Primary intra-articular epithelioid sarcoma of the knee: Unusual presentation of a rare entity

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ABSTRACT

We report a case of a 60-year-old female with severe and progressive pain of her right knee. Physical therapy, pain medication, and arthroscopic debridement were unsuccessful. Finally, pathological examination revealed an intra-articular epithelioid sarcoma, a rare tumor in an atypical location. Patient died within 5 months after initial admission. Despite this unusual clinical course and presentation, we would like to share the valuable clinical lessons we learned from this case. Introduction of a coordinating physician in combination with a multidisciplinary treatment regarding optimal pain management should optimize treatment results in future patients.

1. Case report

A 60-year-old female with a history of an arthroscopy of the left knee, lower back pain, hypertension, and bilateral carpal tunnel syndrome was admitted with pain in her right knee since she fell 6 months before. She reported exercise-induced swelling of her right knee not responding to non-steroidal anti-inflammatory drugs and physical therapy instituted by her general physician. Physical examination of the knee demonstrated a mild hydrops, a limited extension of 10°, no atrophy, and no joint line tenderness on palpation. Plain radiographs of the right knee demonstrated chondrocalcinosis of the medial and lateral meniscus. Magnetic resonance imaging (MRI) revealed a degenerative medial meniscus, popliteal 'synovial chondromatosis', and anterior cruciate ligament thickening which was interpreted as 'muroid degeneration' (Fig. 1). Because of the suspicion of a posttraumatic synovitis, patient received an intra-articular injection with corticosteroids. Patient was readmitted three weeks later with increased severity of right-sided knee pain. Physical examination showed more pronounced hydrops and a further deterioration in range of motion, both in flexion and extension. Because of persisting pain, an arthroscopic partial medial meniscectomy was performed with debridement of chondrocalcinosis and synovium in the femoral notch. The anterior cruciate ligament showed prominent thickening. Following surgery, patient complained of progressive pain and a further reduction in range of movement. An

urgently consulted rheumatologist ruled out rheumatological disease. For optimal pain management a pain specialist was consulted. Laboratory analyses of blood and joint aspirate, including cultures, were negative for an inflammatory process. MRI was repeated and showed extensive mass-forming lesions with nodular synovitis accompanied with deep cortical invasion (Fig. 2). Differential diagnosis included pigmented villonodular synovitis, although hemosiderin deposition was only noted in the popliteal fossa (on the initial MRI interpreted as synovial chondromatosis). Subsequent histological biopsy of the knee revealed an intra-articular located epithelioid sarcoma. On immunohistochemical analysis the tumor cells were focally positive for CD34, strongly keratin AE1 positive, positive for epithelial membrane antigen (EMA), and hepatic encephalopathy necrosis was visible (Fig. 3). Additional oncologic work-up demonstrated multiple hepatic and bone metastases. Palliative care was initiated focusing on pain relief. An above knee amputation was considered, but due to her rapid clinical deterioration she received a chordotomy instead. The patient deceased three weeks after the definitive diagnosis, 5 and 6 months after her first visit to hospital and her family physician, respectively.

2. Discussion

Epithelioid sarcoma is a soft-tissue sarcoma mostly occurring in adolescents and young adults with a significant male predominance.^{1–3}

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Fig. 1. Baseline MRI of the right knee. Sagittal fat-suppressed T2-weighted image depicts a thickened anterior cruciate ligament (arrowheads) and a nodular lesion in the popliteal fossa (arrow), which was interpreted as ‘synovial chondromatosis’, but more likely represents a localized form of pigmented villonodular synovitis considering the low-signal hemosiderin deposits lining the synovium. Some effusion is present in the suprapatellar bursa.

The classic variant consists of a small, slow-growing tumor, which is often painless and involves the distal extremities (i.e. the forearm and hand).^{1,4} The proximal form is aggressive and most often occurs in relatively older patients.^{3,5} An intra-articular location of epithelioid sarcoma is rare and only six cases have been published before, however, Chow (2015)⁶ questioned the intra-articular aspect of four of them.

Since an epithelioid sarcoma is often confused with other benign and malignant processes,^{1,3} a delay in diagnosis and treatment occurs frequently. Chase and Enzinger (1985)¹ reported a time interval between symptom onset and surgery of 2 years and 5 months. Conventional radiographs show calcifications or ossifications in 28% of the cases. Epithelioid sarcomas, however, rarely invade bone. Initial treatment consists of wide and aggressive surgery, often resulting in an amputation. Epithelioid sarcoma has a high local recurrence rate of 65–77%^{1,3} and more than 50% of patients develop metastases, especially to lungs and lymph nodes.^{1–3} Prognosis is often extremely poor. Generally, the more proximal the site of origin, the worse the outcome.¹ Factors associated with worse prognosis are male sex, haemorrhagic tumors, older age at presentation, proximal location, large tumor size, and early metastasis.^{1–3}

Our patient presented with atypical features of epithelioid sarcoma at various levels including demographics (elderly female), clinical

Fig. 2. A-B. Follow-up MRI. Sagittal fat-suppressed T2-weighted (A) and sagittal contrast-enhanced fat-suppressed T1-weighted (B) images show progressive thickening of the anterior cruciate ligament extending to adjacent bony structures (arrowheads) with extensive nodular mass-forming thickening of the synovium (white stars). Note also bone invasion of the patella and proximal tibia (white vector arrowheads). Nodular lesion in the popliteal fossa (arrow) represents a localized form of pigmented villonodular synovitis (susceptibility artifacts were present on gradient echo techniques, not shown).

Fig. 3. A-D. Immunohistochemical analysis. A. Hepatic encephalopathy necrosis is visible on the left. B. The tumor cells are focal CD34 positive. C. The cells are strongly keratin AE1 positive. D. They show positive staining for EMA.

course (rapidly growing tumor with excruciating pain initially non-responsive to pain therapy), and morphological findings (extensive bone invasion), which was rarely reported before. Delay in diagnosis and appropriate treatment in our patient was caused by several factors that include the initial benign appearance of epithelioid sarcoma on imaging, at physical examination, and during arthroscopy and the resulting misdiagnosis of a degenerative posttraumatic knee.

The non-standard procedures performed for knee pain in this elderly patient, including urgent diagnostic steps, arthroscopy, consult of a rheumatologist, and a second MRI scan, showed that there was ambiguity. In retrospect, one could argue there was a reason for suspicion following each of these diagnostic and therapeutic steps. The indeterminate appearances of the anterior cruciate ligament at the first MRI (where contrast enhanced images would have been helpful) and during arthroscopy could have led to diagnostic biopsies during arthroscopy. Whether an earlier diagnosis, however, would have resulted in a more favorable clinical course is doubtful, considering the aggressive clinical behavior that was worse than described previously.

The lessons we learned from this case led to modifications of current management protocols. First, one principal specialist or case manager will be assigned for every patient, who is deemed especially helpful in cases involving different disciplines. Second, unusual or equivocal morphologic findings on imaging are discussed during multidisciplinary meetings. Finally, adequate management for intractable pain that is not responsive to conventional therapy is addressed by consulting an anesthesiologists specialized in pain treatment at an early stage. In conclusion, this case report of an intra-articular epithelioid sarcoma with unusual clinical presentation and morphological findings, engendered optimization of institutional protocols.

Competing interests

The authors declare that they have no competing interests.

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Appendix A. Supplementary data

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.jor.2018.08.009>.

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