Intra-Articular Neurofibroma: An Unusual Source of Anterior Knee Pain

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Abstract

The infrapatellar fat pad of the knee is often implicated as a source of anterior knee pain. We present an unusual case of anterior knee pain in an adult patient from a solitary neurofibroma arising in an intra-articular location associated with the infrapatellar fat pad. Clinical, radiographic, magnetic resonance imaging (MRI), and histopathologic features are presented, as well as, a review of the literature. This case adds to the differential diagnosis of fat pad lesions seen on MRI and contributes to understanding the spectrum of clinical presentations of neurofibromas and pathology involving this region.

Diagnosing the source of anterior knee pain can at times be difficult. A variety of structures, alone or in combination, can be the source of discomfort. The infrapatellar, or Hoffa's, fat pad is often implicated as a source and can itself be affected by a spectrum of disorders. This is most likely due to its unique anatomy and location within the knee joint. The fat pad is an intracapsular and extrasynovial structure. It is subject to not only injury and inflammation, but to benign and malignant neoplasms.[1-3]

We present an unusual source of anterior knee pain in an adult patient from a solitary neurofibroma arising in an intra-articular location associated with the infrapatellar fat pad of the knee. Clinical, radiographic, magnetic resonance imaging (MRI), and histopathological features are presented.

The patient provided written informed consent for print and electronic publication of this case report.

Case Report

A 47-year-old African American woman presented with a 10-year history of slowly progressive right anterior knee discomfort. She first sought medical treatment 2 years prior to our initial consultation and was evaluated by conventional radiographs and MRI scan of her knee. The initial radiographs were interpreted as normal. She was informed that MRI findings revealed 2 cysts in her knee, underwent cortisone injection treatment and an aspiration attempt. The patient received no benefit. She then sought a second orthopedic opinion and received a second cortisone injection with no relief.

At the time of our first evaluation, the patient reported persistent swelling in her right knee for more than 2 years and a remote history of trauma to the right knee, but no injury within the last 20 years. The patient had no instability symptoms and no symptoms of locking or catching. Her level of knee pain on a visual analog pain scale was at times a 9 out of 10.

Physical examination revealed a morbidly obese woman with a body mass index (BMI) of 46 who stood with neutral knee alignment. Examination of the right knee confirmed an obvious mass in the infrapatellar region, with prominence both medially and laterally. The mass was soft, but not cystic. Moderate tenderness was noted diffusely over this area, as well as directly over the patellar tendon itself. Mild patellofemoral compression discomfort was noted. The knee showed no effusion, no ligament instability, and a range of motion from full extension to 110° of flexion, a 10° restriction of flexion compared to the left knee. McMurray's testing was negative. There was no neurologic or vascular

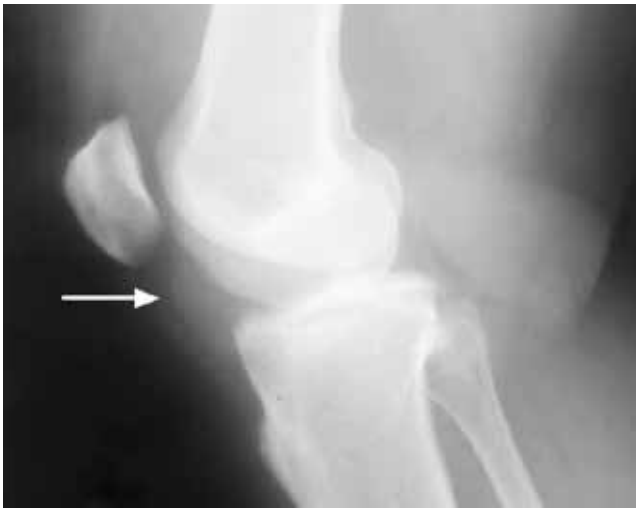

Figure 1. Lateral radiograph of the right knee demonstrates infrapatellar fat pad density (arrow).

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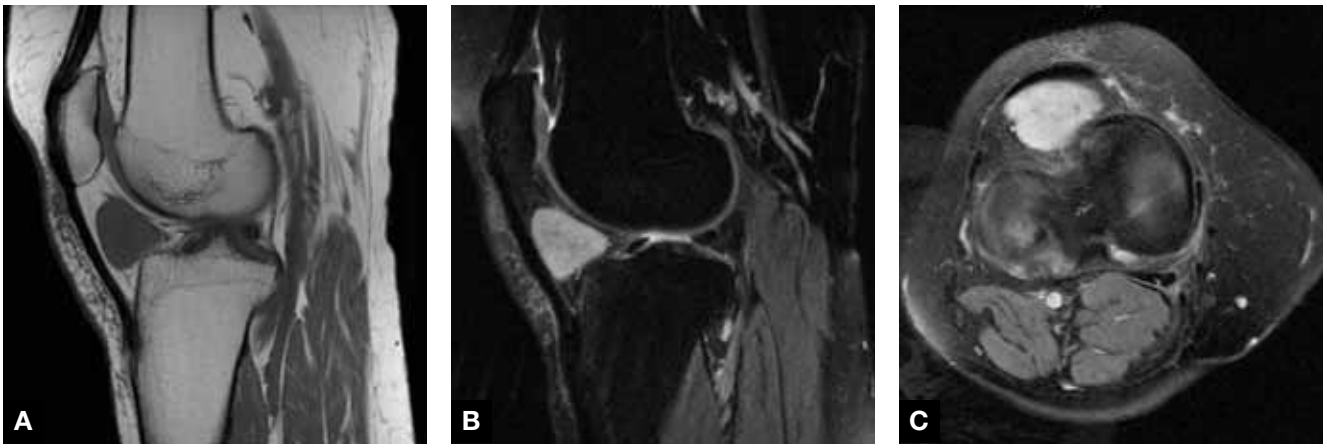

Figure 2. MRI of the right knee shows a well-marginated fat pad lesion of (A) intermediate signal intensity on sagittal T1-weighted sequence, (B) bright signal intensity on sagittal T2-weighted fast spin echo fat suppressed sequence, and (C) subtle heterogeneous signal intensity on axial T2-weighted fast spin echo fat suppressed proton density sequence.

deficit in the right lower extremity. The patient exhibited no skin nodules or abnormal cutaneous findings.

Updated conventional radiographs of the right knee were obtained. The lateral radiograph of the right knee demonstrated a lobular soft tissue density within the infrapatellar fat pad (Figure 1). The MRI study of the knee was reviewed. The sagittal images demonstrated a well-marginated lesion within the infrapatellar fat pad corresponding to the radiographic abnormality, intermediate in signal intensity on the T1-weighted sequence (Figure 2A) and bright in signal intensity on the fast spin echo fat suppressed T2-weighted sequence (Figure 2B). The axial (Figure 2C) and coronal fast spin echo fat suppressed proton density images showed subtle heterogeneous signal intensity within the lesion. Based on the MRI signal characteristics, the principal diagnosis was a complex ganglion cyst. The differential diagnosis included a neurogenic type tumor such as a neuroma or schwannoma, and less likely, a chondroid matrix lesion such as a chondroma. A specific diagnosis could not be made at that time.

A recommendation was made for surgery with a preoperative diagnosis of a large intra-articular ganglion cyst arising from the infrapatellar fat pad. The literature suggested treatment by arthrotomy for cysts of this size.[4] Arthroscopy followed by a planned arthrotomy of the right knee was performed.

Right knee arthroscopy revealed a relative obstruction attempting to pass the arthroscope from a lateral portal to the medial compartment. The infrapatellar fat pad was never clearly visualized and no specific intercondylar lesion identified. At the time of medial arthrotomy, a small portion of the medial fat pad was removed exposing a mass lodged in a posterior fat pad location. Digital manipulation of the mass mobilized it easily to the medial arthrotomy opening (Figure 3A). It was completely mobile with the exception of a small pedicle tethering it to the most posterior aspect of the fat pad like an umbilical cord. The pedicle was incised at the base of the fat pad and the entire tumor easily removed (Figure 3B). Further inspection of the bed showed no other abnormal findings. A ligamentum mucosum was notably absent. The specimen was incised (Figure 3C) and sent to pathology.

The gross pathological examination of the submitted tissue revealed a previously incised, 4.5 x 2.6 x 2.2 cm, well-circumscribed, rubbery, solid, gray-tan, polypoid mass with a gray-tan solid cut surface (Figure 3C) and an apparent site of a vascular pedicle (Figure 3B). On microscopic examination, the hematoxylin/eosin stained section revealed a circumscribed cellular neoplasm composed of wavy spindle cells in a fibrous stroma with a diffuse growth pattern, including occasional mast cells (Figure 4A) and scattered so-called pseudo-Meissner corpuscles (Figure 4B). The histological features included the site of a vascular pedicle and an attenuated peripheral nerve fiber. An immunostain for S-100 protein was diffusely and strongly expressed in the neoplastic cells (Figure 4C). The pathologic features were of a neurofibroma.

The patient's right knee pain nearly completely resolved with a return of full knee range of motion at final follow-up, 4 months postoperatively.

Discussion

Neurofibromas are not uncommon, accounting for about 5% of all benign soft tissue tumors.[5] They are considered to be nerve sheath tumors arising from nonmyelinating Schwann cells and eventually assume 1 of 3 growth patterns: localized, diffuse, or plexiform.[6] The localized form is usually seen as a solitary tumor in normal individuals. Diffuse and plexiform neurofibromas have a close association with the genetic disorder neurofibromatosis type 1 (NF1). The NF1 subtype of neurofibromatosis results from a genetic defect on chromosome 17 and is far more common than NF2.[6]

Our case represents a solitary, localized tumor in an otherwise normal individual. These tumors usually involve small nerves and readily extend into soft tissue. They become circumscribed but not encapsulated.[6] Our

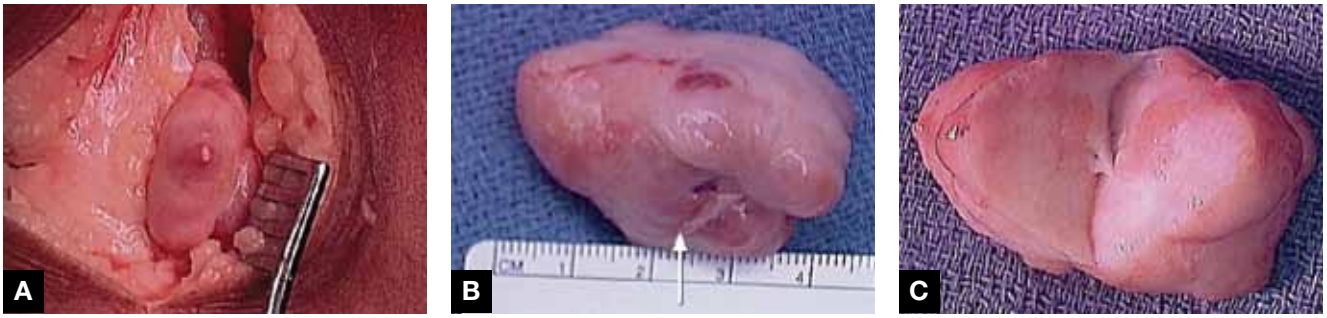

Figure 3. Operative findings show (A) the mass mobilized to the medial arthrotomy, (B) the excised smooth polypoid tumor with a small vascular pedicle (arrow), and (C) the cut surface of the tumor.

patient had no family history of neurofibromatosis. Physical examination revealed none of the other stigmata of this neurocutaneous disorder. This sporadic type presentation appears to be the most common presentation of neurofibromas. In one series, about 90% of neurofibromas were of the solitary or sporadic type, and the remainder were found associated with NF1.[7]

Neurofibromas have been found in a variety of locations. This tumor can potentially arise in any location where nonmyelinating Schwann cells are present. Both boney and soft tissue lesions are well documented, but intra-articular involvement appears quite rare, limited to a few case reports.[8-16] The rare cases of hip or knee joint involvement have occurred in patients with NF1 by a process of intra-articular penetration from neurofibromas originating in extra-articular locations. These extra-articular tumors can cause bony erosion and deformity, and even joint dislocation by intra-articular extension.[13-16]

One unusual case of a plexiform neurofibroma without any association with neurofibromatosis has been documented. This tumor was found in a subsynovial location beneath the iliotibial band of a multiply operated knee.[17] Our case of a true intra-articular neurofibroma, arising with a small pedicle, within the knee joint, is even more unusual.

Our findings suggest that the most likely tissue of origin of this intra-articular tumor is the posterior aspect of the infrapatellar fat pad and/or its extensions. Posteriorly, the fat pad projects into the intercondylar notch via 2 alar synovial folds, which fuse forming the infrapatellar plica or ligamentum mucosum.[1,3] Both the infrapatellar fat pad and the ligamentum mucosum have abundant vascular supplies[1,3,18] as well as rich innervations of type IVa nerve fibers.[19] These nerve fibers present a potential source of nonmyelinating Schwann cells.

This unusual case of an intra-articular neurofibroma originating from the area of the infrapatellar fat pad of the knee adds to the spectrum of pathology involving this region and to the differential diagnosis of fat pad lesions seen on MRI. In our case, the MRI was able to accurately localize and determine the extent of the lesion. It was able to point to a diagnosis of a complex cyst and/or a solid lesion, but not to a specific diagnosis. There is no doubt that a preoperative MRI evaluation with gadolinium enhancement would have added information leading to a more specific diagnosis. However, the specific diagnosis of a neurofibroma is rarely made without MRI demonstrating an unencapsulated soft tissue mass in continuity with a peripheral nerve.[20]

This case demonstrates the value of an arthrotomy in the diagnosis and treatment of this previously unreported condition. The arthrotomy allowed direct visualization of the entire tumor in situ and provided easy access for its complete removal while preserving the gross anatomy of an intact specimen for examination. Finally, this case documents a rare but potential source of anterior knee pain in the adult and contributes to understanding the spectrum of clinical presentations of neurofibromas.

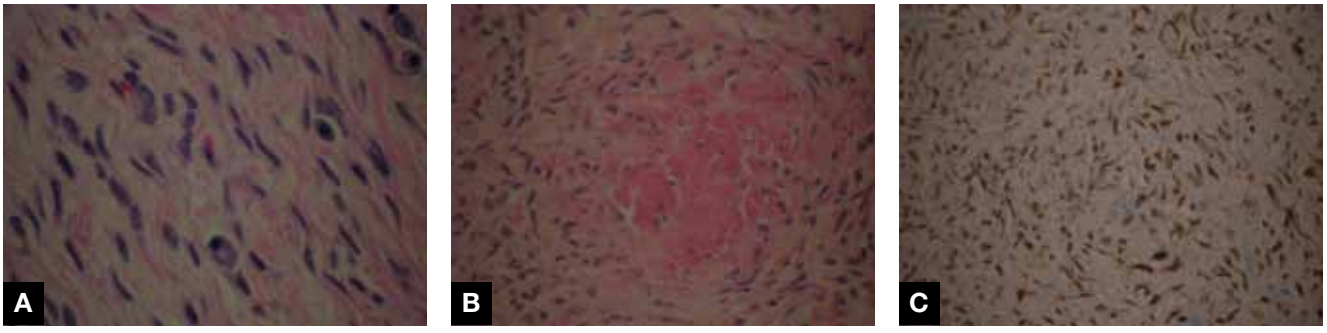

Figure 4. Histology of the tumor. (A) Hematoxylin and eosin (H/E) stained slide, original magnification x40, showing a cellular wavy spindle cell lesion with stromal mast cells. (B) H/E stained slide, original magnification x40, showing a pseudo-Meissner corpuscle, a characteristic finding in neurofibromas. (C) Immunoperoxidase stained slide, original magnification x 40, showing strong, diffuse, nuclear and cytoplasmic staining of the neoplastic cells.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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