Orthopaedic Case of the Month

Painless Right Knee Mass in 32-year-old Man

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History and Physical Examination

A 32-year-old healthy male presented to the hand clinic with a nondisplaced fracture of the right fifth metacarpal neck. During the visit, he mentioned a slowly enlarging painless mass anterior to the right knee and was referred to the orthopaedic oncology service. The mass had been present for approximately 10 years and increased in size for several months before the patient's presentation. He denied constitutional symptoms and neurologic deficits. He was a goalkeeper for an ice hockey team and therefore, the site of the mass was subject to frequent friction and irritation. However, the patient denied specific trauma to the right knee and had no mechanical symptoms of locking or

instability. He had no other masses and had not sought diagnosis and treatment for the knee mass.

Examination of the right knee showed an approximately 4-cm × 2-cm mobile, nontender, well-defined mass located along the medial border of the patella. The overlying skin was normal and the mass was nonfluctuant, noncompressible, did not transilluminate, and did not change in size with knee ROM or quadriceps contraction when the knee was extended against resistance. The right knee had full ROM, no effusion, and otherwise, normal findings regarding stability, alignment, and patellar tracking. There was no popliteal or inguinal lymphadenopathy and the distal neurovascular examination was unremarkable. The laboratory workup was noncontributory. Plain radiographs including AP, lateral, and skyline views (Fig. 1) and MRI with contrast were obtained (Fig. 2).

Based on the history, physical examination, laboratory studies, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

AP, lateral, and skyline plain radiographs of the right knee showed a soft tissue shadow medial to the patella with no calcification (Fig. 1). MRI of the knee showed a superficial, ovoid, well-circumscribed, extracapsular soft tissue mass abutting the medial retinaculum. The mass measured 3.0 cm × 1.7 cm × 2.6 cm. The mass was homogeneously hypointense to muscle on T1-weighted imaging and had a heterogeneous high signal on T2-weighted imaging. Post-contrast images showed enhancement of the mass, which had septations and slightly lobular borders. The surrounding musculature, ligaments, and bones were uninvolved. There was no joint effusion or surrounding edema (Fig. 2).

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Each author certifies that his or her institution approved or waived approval for the reporting of this case and that all investigations were conducted in conformity with ethical principles of research.

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Fig. 1A–C (A) AP, (B) lateral, and (C) skyline plain radiographic views of the right knee show a noncalcified soft tissue mass medial to the patella. The mass is best seen on the AP view (arrow).

Differential Diagnosis

Soft tissue sarcoma (STS)

Fibromatosis

Solitary nodular form of pigmented villonodular synovitis (PVNS), also known as tenosynovial giant-cell tumor

Chronic granulomatous infection

Inclusion cyst

Chronic bursitis

Hemangioma

Lipoma

An ultrasound-guided core needle biopsy of the mass was performed. The specimens were stained with hematoxylin and eosin and immunohistochemical staining also was performed (Fig. 3).

Based on the history, physical examination, laboratory studies, imaging studies, and histologic picture, what is the diagnosis and how should the patient be treated?

Histology Interpretation

Microscopic examination of the biopsy specimen showed bundles of elongated cells with eosinophilic cytoplasm and oblong nuclei with blunt ends. There was benign focal vascular proliferation without evidence of increased mitotic activity, cellular atypia, hemorrhage, or necrosis. There were no thick-walled vessels or calcification. The predominant cell type was smooth muscle confirmed by positive immunohistochemical staining with desmin and smooth muscle actin (SMA).

Fig. 2A–D (A) A preoperative T2-weighted, fat-suppressed, precontrast sagittal image, shows a well-circumscribed, extracapsular, heterogeneously enhancing mass. (B) The T1-weighted, fat-suppressed axial image, and the (C) T1-weighted, fat-suppressed axial image with contrast, show enhancement with some septations and slightly lobular borders. (D) Proton density sagittal imaging reveals a well-circumscribed, extracapsular mass that is hypointense to skeletal muscle and slightly heterogeneous.

Diagnosis

Leiomyoma of deep soft tissue

Discussion and Treatment

Leiomyoma was not considered in the differential diagnosis owing to its rarity. It was diagnosed only on histologic examination and immunohistochemical stains. In STSs, 46% of lesions occur in the lower extremities as a slow-growing mass [23] with similar MRI characteristics to leiomyoma.

The patient's age and a mass for several years may suggest synovial sarcoma, which may be monophasic showing spindle cells or biphasic that stains positive for epithelial markers [14]. STSs show mitoses and necrosis not identified in this lesion. Fibromatosis is a benign, locally aggressive soft tissue tumor [15] and 1/3 of extra-abdominal tumors involve the lower extremities [22]. They are more heterogeneous on MRI. Although fibromatosis can stain positive with actin and desmin [12], it consists mainly of fibroblasts and collagen [22]. Nodular PVNS of the knee occurs at an average age of 33 years [1, 10, 16]

and would be hypointense on MRI with giant cells and hemosiderin-containing macrophages on microscopy [1, 10]. The patient had no clinical, laboratory, or histologic findings suggesting chronic granulomatous infection [19]. Kneeling in internal rotation as a hockey goalkeeper could result in an unusual location for bursitis. However, the lesion was not painful, and not cystic. Histologic examination was not suggestive of an inclusion cyst or bursitis. A synovial hemangioma may affect the knee in younger patients [1, 18]. MRI shows a lobulated serpentine appearance and histologic examination shows greater vascularity [18]. Lipomas represent 1/2 of soft tissue tumors in older patients. Synovial lipoma in a joint or tendon sheath is uncommon and could be diffuse, known as lipoma arborescens. The reported lesion however, was extraarticular. In addition, lipomas appear as fat on MRI and mature adipocytes on histologic examination [13, 15].

Leiomyomas outside the reproductive tract occur in 5% [4, 5, 8, 15] in the fourth to sixth decades of life equally in males and females [2, 4, 5]. Extraabdominal leiomyomas are rare and mostly smaller than 2 cm in diameter [2, 4, 5, 7, 15]. Leiomyomas rarely show mitotic figures and atypia [21], are positive for SMA, and rarely negative for desmin [2, 3, 6, 7, 9, 17, 25, 26]. Leiomyoma cutis associated with the erector

Fig. 3A–D (A) The permanent histologic section shows bundles of elongated cells with eosinophilic cytoplasm and oblong nuclei and the thick fibrous capsule (bottom right) (Stain, hematoxylin and eosin; original magnification, $\times 4$). (B) A higher magnification view shows bundles of smooth muscle cells (Stain, hematoxylin and eosin;

magnification, $\times 40$). (C) Desmin immunohistochemical staining shows intense staining of neoplastic cells (Original magnification, $\times 40$). (D) SMA immunohistochemical staining shows cells to be positive for SMA (Original magnification, $\times 40$).

pilli muscles around the genitals or extensor surfaces of the extremities is the most common and smallest [9]. Vascular leiomyomas are more common in the extremities of females, and 58% present with pain or tenderness [5]. Deep soft tissue leiomyomas are the largest, most uncommon, and most concerning. Retroperitoneal lesions could be confused with leiomyosarcomas [17, 26]. In the extremities, men and women are affected equally [2, 11, 17]. Deep soft tissue leiomyomas have a propensity to calcify [3, 6, 7, 24, 26] and are the only subtype reported in an intraarticular location [20]. The natural history of a leiomyoma is indolent growth without metastasis or transformation to leiomyosarcoma. Treatment consists of marginal excision, and the local recurrence rate after excision of a leiomyoma ranges from less than 1% to 9% [2, 5, 7, 9, 17, 27]. One study showed only one recurrence in 11 tumors at 4.5 years of followup [7], and another series had one recurrence in 36 cases at 4 years of followup [2].

In the reported lesion, leiomyoma cutis was excluded as the lesion was relatively large, painless, and not associated with a hair follicle. Vascular leiomyoma was unlikely

owing to the lack of thick-walled vessels on microscopy. Therefore, the mass may represent a deep leiomyoma. The scant subcutaneous tissue medial to the patella might have led to the unusual superficial appearance.

Treatment was approached with caution owing to tumor rarity and unusual presentation. Although marginal resection of a leiomyoma is adequate, STSs and fibromatosis require wider margins to enhance local control. Based on lack of features of malignancy, it was decided to perform a negative, yet close, marginal resection. Violation of the knee capsule and retinaculum was avoided to preserve the joint in the event that the final diagnosis required tumor bed excision. Microscopic examination confirmed leiomyoma with negative resection margins [2, 3, 6, 7, 9, 17, 25, 26].

We report a rare extraarticular, soft tissue mass of the knee. We describe the caveats and the desired caution in the diagnosis and treatment of unusual soft tissue masses particularly when the initial diagnosis is rare and the more common diagnoses are aggressive or malignant and therefore, may require wider resection margins. Emphasis should be placed on avoiding unnecessary contamination

of nearby tissues in anticipation of tumor bed excision. Our patient had no recurrence of the mass at 8 months followup.

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