Journal of Bone Oncology 3 (2014) 80–83

Contents lists available at ScienceDirect

Journal of Bone Oncology

journal homepage: www.elsevier.com/locate/jbo

Case Report

Solitary intraosseous myofibroma of the tibia in an adult patient: A case report

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ARTICLE INFO

Available online 30 September 2014

Keywords:
Myofibroma
Intraosseous
Tibia
Adult

ABSTRACT

Myofibromas are mesenchymal tumors showing myofibroblastic differentiation and found most frequently in the head and neck region. While several cases of myofibromas have been reported in adults, they have not been described in long bones of the appendicular skeleton. We describe an otherwise healthy young woman who presented with a progressive incapacitating pain in her right shin. Imaging studies revealed a well-circumscribed osteolytic lesion with slight marginal sclerosis confined to the proximal tibia metaphysis, without a soft tissue component. Surgical intervention was performed and histological examination identified a myofibroma.

This case represents an extremely rare occurrence of an intraosseous myofibroma involving a long bone in an adult patient. Although solitary myofibroma is a rare lesion in the skeletal bones of adults we believe it should be included in the differential diagnosis of a solitary lytic mass in bone, especially if it is associated with pain.

1. Background

Solitary adult myofibromas are infrequent lesions with a predilection for the head and neck [1,2]. The lesion is considered to be completely benign but there is the potential for it being confused with more aggressive spindle cell tumors. Histologically, myofibroma has two characteristic microscopic components: (a) a central, highly vascular and cellular, hemangiopericytoma-like component and (b) coalescent clusters and bundles of spindle-shaped myofibroblasts [5].

Myofibromas usually occur as painless solitary masses, most often in the skin and subcutaneous tissues of the head and neck [3]. Most of these tumors present prior to the age of two years, but may be observed in older children and even in adults [4]. Oudijk et al. have investigated 97 cases of MF and 17 cases of MMF and have shown that these lesions occur predominantly, but not exclusively, in infants and children [3].

There are three distinct presentations [5]: solitary, multicentric without visceral involvement and multicentric with visceral involvement. The solitary form tends to occur predominately in males [6] and is typically identified in the dermis, subcutis or deep soft tissues. The reported incidence of solitary osseous myofibromatosis is rare [2,7–10]. The distribution is predominantly on the head, neck and torso, with only a rare involvement of the extremities [4,5]. Wu et al. recently reported two cases of solitary osseous myofibromas in the upper extremities of two female infants older than two years of age [1].

There are a few reports of myofibromas in adults [2,3,7,9,11–15]. Daimaru et al. reported five cases of myofibromas in adult patients [13]. The tumors clinically presented as superficial, painless, and slowly enlarging nodules, usually of more than 10 years' duration, that occurred in the upper (two cases) and lower (two cases) extremities or the buccal mucosa (one case). However all of these tumors involved the dermis and subcutis, and did not develop in deeper structures.

Intraosseous myofibromas were described in children and in adults. Still the occurrence of myofibromas in long bones is extremely rare, even in the pediatric population, and they usually occur in craniofacial bones.

We present the clinical, histologic, and radiographic findings of an intraosseous myofibroma recently diagnosed in a 23 year old woman at our hospital. To our knowledge, an adult with such an intraosseous lesion myofibroma has not been previously reported in the world literature.

2. Case report

A previously healthy 23-year-old woman was referred to our outpatient clinic with pain around her left knee. The pain had

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http://dx.doi.org/10.1016/j.jbo.2014.06.002

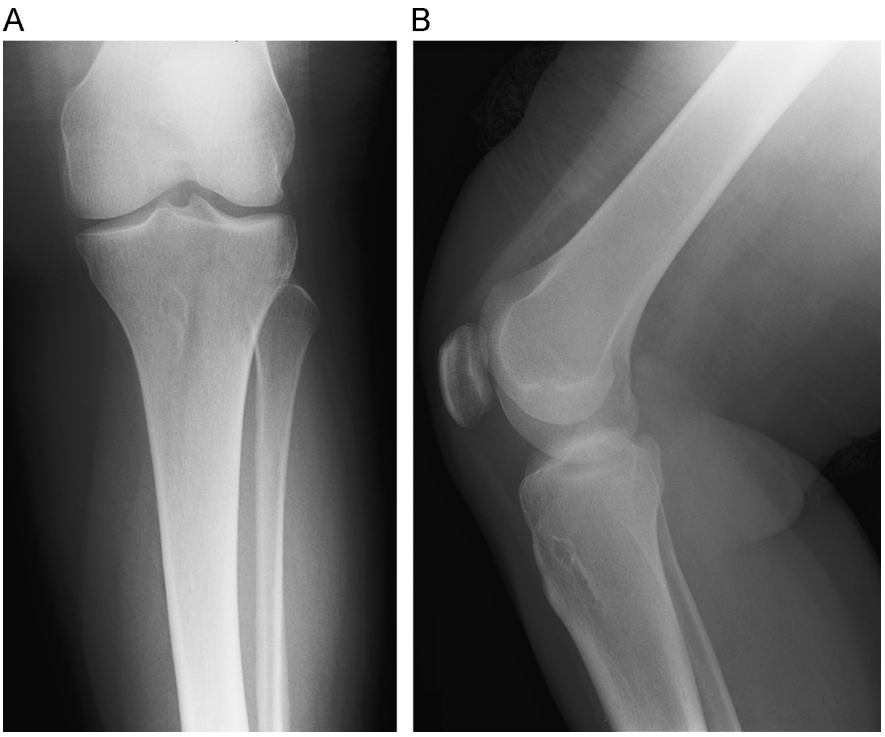

Fig. 1. Anteroposterior (A) and lateral (B) radiographs of the proximal tibia demonstrating a well-bordered lytic lesion with a sclerotic rim.

started 4 years before and steadily worsened. During the last year she had been experiencing night pain as well.

The patient did not feel any muscle weakness in her legs and the pain did not radiate to her foot. She denied any recent febrile illness, loss of weight, or loss of appetite. There was no history of trauma or infection, local injection, foreign body, or wound. Family history and medical history were otherwise unremarkable. Physical examination at the time of her visit revealed mild tenderness to palpation of the antero-medial proximal tibia, with no redness, fluctuation or ulcerations of the overlying dermis. The knee had no effusion and demonstrated normal range of motion. Normal deep tendon reflexes were elicited in both legs. Sensation and strength were normal in the entire extremity.

Levels of both erythrocyte sedimentation rate (ESR) and C-reactive protein were within normal limits.

Radiographs of the tibia revealed a well-bordered lytic lesion with a sclerotic rim (Fig. 1).

Computerized tomography (CT) scan of the proximal tibia showed an eccentric, purely lytic mass expanding into the proximal tibia with marginal sclerosis (Fig. 2). The lesion involved the anteromedial cortex, and yielded no evidence of a significant abnormality in the surrounding soft tissues. To rule out further tissue involvement a bone scan and a chest radiograph were performed, which failed to demonstrate any visceral or distant involvement.

A CT guided biopsy revealed a spindle cell tumor with a differential diagnosis of a leiomyoma, myofibroma or a myofibroblastic tumor. Under general anesthesia, the patient underwent curettage with fluoroscopic guidance. The tumor was identified in the anteromedial tibial metaphysis. Macroscopically, the tumor presented as firm gray-white fibrous nodule. The specimen on histological examination was reported as myofibroma.

Histologically, the cells had a characteristic spindle shaped fibroblast appearance with pale pink cytoplasm and elongated nuclei when stained with hematoxylin and eosin (Fig. 3A, B). On immunohistochemical examination, the tumor cells were diffusely immunoreactive for smooth muscle actin (Fig. 3C). Immunostaining for Ki-67, a proliferation marker, showed a low proportion of Ki-67 positive cells, indicating a low tumor proliferation activity (Fig. 3D).

On the basis of both pathologic and radiologic findings, we made a diagnosis of solitary myofibroma originating in the proximal tibia. The patient has been well without disease for 8 months following the operation and free from the pain.

3. Discussion

To the best of the authors' knowledge there are no previous reports of a solitary myofibroma arising within a long bone in an adult patient. Most previous reports of solitary myofibromas occurring at intraosseus sites have been in patients under 18 years of age.

Furthermore most of the previously described cases with a solitary lesion affecting bone involved the craniofacial bones [2–7], and only a few cases affected the appendicular skeleton. Several affected extra-craniofacial sites have been sporadically reported, for example: femur [8], tibia [9–11], humerus [1], ulna [12,1], and clavicle [13]. The age of the patients was usually less than 2 years [3], and no adult case has been reported, to our knowledge.

Konishi et al. reported the only known adult case of a solitary myofibroma affecting a lumbar vertebrae in a 33 years old male [14]. They described an expansile lytic lesion with a sclerotic rim located in the right pedicle of L1 vertebrae. As in our case, their patient sough medical attention due to severe pain in his lower back and abdomen and was pain free after excising the tumor.

Other cases described in adult patients were confined to non-osseous sites, like subcutaneus or intramuscular tumors [15–18].

The differential diagnosis of myofibroma is extensive. Microscopically, the lesion may bear a resemblance to nodular fasciitis, neurofibroma, fibrous histiocytoma, desmoid tumor, lipofibromatosis, other fibromatoses, inflammatory myofibroblastic tumor, congenital infantile fibrosarcoma, and various types of sarcoma [5]. Radiologically, the differential diagnosis may include

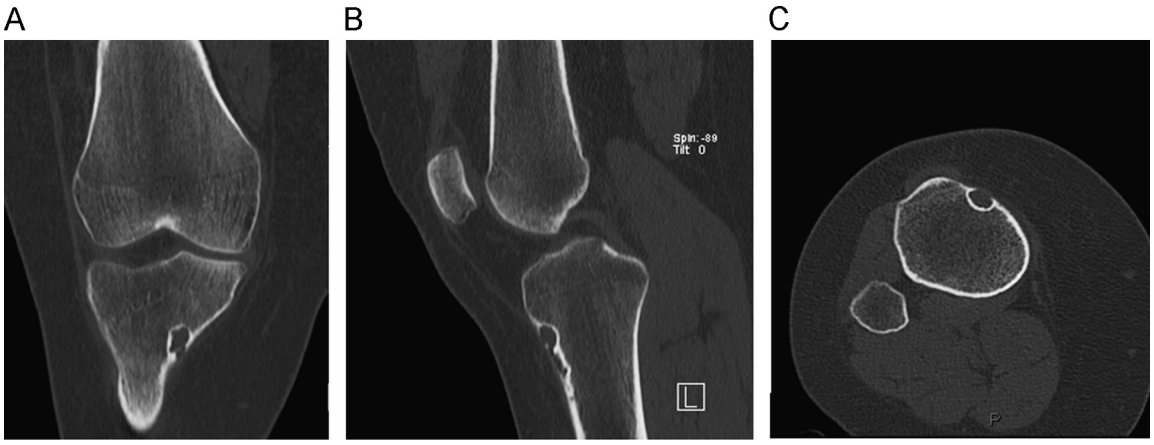

Fig. 2. Computerized tomography scan axial (A), sagittal (B) and coronal (C) images showing an eccentric, purely lytic mass expanding into the proximal tibia with marginal sclerosis. The lesion involves the anteromedial cortex, and has no soft tissue component.

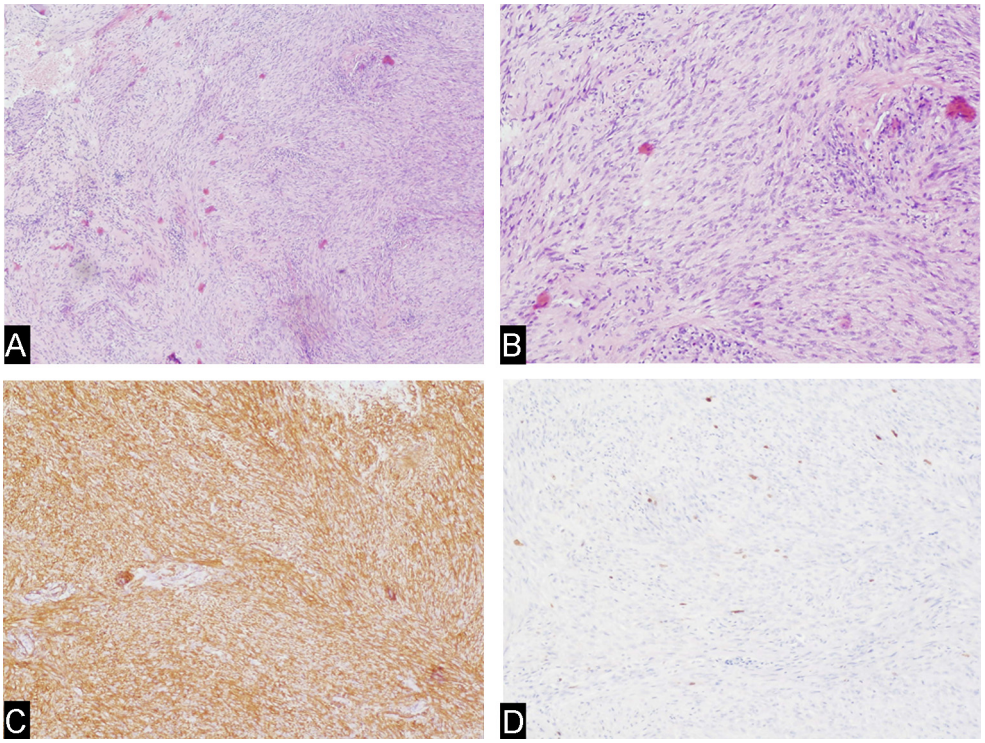

Fig. 3. Histopathology from the intraoperative specimen (A). At low power, a nodular arrangement is observed with hypocellular and hypercellular spindle cell areas in a hyalinized matrix (stain, hematoxylin and eosin, magnification 4 ×). (B) The nodule is composed of myoid spindle cells, having eosinophilic fusiform cytoplasm and spindling nuclei. Hyalinized pinkish matrix is present (stain, hematoxylin and eosin, magnification 10 ×). (C) Immunohistochemistry demonstrating a smooth muscle actin positivity within the tumor (immunoperoxidase–haematoxylin, magnification 10 ×). (D) Nuclear immunostaining for Ki-67, a cell proliferation marker, was rare and accounted for less than 1% of tumor cells (magnification 10 ×).

non-ossifying fibroma, fibrous dysplasia and histiocytosis X [8]. Given the patient's age and the lesion location, this lesion could be easily confused with a non-ossifying fibroma. Nevertheless our primary concern was the patient's worsening pain, which led us to offer her a surgical treatment.

As observed in previously reported cases, the plain radiograph and CT for our case showed a sharply demarcated, non-specific lytic mass with a sclerotic margin [1,14]. Radiological examinations can outline the tumor extent but are not sufficiently specific to allow definitive diagnosis, which requires histopathologic examination. Our case revealed the typical pathological features of myofibroma as described above.

In the majority of cases the histology of myofibroma is distinctive, and additional studies are not needed [5]. However, immunostaining and clinical signs can support the diagnosis and differentiate it from the aforementioned pathologies.

Surgical excision of solitary bone lesions usually results in cure [14]. Spontaneous regression may occur in soft tissue lesions as well as is solitary bone lesions [14].

In summary our patient was found to have a solitary lesion with a progressive clinical course, for which local excision appeared sufficient. The outcome was favorable.

Although solitary myofibroma is a rare lesion in the skeletal bones of adults we believe it should be included in the differential

diagnosis of a benign-appearing, solitary lytic mass in bone, especially if it is associated with pain.

Conflict of interest statement

The authors declare that there are no conflicts of interest.

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