peroneal shaft, but clearly separated from the nearby bony structures. The tumour was sharply circumscribed with multiple spotty calcifications (Fig. 2). The magnetic resonance image demonstrated a solid, well-outlined 6×3.5 cm tumour located within the soleus muscle. The lesion was isointense compared with neighbouring muscles on T1-weighted images and hyperintense on T2-weighted images. At surgery, the mass was found within soft tissue without any relationship to bony structures. After a diagnostic biopsy, complete resection with surrounding soft tissue was performed. The patient is currently well without recurrence 1.5 years after the excision.

The excisional biopsy consisted of a well-circumscribed 5.5×3.5×3 cm ovoid mass, surrounded by soft tissues. On section, a white solid tumour stippled with multiple yellow calcifications was observed (Fig. 3A, B). Microscopically, the lesion consisted of a proliferation of spindle cells arranged in interlacing bundles. Tumoural cells showed abundant eosinophilic cytoplasm with elongated blunt-ended nuclei (Fig. 4A). Between the muscle cells there was a variable amount of fibrous tissue and calcification. The calcification ranged from big irregular masses to spotty round psammoma bodies (Fig. 4B). Some of the larger masses were surrounded by a foreign body granulomatous reaction. The vascular pattern was inconspicuous, with thin-walled vessels. No mitotic activity or necrosis was present.

Histochemical staining demonstrated intracellular alkaline phosphatase, especially in subsarcolemmic regions. The tumour cells showed strong immunoreactivity to vimentin, desmin and smooth-muscle actin (SMA).

Ultrastructurally, the tumour consisted of spindle cells arranged in fascicles. The cells showed numerous pinocytotic vesicles, well-defined basal lamina and fusiform nuclei. The cytoplasms were filled with numerous filament bundles with smooth muscle dense bodies that pushed the cytoplasmic organelles to the periphery (Fig. 5).

Discussion

Leiomyoma is a benign, usually solitary, tumour of smooth muscle origin that most often involves those organs of the body composed of smooth muscle, such as the uterus or gastrointestinal tract. When found in soft tissue it usually involves dermis and subcutis and very rarely the deep soft part. It may be classified as cutaneous leiomyoma, vascular leiomyoma, or leiomyoma of deep soft tissue [3]. Eleven deep soft tissue leiomyomas have been described (Table I) and only seven were in the extremities: four in the thigh [1, 5, 7], two in the leg [2, 10] and one in the arm [1]. The clinicopathologic characteristics of the deep soft tissue leiomyomas including our case, are presented in Table I.

The mean age of patients with deep soft tissue leiomyoma was 25 years (range: 3–74-years-old) and they

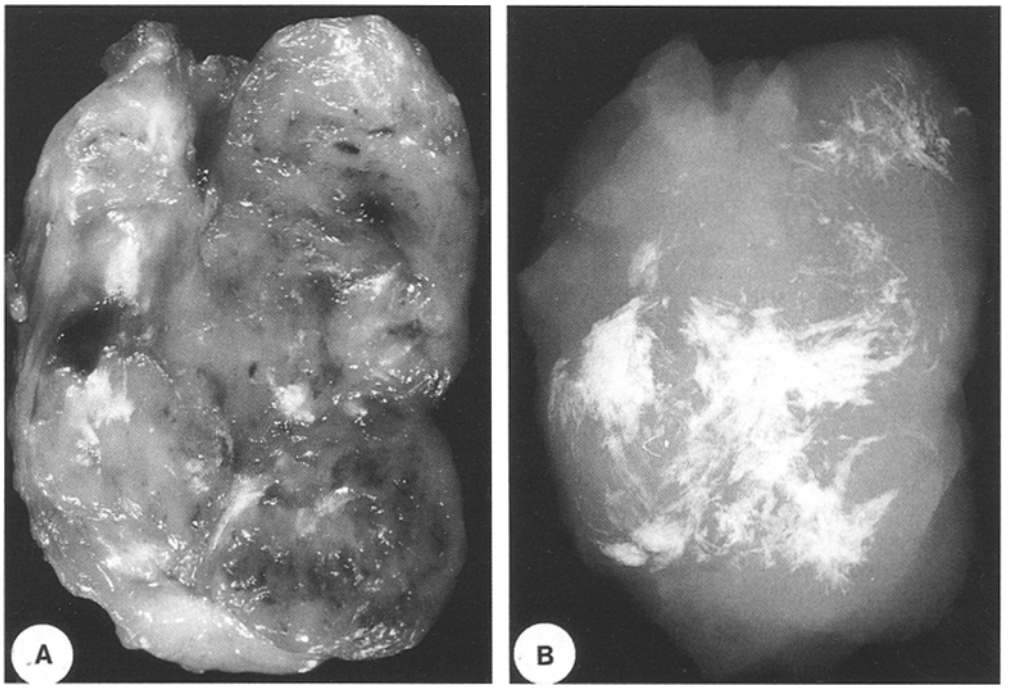

Fig. 3A Sectioned surface of the tumour composed of a solid white mass with calcified zones. **B** Radiograph of the surgical specimen

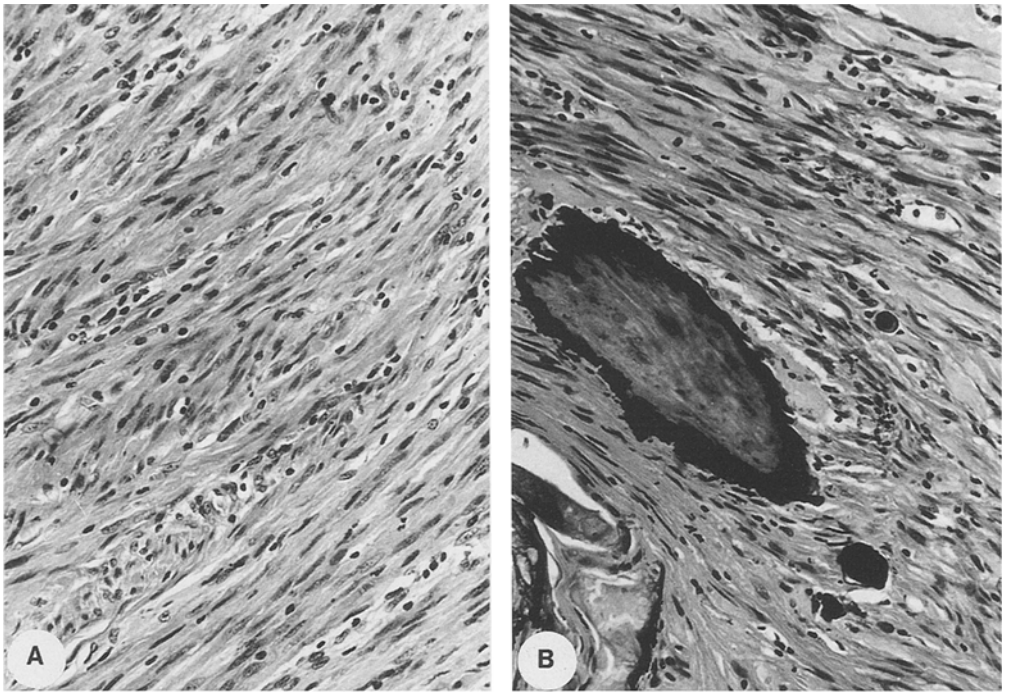

Fig. 4A Microscopic view of the tumour. Interlacing spindle cell fascicles with elongated blunt-ended nuclei, haematoxylic and eosin (H&E) ×120. **B** Haphazard deposits of amorphous calcium within smooth muscle proliferation, H&E ×120

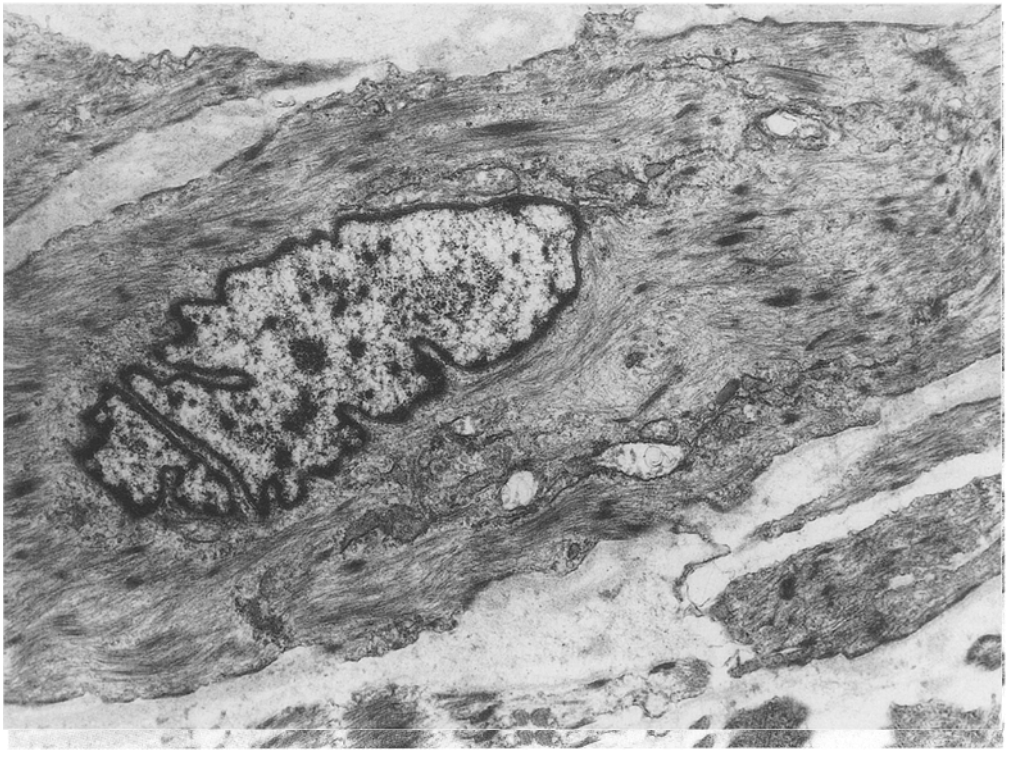

Fig. 5 Tumour cell containing bundles of filaments with smooth muscle dense bodies. Electron microscopy, ×3,000

were younger than patients with superficial angioleiomyomas (47-years-old) [6]. Deep soft tissue leiomyomas affect men more often than women (1.4:1), in contrast to their superficial counterpart (1:1.7). Pain was one of the most frequent clinical symptoms in superficial leiomyomas (58%), however, only 5 of the 12 deep tumors (41%) were painful. The lower extremity is the most frequent location in both groups of tumours [4, 6], and all deep leiomyomas have been found within skeletal muscles. The mean diameter of intramuscular leiomyomas is 7 cm at the time of surgical removal; while superficial tumours measured between 0.2 and 4.3 cm, 78% of the cases were smaller than 2 cm. Seven of the 11 (58%) deep leiomyomas showed calcification in contrast to 1.9% of the cutaneous superficial ones. All these clinical data clearly differentiate superficial from deep

Table 1 Relevant details in 12 cases of deep soft tissue leiomyomas

Author	Case number	Age (years)	Sex	Pain	Length of history	Site	Calcification	Size (cms)
Bulmer	1	68	Female	No	30 years	Thigh	Yes	12
	2	7	Male	No	3 weeks	Arm	No	3
Duchateau	3	59	Female	Yes	43 years	Leg	No	3
Goldman	4	5	Female	No	5 months	Thigh	Yes	7.5
Herrlin	5	74	Male	No	2 months	Ischiorectal	No	10
	6	19	Female	Yes	3 years	Thigh	Yes	6
	7	29	Female	No	1.5 years	Trapezius	No	8
	8	20	Male	No	2 months	Thigh	No	9
Ledesma	9	6	Male	No	–	Axilla	Yes	5
	10	3	Male	No	6 weeks	Subscapular	Yes	8
Ross	11	6	Male	Yes	Some weeks	Leg	Yes	7
Present case	12	6	Male	Yes	6 months	Leg	Yes	5.5

leiomyomas. Thus, deep soft tissue leiomyoma involves men more frequently than women, is more common in patients younger than 10-years-old, usually reaches a large size and frequently shows calcification. From the clinical point of view, the differential diagnosis includes all lesions containing calcification (haematoma, myositis ossificans, calcifying fibrous pseudotumour, synovial sarcoma, extraskeletal osteosarcoma and mesenchymal chondrosarcoma).

Microscopically, classic leiomyoma is composed of interlacing bundles of smooth muscle cells with elongated blunt-ended nuclei and abundant eosinophilic cytoplasm. Nevertheless, when the tumour displays some atypical features such as nuclear palisading, cytoplasmic perinuclear vacuolization, abundant clear cells or myxoid change [3], the right diagnosis may be difficult. Moreover, 58% of these deep soft tissue leiomyomas show calcification. These calcareous deposits may be small, big or psammoma-like bodies. The biggest ones may develop foreign body reactions similar to those in tumour calcinosis [3]. Demonstration of the smooth muscle nature of the cells by ultrastructural or immunohistochemical studies are keys in the diagnosis.

The precise nature and pathogenesis of calcification in deep leiomyoma is uncertain. Classically, they have been considered dystrophic changes following tumoral haemorrhage, necrosis or liquefaction [1, 3, 7, 8]; but the case reported here does not display haemorrhage or necrosis. Alkaline phosphatase has been detected in the present case and also isolated and purified from uterine myoma [9] suggesting that alkaline phosphatase may play a role in the pathogenesis of calcification.

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