



Intraosseous leiomyoma in a rib A case report

Ali Kursad GANYUSUFOGLU, Kemal AYALP, Cagatay OZTÜRK, Unal SAKALLIOGLU, Ozden OZER

From Istanbul Bilim University, Florence Nigthingale Hospital, Istanbul, Turkey

Intraosseous leiomyomas are very rare. To the best of our knowledge, this is the first published case of primary intraosseous leiomyoma in a rib. This rare benign tumour should be included in the differential diagnosis of any relatively small intraosseous lesion with benign imaging findings, but with gradually worsening, long-standing pain.

Keywords : intraosseous leiomyoma ; rib ; smooth muscle tumour.

INTRODUCTION

Leiomyomas are benign tumours originating from smooth muscle. The uterus is the most common site of origin, but they may also originate from the gastrointestinal tract, the skin and the urinary system. Although leiomyomas constitute 70-80% of all benign mesenchymal tumours, intraosseous leiomyomas are very rare (6). Other than the secondary involvement of bone in disseminated leiomyomatosis cases, 15 primary intraosseous leiomyomas have been reported in the oral cavity, 6 in the skull, 4 in the peripheral skeleton and 3 cases in the pelvic bones (1,2,6-9,13,14). To the best of our knowledge, this is the first published case of a primary intraosseous leiomyoma occurring in a rib.

CASE REPORT

A 43-year-old woman was admitted to our hospital with pain localized to the left rib cage over the

past eleven years. The pain was initially insidious, but it had become more severe about two years before her consultation. Intercostal neural block was performed in another center, one year previously, but it failed to relieve the pain. Her pain worsened over the last three months, interfering with her normal daily activities. Her past medical history was unremarkable and gynaecologic routine controls were normal. On physical examination, extreme pain was detected by the palpation of the costochondral junction of the left lower ribs ; there was no palpable mass.

On admission, her readily available standard rib radiographs and MR imaging of the dorsal vertebral column did not disclose any pathology. MR imaging (Symphony 1.5 Tesla, Siemens, Erlangen, Germany) was done, with a marker placed on the

-
- Ali Kursad Ganyusufoglu, MD, Consultant.
 - Kemal Ayalp, MD, Consultant.
 - Cagatay Oztürk, MD, Consultant.
 - Unal Sakallioğlu, MD, Consultant.
 - Ozden Ozer, MD, Consultant.

Istanbul Bilim University, Florence Nigthingale Hospital, Istanbul, Turkey.

Correspondence : Cagatay Oztürk, MD, Istanbul Bilim University, Florence Nigthingale Hospital, Abide-i Hurriyet Caddesi, No : 290, Şişli, Istanbul, Turkey.

E-mail : cgtyztrk@yahoo.com

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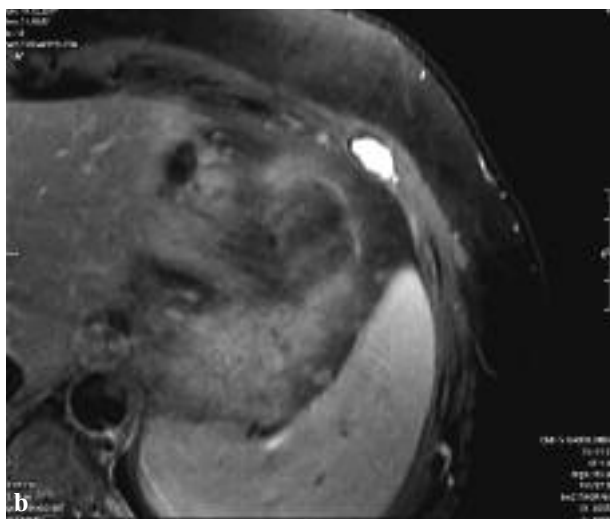
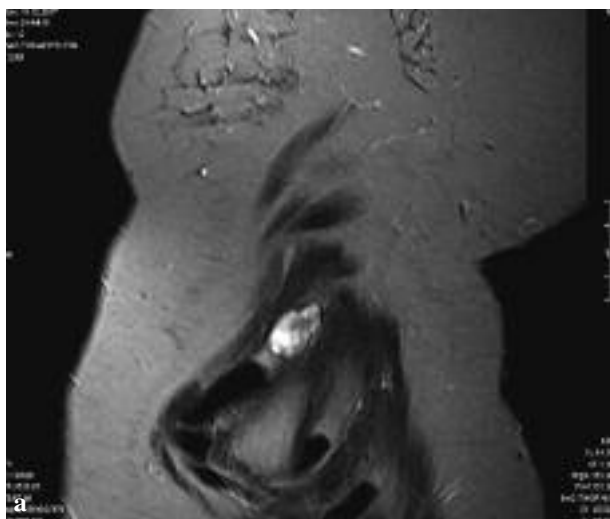


Fig. 1. — a) Sagittal STIR image shows that the lesion has high signal. b) Axial post-contrast fat-suppressed T1-weighted image shows homogenous enhancement of the mass.

clinically tender point. An intramedullary 2.5×1.5 centimeter mass was detected in the distal part of the 7th left rib, 5 millimeters proximal to its costochondral junction. The lesion was mildly expansile and well marginated (fig 1 a-b). Its signal intensity was low, similar to muscle on T1-weighted images. It showed prominent high signal on T2-weighted images. Homogenous contrast enhancement was present. There was no perilesional oedema and no extension into the soft tissue. CT



Fig. 2. — Axial CT image shows focal cortical discontinuity (arrow).

examination (Sensation 16, Siemens, Germany) was performed to assess the lesion matrix and cortical continuity in greater detail. It revealed a soft tissue mass of 50 Hounsfield unit (HU), delineated by a sclerotic rim, with fine internal septation (fig 2). Although cortical thinning with focal disruption of the lateral cortex was detected, CT imaging also confirmed that the borders of the lesion were well defined. It did not contain any calcification. There was no associated periosteal reaction. CT and MR imaging did not demonstrate a pathological fracture. The constellation of findings with the small size of the mass, long duration of symptoms and sclerotic rim were consistent with a benign lesion. In the differential diagnosis, benign rib tumours or tumour-like lesions such as fibrous dysplasia or enchondroma were considered. Although it is very rare, intraosseous neurilemmoma, which is a painful benign osseous tumour, was also included in the differential diagnosis. Surgical excision was recommended to establish a definite diagnosis.

Complete removal of the mass was performed by segmental resection of the left 7th rib. The patient was pain free postoperatively. The resection specimen was represented by multiple segments of rib with the attached soft tissue mass. The largest and smallest pieces measured $3 \times 1 \times 0.5$ cm and $1.8 \times$

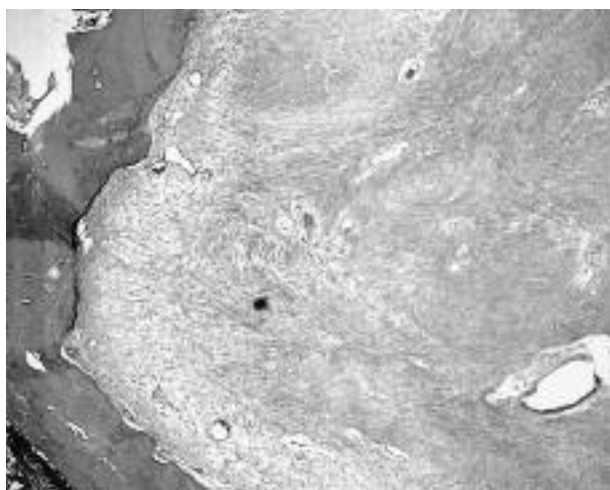


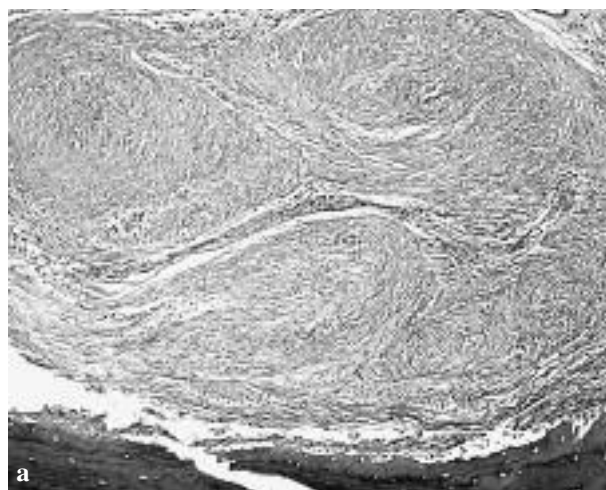
Fig. 3. — The lesion is a soft tissue mass, completely surrounded by lamellar bone.

1.5 × 0.8 cm respectively. Sections were studied following fixation in formalin and brief decalcification in formic acid. H&E stained sections revealed an intraosseous, well-circumscribed mass (fig 3), characterized by bundles of smooth muscle, consistent with intraosseous leiomyoma (fig 4a-b). Individual cells exhibited blunt nuclei without cellular atypia, mitotic activity or necrosis, suggesting a benign lesion. Smooth muscle actin showed diffuse, strong staining to the muscle bundles, confirming smooth muscle differentiation in the lesion (fig 5).

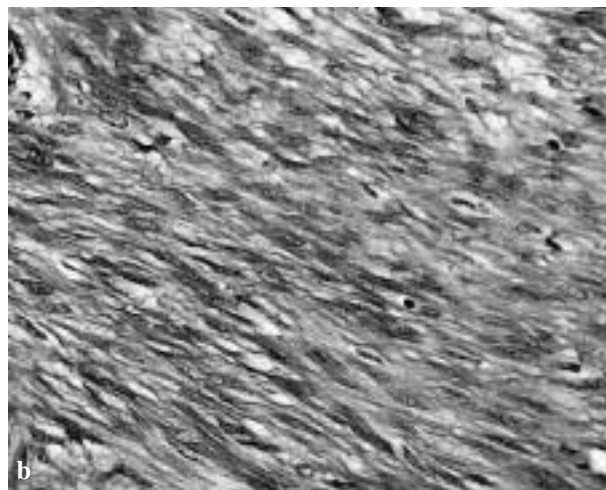
DISCUSSION

Intraosseous leiomyomas are exceedingly rare benign tumours. Most of the reported cases were primary intraosseous leiomyomas, but cases of disseminated leiomyomatosis secondarily involving bone have also been reported in literature (3,10-12).

In primary intraosseous leiomyoma, smooth muscle cells of vessel walls are claimed to be the site of origin (6,8,14). In disseminated leiomyomatosis, lesions are mostly seen in the axial skeleton, whereas primary intraosseous leiomyomas are most common in the oral region. Fifteen oral intraosseous leiomyomas have been reported, 13 of them involving the mandible. Three cases have



a



b

Fig. 4. — a) The lesion is composed of intersecting bundles of smooth muscle. b) At high power magnification, the cells look blunt with no atypia, mitosis or necrosis.

been reported in the pelvis. Rest of the reported cases reveal involvement of the appendicular skeleton, including two cases in the tibia, one case in the ulnar styloid and one case in the fibula. To the best of our knowledge, our case is the first reported case of a primary intraosseous leiomyoma in a rib.

Our patient presented with a clinical history and imaging findings, compatible with a benign tumoral lesion of the rib. The lesion was small in contrast to the long standing symptoms. Imaging findings such as sclerotic rim, well-defined borders without extension into extraosseous soft tissues suggested a

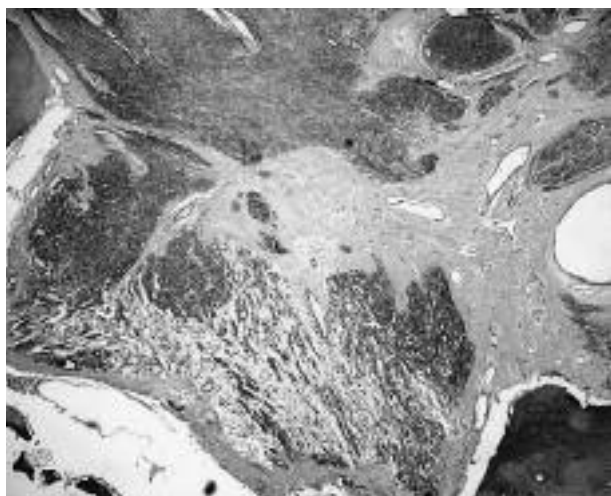


Fig. 5. — Smooth muscle actin shows strong positivity, supporting the diagnosis of leiomyoma.

benign nature. Benign mass lesions of the rib such as fibrous dysplasia, enchondroma, haemangioma and intraosseous neurilemmoma were considered in the differential diagnosis prior to resection. However, no definitive diagnosis could be established. Intraosseous neurilemmoma is a very rare painful benign tumoral lesion of bone, having clinical and imaging findings similar to our case, but it is mostly located in the mandible and, to the best of our knowledge, no rib involvement has been reported before (5). Fibrous dysplasia is the commonest benign tumour of the ribs (4). While its imaging findings are variable, monostotic fibrous dysplasia of the rib could appear as an expansile, well-margined osteolytic lesion, similar to our case. However, fibrous dysplasia is usually asymptomatic and is detected incidentally. It becomes symptomatic only if it is large enough to cause local pressure or in case of pathological fracture. Neither of these was present in our case.

Enchondroma, as the second most common benign rib tumour, was also considered in the differential diagnosis. It presents as an expansile, osteolytic mass with cortical thinning, exhibiting similar MRI signals. However, our lesion lacked any calcifications. Haemangioma is another benign lesion of rib that was considered. Although the expansile, well-defined osteolytic nature of our

lesion would fit with this entity, the typical coarse trabecular pattern was absent and the extreme pain which our patient experienced was not compatible with this diagnosis.

The ribs represent an unusual location for osteoid osteoma and osteoblastoma. The osteoid osteoma nidus causes a marked sclerotic reaction even in neighbouring ribs. Partial or extensive calcification and perilesional oedema which are the expected imaging findings of osteoblastoma, were not detected in our case. Eosinophilic granuloma appears as an erosive osteolytic lesion in patients between the ages of 5-15 years. Aneurysmal bone cyst is also seen in younger age groups as a markedly expansile multiseptated lesion with a potential for extraosseous soft tissue extension. Also, MR imaging of our case did not demonstrate typical fluid-fluid levels. Malignant rib tumours such as metastasis, chondrosarcoma, Ewing's sarcoma and plasmacytoma were not considered in the differential diagnosis, because there was no destructive pattern, no extraosseous soft tissue mass nor any sign of malignancy.

Pathological examination of the resected mass instantly ruled out all above mentioned entities. Bundles of smooth muscle strongly staining for smooth muscle actin were diagnostic of leiomyoma. The typical histomorphology of fibrous dysplasia, characterized by osteoblastic rimming, irregular bony spicules simulating Chinese letters were not seen. The mass was paucicellular without formation of bone or cartilage, ruling out tumours of osseous or chondroid origin. The vessels present, were thick walled with no arborization or lobular configuration to suggest a haemangioma. The ribs are a typical location for Ewing sarcoma which is a small round blue cell tumour of young age. That was easily ruled out by lack of a small round blue cell infiltrate.

Our case shares similar clinico-radiological characteristics with the cases published by Laffosse *et al* (6). They reported one case in the distal metaphysis of the fibula and one in the pelvis. Imaging characteristics of those lesions were suggestive of benign osseous lesions, non-ossifying fibroma and histiocytofibroma, respectively, but the presence of gradually worsening pain was inharmonious with

these diagnoses, as in our case. Although fibrous dysplasia was the main differential diagnosis, the extreme pain reported by the patient hampered this diagnosis. Zikria *et al* (14) also reported another benign intraosseous lesion of the distal ulna with marked symptoms.

In conclusion, although intraosseous leiomyoma is a very rare entity, it should be included in the differential diagnosis of any relatively small intraosseous lesion with benign imaging findings, but with long-standing and gradually worsening pain.

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