Intraosseous Leiomyoma of the Tibia. A Case Report

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Abstract

Leiomyomata are infrequently seen in the extremities and rarely seen in the bone. It is usually presented by a gradually increasing pain with non-specific radiological findings, and could be a differential diagnosis for wide range of bone tumors. We report a case of a 73-year-old Japanese female patient with a painful intraosseous leiomyoma involving the proximal tibia. The patient had undergone tumor excision with wide margin, immediate weight bearing was allowed, pain had been relieved and the patient was satisfied with no recurrence, malignant change, distant metastases or functional impairment. We reviewed all published cases of intraosseous leiomyomata in English literature. Diagnosis of Intraosseous leiomyoma of the extremities is difficult due to extreme rarity of the tumor and absence of pathognomonic radiological sign in X-ray, CAT, or even MRI. While the exact diagnosis is only achieved by histopathological examination and with immunohistochemistry stains, which can differentiate it from malignancy, especially from the much less rare leiomyosarcoma. Orthopedic oncologists have to include this rare benign tumor in the differential diagnosis of any intraosseous lesion with gradually worsening and long-standing pain, despite of benign imaging characters. Different histological patterns of leiomyoma do exist, however there is no difference in prognosis or treatment options. Treatment standard includes wide excision with autologous bone graft whenever possible. Internal fixation may be necessary if the bone defect is large or there is thinning out of the cortex that may lead to pathological fracture. Keywords: Intraosseus, Benign bone tumors, leiomyoma.

Introduction

Leiomyoma is a benign smooth-muscle tumor that most commonly arises in the uterus, the gastrointestinal tract, and the skin. It constitutes 70% to 80% of all benign mesenchymal tumors [1], with uterine leiomyomata being the most common smooth-muscle tumor in women [1]. Leiomyomata are infrequently seen in the extremities and rarely seen in bone [2]. To the best of our knowledge, the eleven reported cases occurred in peripheral skeleton. Five cases were intraosseous lesion in long bones, including neck of the femur [3], ulna [4], fibula [5], tibia [6] and distal femur [7]. Three cases were periosteal leiomyomata [8] and two cases arose from the hip bone [6]. In addition to three cases of intraosseous leiomyomatosis secondary to disseminated leiomyomatosis have been reported [9]. We describe a case of a patient with painful intraosseous leiomyoma involving the proximal aspect of the tibia and review the literature about intraosseous leiomyoma.

What to Learn from this Article?
Leiomyoma could be a possible differential diagnosis of any intraosseous lesion with a gradually worsening and long-standing pain despite of benign imaging characteristics and orthopaedic oncologists need to keep this in mind in order not to miss it. Though different histological patterns of leiomyoma exist, there is no difference in the prognosis or treatment options which include wide excision with autologous or artificial bone grafting.

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Case Presentation

A 73-year-old female presented with a four year history of a painful swelling in the left knee that was slowly growing, with a gradual increase in pain at the same time. She had an unnoticeable medical history. Pain was aggravated by weight bearing, sitting straight, but she enjoyed full range of motion, and her general health was not affected. Osteoarthritis of left knee joint was suspected in another clinic, and she was treated conservatively with analgesic drugs. The pain persisted despite administration of nonsteroidal anti-inflammatory drugs (NSAIDs) for a long time; she visited our outpatient clinic for consultation. Clinical examination revealed local tenderness over the medial aspect of the proximal tibia, a small well-defined swelling over the medial proximal tibia was palpable. It was firm in consistency and was not red or hot to touch. Roentgenography revealed a small well-defined osteolytic lesion with thinning out of the cortex over the proximal part of the tibia, surrounded by a thin sclerotic margin (Fig. 1A, 1B). Computed tomography (CT) scan revealed a radiolucent lesion eroding the cortex, with central calcification (Fig. 1C, 1D). Magnetic resonance imaging (MRI) revealed a hypechoic mass in both T1 and T2 weighted images, not suppressed on T2 fat suppression view T2FS, no fluid-fluid level, it was enhanced with Gadolinium, the lesion had extra osseous extension (Fig. 2A-2D). Technetium99 (Tc99) bone scan revealed an increased uptake. Laboratory studies were within normal range. CT-guided biopsy was performed, and the pathology showed a picture of leiomyoma, consisting of moderate cellular proliferation of smooth-muscle cells with little cellular pleomorphism, with no mitoses or necrosis. Immunohistochemical stains showed positive stain for smooth-muscle actin (SMA). The diagnosis of a benign intraosseous leiomyoma was made. Since the definite diagnosis was established, surgical excision with wide margin was decided to avoid local recurrence. The specimen consisted of gray-white soft tissue mass without apparent central necrosis or fluid content by macroscopic examination. Microscopically, the tumor composed of fascicles of spindle-shaped cells with elongated nuclei and fusiform or blunt ends. No atypical mitosis was seen. Immunohistochemical stains on SMA were diffusely positive. The surgical margin was free of tumor. Otherwise, the histological exams were compatible with the results of biopsy and confirmed the diagnosis of intraosseous leiomyoma. The patient symptoms had improved after surgery, pain gradually improved postoperatively until it disappeared by two months after surgery, weight bearing was allowed from next postoperative day, convalescence passed uneventfully. At the latest follow-up after 18 months following surgery, the range of motion of left knee achieved full extension and flexion to 130 degrees without discomfort without any instability or residual pain of the knee. No local recurrence, malignant change, or distal metastasis had occurred.

Discussion

Leiomyomata are very rare in the bone. Most of reported intraosseous leiomyomata arise from axial skeleton. First case of intraosseous leiomyoma was in the maxillary tooth socket and it was described in 1976 by Rhatigan and Kim [10]. Loyola et al. revised 11 cases involving the mandible and maxilla [11]. The lesion is recorded in other areas of the axial skeleton such as skull, spine, as well as a single case of the rib has been reported [12], while only limited cases of intraosseous leiomyoma in peripheral skeleton have been published. In published English literature, eleven cases were found with peripheral skeleton...
defined, irregular osteolytic lesion with a moth-eaten or permeative pattern of osseous destruction radiographically, and diffuse involvement and destruction of the medullary bone histologically. On the other hand, low grade tumors exhibit diffuse involvement of the marrow spaces, and have radiographic features, including a geographic pattern of bone destruction and a sclerotic rim ([14]. Calcification is exceedingly rare in leiomyosarcoma. In MRI, it may frequently demonstrate cystic foci within. The lesion appears so intense to muscle T1, while inT2 intermediate to hypo intense to neighboring fat, and predominantly hyper intense in T2FS. Metastasis is not a far possibility, it would occur from primary carcinomas, including lung, breast, kidney, and pancreas. The thin capsule seen around the lesion, as well as its oval shape and sharply marginated edge made metastasis less likely. We considered adamantinoma far less likely in the differential diagnosis, as histologically this neoplasm is noted for its heterogeneity and presence of epithelial islands. A malignant spindle cell tumor was not considered because the tumor lacked pleomorphism, high rate of mitoses, and necrosis. The sclerotic margins seen on the conventional radiograph and in the histologic sections also supported the benign nature of this tumor. However, absence of signs of malignancy must be carefully documented in order to rule out leiomyosarcoma.

Histologically, leiomyoma is classified into three histological categories: solid leiomyoma, vascular leiomyoma (or angioleiomyoma), and epithelioid leiomyoma. While angioleiomyoma is the most prevalent type in soft tissues,
solid leiomyoma represents the majority of cases of intraosseous leiomyomata [15], it exhibits the same histological features as leiomyoma at other sites. It is composed of a proliferation of spindle-shaped cells arranged in thick intersecting bundles. Mitoses are very rarely seen (low rate of mitosis 0-4 per ten high power fields) with moderate cellularity and no necrosis. Angioleiomyomas are further classified into three histological categories: solid (the most common type in soft tissues), venous, and cavernous [15]. However, all three patterns are often present within the same tumor. Similarly, reported cases of intraosseous angioleiomyoma showed no distinctive histological features compared to soft-tissue angioleiomyoma [15]. In published cases, three cases out of eleven were histologically angioleiomyoma, two were in tibia and one in the iliac bone.

The use of immunohistochemical staining was helpful in this case, the tumor cells stained strongly for muscle markers, including smooth muscle actin and desmin, thus confirming this tumor's smooth muscle origin. The neoplastic cells did not stain for neural, fibrous, or epithelial markers (S-100, CD34, and AE1/3, respectively).

The standard treatment for intraosseous leiomyoma is extensive tumor excision with wide margin followed by packing of the cavity with autologous cancellous bone graft [4]. Internal fixation is required when the residual bone defect is large. Complete excision of intraosseous leiomyoma is typically curative with excellent prognosis. Till now, no cases of recurrences have been reported after complete excision [4,5], however, regular follow-up is necessary to observe the risk of recurrence.

In only one of the previously reported cases, reconstruction using bone graft alone was not enough, and reconstruction by TKA was performed due to difficulty in preserving the articular cartilage and thinning out of the bone [8]. Otherwise, all known cases were treated either by en-bloc excision, surgical resection or curettage. In addition to bone graft, and in all cases, results were satisfactory to excellent with no recurrence, malignant change, or distant metastases, early recovery and restoration of function.

In our case, we performed wide tumor excision, the bone defect was not large enough to necessitate internal fixation. Patient satisfaction after surgery was good, with improvement of pain, and preserved function of the knee. Pain was completely relieved by two months post-operatively. There was no recurrence, malignant change or distant metastases at the latest follow up visits.

**Conclusion**

Diagnosis of Intraosseous leiomyoma of the extremities is difficult due to extreme rarity of the tumor and absence of pathognomonic radiological signs in X-ray, CAT, or even MRI. The exact diagnosis is only achieved by histopathological examination and with immunohistochemistry stains, which can differentiate it from malignancy, especially from the much less rare leiomyosarcoma. Orthopedic oncologists have to include this rare benign tumor in the differential diagnosis of painful benign intraosseous lesions.

**Clinical Message**

Intraosseous leiomyoma is a rare tumor; it lacks a pathognomonic radiological sign which makes its diagnosis quite difficult. It is better to include this tumor in the differential diagnosis of painful benign intraosseous lesions.

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### Table 1

| No | Author | Year | Country | Age | Sex | Bone        | Location       | Histological type              | Treatment                        |
|----|--------|------|---------|-----|-----|-------------|-----------------|-------------------------------|----------------------------------|
| 1  | Taxy at al. | 1981 | USA     | 37  | ♂   | Tibia       | Periosteal       | Solid leiomyoma               | Surgical excision                |
| 2  | Mirra JM | 1989 | USA     | 39  | ♂   | Tibia       | Periosteal       | Solid leiomyoma               | Surgical excision                |
| 3  | Tomoda and Iyama | 1992 | Japan | 64  | ♂   | Tibia       | Proximal          | Angioleiomyoma                | Marginal excision and autologous bone graft |
| 4  | Braun W et al. | 1994 | Germany | 54  | ♂   | Femur       | Proximal(neck)    | Solid leiomyoma               | Curettage and autologous bone graft |
| 5  | Vanillo-Vinagre et al. | 1999 | Spain  | 28  | ♂   | Iliac bone  | Iliac crest       | Angioleiomyoma                | Curettage, autologous cancellous bone graft |
| 6  | Zikria BA et al. | 2004 | USA     | 38  | ♂   | Ulna        | Distal end       | Solid leiomyoma               | En bloc excision                 |
| 7  | Lafosse J-M, Gomez-Brouchet A | 2007 | France  | 42  | ♂   | Fibula      | Distal end       | Solid leiomyoma               | Curettage, autologous bone graft & internal fixation by plate |
| 8  | Lafosse J-M, Gomez-Brouchet A | 2007 | France  | 46  | ♂   | Pubic bone  | Superior pubic ramus | Solid leiomyoma               | Surgical excision and autologous bone graft |
| 9  | Seena C. Aisner et al. | 2008 | USA     | 37  | ♂   | Tibia       | Diaphyseal(periosteal) | Solid leiomyoma               | En bloc tumor resection          |
| 10 | Yu-Hsiang et al. | 2013 | Taiwan  | 57  | ♂   | Femur       | Distal end       | Solid leiomyoma               | En bloc excision & TKA          |
| 11 | Sean K. Lau | 2014 | USA     | 48  | ♂   | Tibia       | Distal end       | Angioleiomyoma                | Curettage and allograft          |
| 12 | Our case | 2015 | Japan   | 73  | ♂   | Tibia       | Proximal end      | Solid leiomyoma               | Excision with wide margin        |
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