Case report

Intraarticular synovial sarcoma of the knee rising from a lateral meniscus – a case report

M. Bergovec a, b, *, M. Smerdelj a, F. Bacan a, S. Seiwerth c, D. Herceg d, M. Prutki e

a Department of Orthopaedic Surgery, Clinical Hospital Centre Zagreb, University of Zagreb Medical School, Salata 7, HR-10000 Zagreb, Croatia
b Department of Orthopaedics and Traumatology, Medical University Graz and University Hospital Graz, Auenbruggerplatz 5, A-8036 Graz, Austria
c Department of Pathology, School of Medicine, University of Zagreb, Salata 3, HR-10000 Zagreb, Croatia
d Department of Oncology, Clinical Hospital Centre Zagreb, University of Zagreb School of Medicine, Kipaticeva 12, HR-10000 Zagreb, Croatia
e Department of Radiology, Clinical Hospital Centre Zagreb, University of Zagreb School of Medicine, Kipaticeva 12, HR-10000 Zagreb, Croatia

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A B S T R A C T

Synovial sarcoma (SS) is a rare mesenchymal tumor, accounting less than 10% of soft tissue sarcomas. We report a case of intraarticular SS mimicking nodular synovitis and lateral meniscus rupture. Due to clinical and radiological presentation, arthroscopic synovectomy was performed, and histology confirmed nodular synovitis. After four years the lesion recurred and new arthroscopic biopsy was performed, revealing a monophasic SS with SYT/SSX translocation. Repeated histology of the first specimen confirmed appearance of a nodular synovitis microscopically, with no morphological criteria for a sarcoma, but molecular analysis showed positive SYT/SSX1 translocation. Wide extraarticular knee resection and reconstruction with a tumor megaendoprosthesis-allograft composite was performed with a negative tumor margins. This case report showed that in a case of benign histological appearance, underlying sarcoma is possible and could be identified in early stages only with an advanced pathology methods.

Level of evidence: Level IV historical case.

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1. Introduction

Synovial sarcoma (SS) is a rare mesenchymal tumor with variable degrees of epithelial differentiation (monophasic or biphasic), accounting for less than 10% of soft tissue sarcomas, whereas intraarticular SS is extremely rare [1–3]. Patients with localized SS and younger age are considered as positive prognostic factors.

The absence of characteristics clinical and/or radiological presentation poses a diagnostic challenge. While larger lesions tend to be on MRI more heterogeneous in signal intensity, smaller synovial sarcoma (<5 cm) may have well-circumscribed margins and homogeneous signal intensity on all MRI images, regardless of pulse sequence, the findings that simulate benign lesions, such as intraarticular localized nodular synovitis [4]. Molecular analysis revealed that more than 95% of cases of SS have a specific chromosomal translocation t(X;18)(p11;q11) that results in a SYT-SSX gene fusion, considered to be the main oncogenic event in the SS oncogenesis [5,6]. For that reason molecular analysis of biopsy material should be the gold standard in SS diagnosis.

2. Case description

A 26-year-old man presented with a 6-month history of recurrent dull pain and a localized swelling localized at a lateral side of the left knee. MRI of the knee showed a lobulated mass at the lateral compartment, rising from the lateral meniscus, affecting lateral collateral ligament, and measuring 3 × 1.5 cm. The mass was well circumscribed, isointense relative to skeletal muscle on T1-weighted images and hyperintense on T2-weighted images. Punctiform voids seen on gradient-echo images matched calcifications (Fig. 1). Arthroscopy of the knee was performed, and a lateral meniscus tear and a nodular synovitis have been revealed (Fig. 2). A part of the tumor was taken for a histology analysis. Synovectomy with a motorized shaver of lateral part of joint was performed, and lateral meniscus was sutured. Histological analysis revealed a spindle cell proliferation composed of monotonous cells with ovoid nuclei and smooth nuclear contours and scant cytoplasm embedded in the collagenous tissue with areas of increased vascularization and ossification. Mitosis was rare. Cytological atypia

* Corresponding author. Department of orthopaedic and Traumatology, Medical University Graz and University Hospital Graz, Auenbruggerplatz 5, A-8036 Graz, Austria. E-mail address: marko.bergovec@medunigraz.at (M. Bergovec).

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and area of necrosis were not present (Fig. 3), thus rendering the diagnosis of nodular synovitis.

Four years later the patient got a recurrence with a similar symptoms. MRI revealed lobulated intraarticular mass with mild rim enhancement surrounding lateral meniscus indicating the formation of a tumor (Fig. 4). A re-biopsy was performed arthroscopically. Histological findings showed atypical spindle cell proliferation arranged in sheets and fascicles that occasionally presented with a typical “herring-bone” pattern (Fig. 5A). Immunohistochemical staining with AE1/AE3 (Fig. 5B) and vimentin revealed focal positive reaction suggesting the diagnosis of intraarticular monophasic SS. Molecular analysis using PCR showed SYT-SSX1 fusion (Fig. 5 A and B) confirming the diagnosis.

We repeated a histological analysis of a specimen obtained during first arthroscopy four years ago, which again confirmed finding of a spindle cell proliferation with rare mitotic figures, without cytological atypia and necrosis suggesting a benign lesion. No solid histological criteria for sarcoma were found. Still, molecular analysis of that first specimen was done and confirmed the diagnosis of monophasic SS with a finding of a SYT-SSX1 gene fusion.

Surgical treatment consisted of an extraarticular en block resection of the knee joint, resection margins were tumor-free. Reconstruction of the knee was made with a tumor endoprosthesis and a bone-tendon allograft (Figs. 6 and 7). A clinical result of the operation was good: the patient was full weight-bearing, mobile without an aid, with an active knee range of motion 0°–100°.

Fig. 1. First MRI of the knee (coronal PD fat suppressed MR image).

Fig. 2. Intraoperative arthroscopy finding of the lateral knee compartment: an appearance of a nodular synovitis and a lateral meniscus rupture.

Fig. 3. The first histology examination of the arthroscopically obtained specimen. (H & E staining, objective ×10).

Fig. 4. MRI performed 4 years after the primary operation. Intraarticular lobulated mass surrounding lateral meniscus has high signal intensity on T2-weighted MR image (a) and is isointense to muscle on T1-weighted image (b) (arrow). T1-weighted MR image following intravenous gadolinium-based contrast administration shows mild rim enhancement of the lesion (c) (arrowhead).
Thirty-nine months after the wide resection, a local metastasis in a proximal part of the quadriceps muscle with an erosion of the proximal femur was found. The patient was treated with a hip disarticulation. At the latest follow-up 9 months after the amputation, the patient is alive, with no evidence of local recurrence or distant metastasis.

3. Discussion

To our knowledge, this is a first report of a patient with a positive molecular finding for a sarcoma by seemingly non-malignant primary lesion. Even more, this case report showed that in a case of a benign histological appearance, underlying sarcoma is possible and could be in early stages identified with an ancillary (molecular) methods.

Orthopaedic surgeons should be acquainted with a variety of different tumor presentations. By suspicion of a tumor, patient should be forwarded to a hospital specialized in orthopaedic oncology. Additionally, radiologists and orthopaedic surgeon should be aware that the majority of intraarticular synovial sarcomas less than 5 cm in size have presented with non-specific MRI features, leading to their misinterpretation [2]. If clinical presentation and non-specific MR findings raise suspicion of intraarticular sarcoma, an excision biopsy should not be performed arthroscopically. Intraarticular insufflation of the knee with saline would risk spread of tumor cells, which may significantly alter any future limb salvage reconstruction. Evaluation and treatment of all patients with suspected sarcomas should be undertaken by a musculoskeletal oncologist.

Surgical wide resection with tumor-free margins is a must in a soft tissue sarcoma treatment [1]. For the intraarticular located tumors achieving clear margins is possible only with an extraarticular resection. Retaining any part of a joint capsule carries a risk of sarcoma recurrence [7]. Since use of large allografts carries a significant risk of deep infection, especially in knee region [8], we decided to perform reconstruction of the extensor mechanism with an allograft: the tibial tuberosity, patellar tendon, patella, and quadriceps tendon in continuity, sutured to quadriceps in the proximal part and fixed to endoprosthesis distally, covered with a rotational medial gastrocnemius flap. This true extraarticular knee resection and reconstruction with endoprosthesis and a patella-tibial tuberosity allograft worked well in this patient in a 3.5 years follow-up, before the occurrence of the local metastasis and the need for amputation.
4. Conclusion

Future development in bone and soft tissue sarcoma diagnosis will highly depend on an advanced molecular analysis. The main benefit from these tests is high specificity for some tumors. This case report shows the possibility of discovering the sarcoma in very early stages, before positive histological findings. Cost benefit analysis of molecular analysis is yet to be determined.

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Disclosure of interest

The authors declare that they have no competing interest.

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