Case Report

Including MRI of a Primary Bone Leiomyosarcoma that Radiologically Mimics a Giant Cell Tumor

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The authors present a case of a 42-year-old female who developed a leiomyosarcoma of the right proximal tibia that appeared radiologically similar to a giant cell tumor. Histology revealed spindle cells running in whorl-like fashion with focal atypia and low mitotic figures. The immuno-stains revealed positive reactivity for alpha-smooth muscle (SMA), muscle actin and cytokeratin (AE1/AE3). The authors rendered a diagnosis of low-grade leiomyosarcoma of bone. The lesion was considered a primary lesion since the patient did not have other leiomyomatous tumors. The MRI showed hypo- to iso- signal intensity on T1-weighted imaging and heterogeneous intensity on T2-weighted imaging. This was likely due to admixed fibrotic tissue in the lesion. The tumor cells were not positive for Ebstein-Barr virus by in-situ hybridization as seen in leiomyomatous tumors in immunodeficiency patients.

Keywords: *Leiomyosarcoma, Bone, MRI, Imaging, Immunostains*

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Giant cell tumors of bones often present specific radiographic features. However, they may also radiologically mimic other lesions ranging from aneurysmal bone cysts[1], telangiectatic osteosarcomas[2] and chondroblastomas[3,4]. Even though there are approximately 120 reported cases of primary leiomyosarcoma of bone, only about 20 of these include imaging studies and the issue of lesions that mimic giant cell tumors was not addressed.

Case Report

A 42-year-old female suffered pain in the right knee for two years. Treatment for tendinitis did not improve her symptoms and the pain became progressively worse until she could no longer walk. Physical examination revealed swelling and mild tenderness of the right knee and flexion contracture of 30 degrees. The plain film of the right knee showed an expansile osteolytic lesion with internal "pseudotrabeculae" at the distal femur. The preliminary diagnosis was a giant cell tumor (Fig. 1). Magnetic resonance imaging (MRI) of the lesion revealed hypointense signals relative to muscle in T1 and inhomogeneous intensity relative to fat in T2 (Fig. 2-3). Following curettage of the lesion, it was found that the tissue comprised approximately an equal number of cellular spindle cells and dense fibrous tissue cells. The spindle cells had elongated nuclei with blunt edges and slender eosinophilic cytoplasmic extensions that formed interlacing bundles and intersected with each other at wide angles. The dense fibrous tissue comprised of sparse fibroblasts intermingled with dense collagenous cells (Fig. 4-5). Immunohistochemical staining revealed positive reactivity for AE1/AE3 and SMA.

Discussion

Primary osseous leiomyosarcoma is very rare. To the authors' knowledge, there are only 120 cases in the English literature. Among these, the most common patient age was the fourth decade and the sites of predilection were around the knee. Conventional radiographs show what appear to be purely osteolytic

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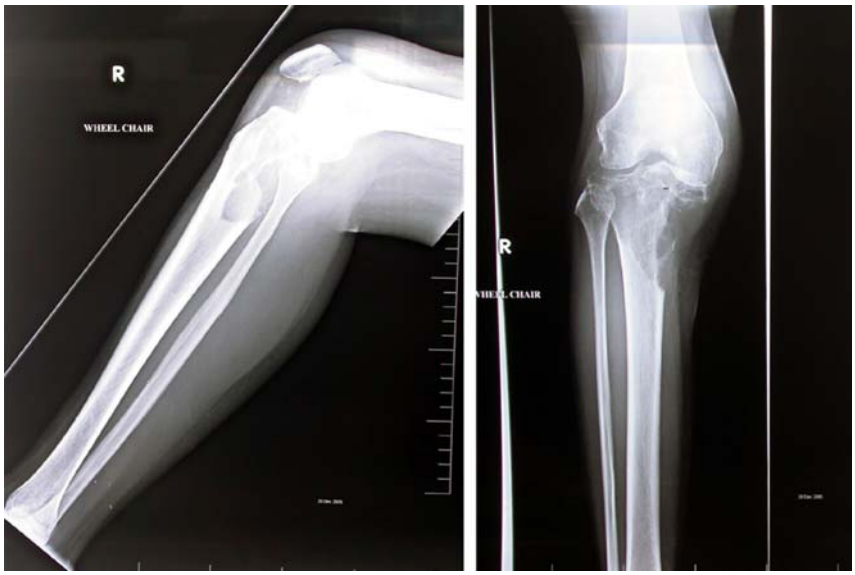

Fig. 1 Plain films of the right knee in lateral (left hand side) and AP views (right side) showing an osteolytic expansile lesion involving the metaphysis and epiphysis with internal pseudotrabeculae

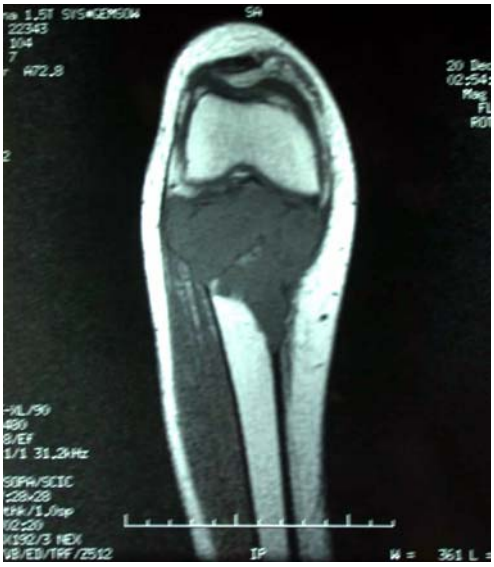

Fig. 2 MRI showing hypointense signal of the lesion on T1W image

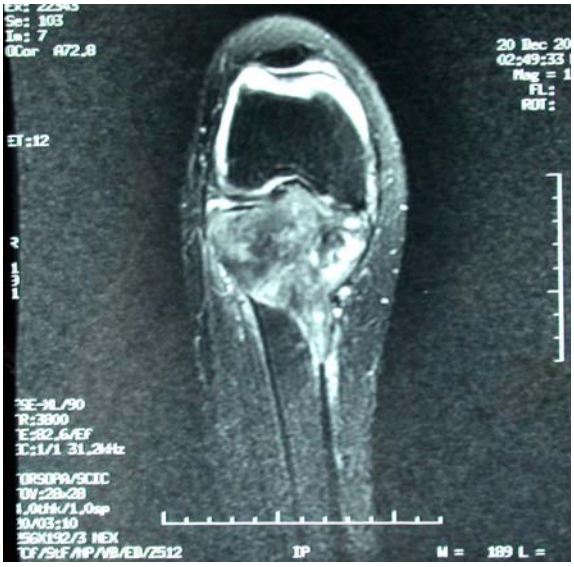

Fig 3. MRI showing an inhomogeneous signal intensity of the lesion on T2W image

lesions with at least one of the following characteristics: indistinct margins; moth-eaten or permeative patterns; cortical breakthroughs and lack sclerotic rims[5]. However, conventional radiographs do not give specific features that allow differentiation from other aggressive osteolytic lesions. The present lesion showed similar features to osteolytic lesions in that it possessed an ill-defined margin and showed cortical breakthrough. Thus, in the plain film, it had an appearance quite similar to that of a giant cell tumor and was initially diagnosed erroneously as such. It is important for radiologists and clinicians to be aware of this pitfall and understand

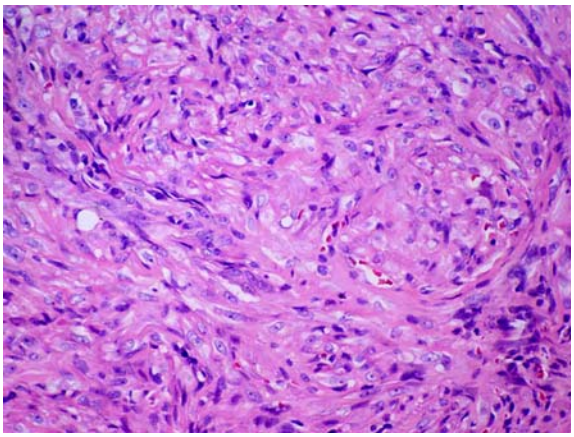

Fig. 4 (H&E; x200) Photomicrograph of the lesion showing spindle cells running in interlacing bundles
The cells have elongated pale stained nuclei with prominent nucleoli and blunt edges
The cytoplasm is eosinophilic and contains occasional vacuoles

that tissue diagnosis is necessary to distinguish amongst lesion types, since the treatment and prognosis for each is different. It was well documented that less than 10% of giant cell tumors are able to metastasize. By contrast, primary osseous leiomyosarcomas had metastatic rates of 30 to 70% and approximately 50% of patients developed lung metastasis within 5 years[6-9]. In addition, the survival time for leiomyosarcomas was shorter, sometimes as short as 1 to 2 years [10,11]. The conventional treatment for giant cell tumors is curettage while leiomyosarcomas, similar to other sarcomas, often require resection with or without adjuvant chemotherapy[6-8]. The MRIs of previously reported cases are inconclusive. Ganau et al[12] described isointense with focal hyperintense results with T1-weighted imaging and hyperintense results with T2-weighted imaging. Sundaram et al[13] found isointense skeletal muscle with T1-weighted images and isointense results relative to fat with T2-weighted images. Shu-Huei Shen et al[14] described isointense

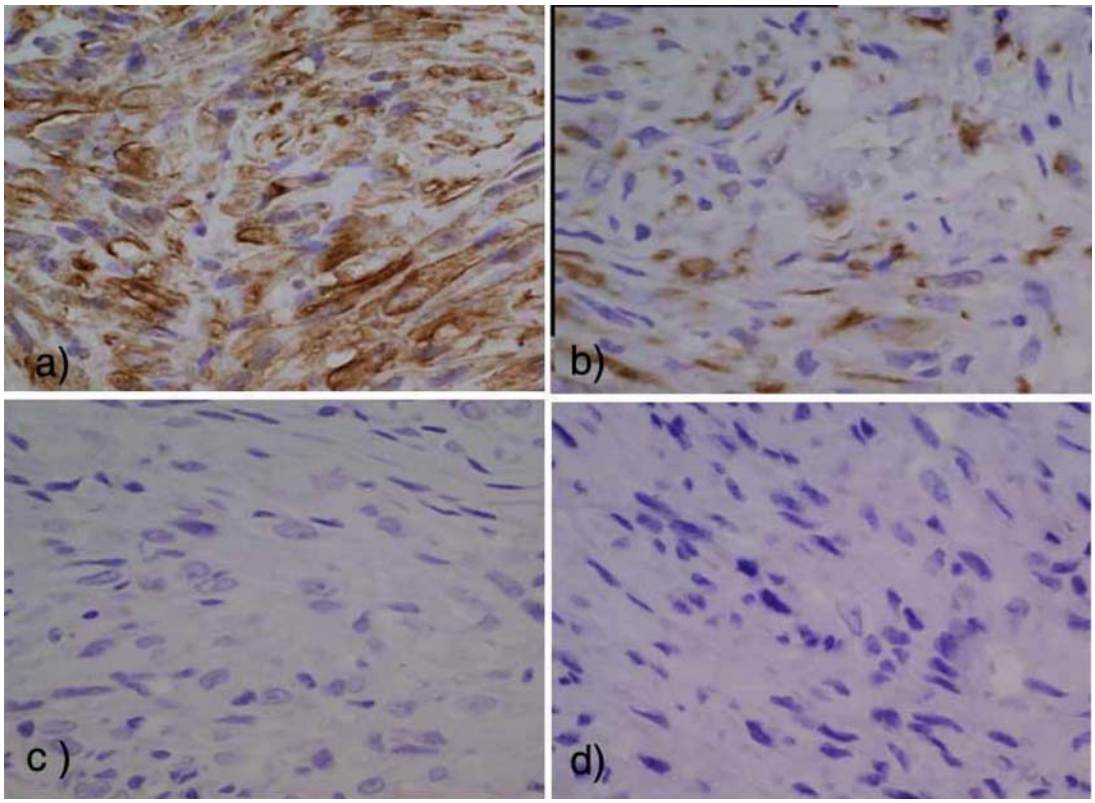

Fig. 5 Photomicrographs of immunohistochemically stained tumor cells
a) Staining for alpha smooth muscle actin (SMA) showing an intense cytoplasmic reaction; b) Staining for cytokeratin (AE1/AE3) showing a strong cytoplasmic reaction; c) Staining for desmin showing a negative reaction; d) Staining for osteocalcin showing negative cytoplasmic reaction

results relative to muscle with T1 and iso-hypointense results relative to fat with fast, T2-weighted imaging. Lo et al[15] reported intermediate signal-intensity with T1-weighted images and high signal-intensity with T2-weighted images. Most of the reported cases showed iso-signal intensity relative to muscle with T1-weighted imaging, and that was similar to what the authors found in the presented case. However, there was a discrepancy in T2-weighted images. The presented case exhibited inhomogeneous intensity, and this was rarely mentioned in previous reports. The authors propose that the presented lesion contained a large proportional area of fibrosis and that this contributed to the inhomogeneous intensity with T2-weighted imaging. Pilavaki et al[16] described similar findings with T2-weighted imaging of a soft tissue leiomyosarcoma and they proposed that the foci of decreased signals were due to hyalinization and deposition of hemosiderin or metaplastic bone. In the presented case, immunohistochemical results were positive for most of the essential markers associated with leiomatous tumors including muscle actin HHF and SMA. The lack of desmin expression in the presented case might suggest that the tumor cells did not originate from vascular smooth muscle cells but probably from multipotential mesenchymal cells[5]. During this decade, there have been reports regarding an association between Epstein-Barr viral infection and leiomyomatous tumors in patients with AIDS, especially from countries with a high prevalence of HIV infection[17-19]. Suankratay et al from Thailand reported nine patients with unusual manifestations of smooth muscle tumors in AIDS patients during 2001-2003. The sites of involvement included epidura, vocal cords, adrenal glands, iris, liver, lungs, orbit and thigh[19]. Although there have been no reports of osseous leiomyomatous tumors associated with Epstein-Barr viral infection in literature, the authors examined the possibility by performing a serum test for HIV infection and an in-situ hybridization test for EBV infection with the tumor tissue. Both tests gave negative results. The authors, therefore, concluded that the leiomyosarcoma in the presented case was unlikely to be associated with EBV infection. However, the authors cannot completely negate the possibility and recommend that any future case include testing for EBV infection, especially if the patient has AIDS.

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เนื้องอก leiomyosarcoma ของกระดูกซึ่งมีลักษณะทางเอกซเรย์คล้ายเนื้องอกชนิด giant cell tumor: รายงานผู้ป่วย 1 ราย

วรชัย ศิริกุลชยานนท์, สุภนีวรรณ เชาว์วิศิษฐ

รายงานผู้ป่วย 1 รายซึ่งมีเนื้องอกชนิด leiomyosarcoma ของกระดูกtibia ซึ่งมีลักษณะทางเอกซเรย์คล้าย giant cell tumor ลักษณะทางจุลพยาธิวิทยาของเนื้องอกประกอบด้วยเซลล์ คล้ายรูปกระสวยเรียงตัวเป็นมัด และวนคล้ายก้นหอย เซลล์บางตัวมี atypia และ mitosis การย้อมทางอิมมิวโนพยาธิวิทยาพบว่า เซลล์ของเนื้องอกให้ผลบวกกับ alpha smooth muscle actin (SMA), cytokeratin (AE1/AE3) และให้ผลลบต่อ desmin และ osteocalcin ลักษณะของภาพที่ตรวจด้วยคลื่นแม่เหล็กไฟฟ้าพบว่าเป็นแบบ hypo และ iso signal intensity ของ T1-weighted image และ heterogeneous intensity ของ T2-weighted image ซึ่งอาจเกิดจากการที่พยาธิสภาพมีการเกิด fibrosis ร่วมด้วย และจากการตรวจในห้องปฏิบัติการด้วยวิธี in-situ hybridization พบว่าเซลล์เนื้องอกไม่ให้ผลบวกต่อการมีเชื้อไวรัสชนิด Epstein-Barr ซึ่งให้ผลแตกต่างจากเนื้องอกของกล้ามเนื้อเรียบที่เกิดขึ้นที่อวัยวะอื่น ๆ ที่พบในโรคภูมิคุ้มกันบกพร่อง