Synovial sarcoma: a case report and review of the literature

Case report/Opis przypadku © J ORTHOP TRAUMA SURG REL RES 2 (32) 2013

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Summary
Synovial sarcoma is the fourth most common type of soft-tissue sarcoma, accounting about 10% of all primary soft-tissue malignancies worldwide. Synovial sarcoma most often affects the extremities, particularly the knee in the popliteal fossa, of adolescents and young adults (15–40 years of age). Despite its name, the lesion does not commonly arise in an intraarticular location but usually occurs near joints. Although radiographic features of these tumors are not pathognomonic, findings of a soft-tissue mass, particularly if calcified (30%), near but not in a joint of a young patient, are very suggestive of the diagnosis. Two features associated with synovial sarcoma that may lead to an initial mistaken diagnosis of a benign indolent process are slow growth (average time to diagnosis, 2–4 years) and small size (< 5 cm at initial presentation). Synovial sarcoma is an intermediate- to high-grade lesion, and, despite initial aggressive wide surgical resection, local recurrence and metastatic disease are common and prognosis is guarded. Understanding and recognizing the spectrum of appearances of synovial sarcoma are important for optimal patient management.
Key words: synovial sarkoma, soft tissue sarkoma

INTRODUCTION

Synovial sarcoma was first reported in 1893 and represents a relatively common type of primary soft-tissue malignancy [1]. Synovial sarcoma accounts about 10% of all primary malignant soft-tissue neoplasms [2,3]. Despite this nomenclature, these lesions do not arise in an intra-articular location but usually occur near joints. Although it is often found to be in close association with tendon sheaths, bursae and joint capsules [4]. Synovial sarcomas typically affect adolescents and young adults. The extremities, particularly the knee in the popliteal fossa, are most frequently affected. Synovial sarcoma is an intermediate- to high-grade neoplasm with extensive metastatic potential. There are three main histologic subtypes of synovial sarcoma: biphasic, monophasic, and poorly differentiated. Biphasic synovial sarcoma represents 20%–30% of lesions and has both a mesenchymal spindle cell component and an obvious epithelial component. Monophasic synovial sarcoma represents 50%–60% (the most common subtype) of all lesions, and in this subtype the mesenchymal spindle cell component predominates. Because of the aggressive potential behavior of synovial sarcoma, pathologic and radiologic assessment is important for staging and evaluating lesion extent to direct appropriate therapy. Imaging findings, although not pathognomonic, frequently suggest the diagnosis. Radiographic findings of a soft-tissue mass near but not in a joint in a young patient (15–40 years old), particularly if calcification is present, are very suggestive of synovial sarcoma [5]. In this article, the clinical features, pathologic characteristics and treatment and prognosis of synovial sarcoma are discussed.

CASE REPORT

A 32-year-old male noticed a palpable mass in his forearm in January 2013. He had no abnormality in the family history. The tumor was growing slowly, had a benign appearance and was giving a pain similar to that associated with trauma. The symptoms suggested a benign process such as tendonitis of biceps muscle. Patient visited our clinic in March 2013. On physical examination, a 1 × 2cm, well defined and movable mass with a smooth surface was palpable in the left forearm near elbow. Imaging studies including MRI and US demonstrated a round solid and inhomogeneus soft tissue (3cm in greatest diameter) associated with biceps tendon. The surgical resection with adequate margin was performed in April 2013. (Wide resection consisted of removal of the tumor with normal tissue completely surrounding it). The histopathology examination revealed that the margin was free of tumor cells. The diagnose was confirmed: Synovial Sarcoma. (Biphasic type)

Patient was referred to oncology center.

CLINICAL CHARACTERISTICS

Synovial sarcoma is the fourth most common soft-tissue sarcoma following malignant fibrous histiocytoma (currently known as undifferentiated high-grade pleomorphic sarcoma), liposarcoma, and rhabdomyosarcoma [1,2,3]. Synovial sarcoma occurs most frequently in adolescents and young adults, with the majority of patients presenting at 15–40 years of age [2,3]. A mild male predominance (1.2:1 ratio) has been described by some authors [6]. No race or ethnic predilection has been reported. Patients with synovial sarcoma usually present with a palpable soft-tissue mass or swelling [2]. These lesions are often slow growing initially. Duration of symptoms before diagnosis varies widely, from weeks to years [2]. In the majority of cases the disease duration prior tosurgery ranges from 2 to 4 years, but a slow-growing mass or pain at the tumor site has been noted for as long as 20 years prior to surgery [1,4]. The long duration of symptoms and initial slow growth of synovial sarcomas may give a false impression of a benign process. This unusual manifestation is important to recognize, because diagnosis may be significantly delayed otherwise. Pain and tenderness at the site of the mass are frequent, and some patients present with pain but no palpable mass [2]. This symptom is unusual compared with other soft-tissue sarcomas that typically manifest as painless masses. The majority of synovial sarcomas (80%–95%) occur in the extremities. The lower extremity is most often affected, accounting for 60%–71% of cases, whereas 16%–25% occur in the upper limb. The single most frequent site of involvement is the popliteal fossa of the knee [2,3,5,6]. However, synovial sarcoma has also been reported in almost all anatomic locations, with rare involvement of head and neck, thorax, and chest wall, pelvis, skin, viscera, central nervous system and bones [1]. The most common site for metastasis is the lung(90-95%). However lymph node involvement has been reported in large studies to occur in 3-27% of patients [4].

Computed tomography and magnetic resonance imaging (MRI) are valuable tools in determining the site of origin and extent of the lesion. The imaging appearance is nonspecific, and in all cases a biopsy is necessary to confirm the diagnosis. It is not infrequent for the imaging studies to be suggestive of a benign process, such as a cyst or hematoma, which often can lead to inadvertent marginal excisions [4,7].

TREATMENT AND PROGNOSIS

As with many intermediate- to high-grade primary malignant soft-tissue neoplasms, local control of synovial sarcoma is primarily achieved with surgery. However, because synovial sarcoma commonly occurs near large joints and neurovascular structures, radical surgical excision that leaves an adequately functional limb may be difficult or impossible. For that reason, the current treatment of choice is wide local excision (removal of the tumor and a normal cuff of surrounding tissue). The surgical margins should be closely evaluated to determine the need for adjuvant therapy. As expected, marginal excision of the synovial sarcoma without removing an

adequate rim of normal surrounding tissue is associated with high local recurrence rates (70%–83%) [8]. The minimal acceptable margin has not been established; however, the surgeon must be aware of the susceptibility for microscopic infiltration of tumor cells into the pseudocapsule of the tumor [9].

Amputation should be reserved for those cases in which gross resection of the tumor and preservation of a functional limb is not possible. The role of adjuvant therapy in the treatment of synovial sarcoma remains controversial [10]. Chemotherapy has been used to treat metastatic or residual disease. Studies have shown a limited survival benefit for high-risk patients following adjuvant chemotherapy [8]. Patients with synovial sarcoma have a 5-year survival rate ranging from 36% to 76%. At 10 years, the survival rate has been reported to range from 20% to 63% [5]. The clinical course of synovial sarcoma is characterized by a high rate of local recurrence and metastatic disease. The majority of metastases occur within the first 2–5 years after treatment. The most frequent metastatic site is the lung. Multiple clinical and pathologic factors, including tumor size, location, patient age, and presence of poorly differentiated areas, have prognostic significance. Tumor size greater than 5 cm at presentation has the greatest impact on prognosis, with studies showing 5-year survival rates of 64% and 26% for patients with tumors less than 5 cm and those with masses greater than 5 cm [5]. Some authors have reported a poorer prognosis for patients with synovial sarcomas located in the proximal extremities. Others have reported better prognosis for patients with tumors located in the upper extremities compared with those with lower extremity lesions. Patient age of less than 15–20 years is also associated with a better long-term prognosis. Some authors reported that the presence of extensive calcifications suggests improved long-term survival. There is considerable controversy about the prognostic significance of tumor cell type (monophasic or biphasic). However, the poorly differentiated subtype is associated with a worsened prognosis, with a 5-year survival rate of only 20%–30% [8].

Synovial sarcomas are characterized by the t(X;18)(p11;q11) translocation. Many studies have delineated the 5-year overall survival rates and 5-year metastasis-free survival rates in patients containing two gene mutations, SYT-SSX1 and SYT-SSX2. Two studies with cohorts of 104 and 243 synovial sarcoma patients showed the overall 5-year metastasis-free survival for patients with SYT-SSX1 ranged from 42% to 53% versus 73% to 89% for those with SYT-SSX2 [11]. Thus, detection of the presence and type of SYT-SSX fusion may be important for diagnosis and prognosis.

In a 2004 study of 271 patients with synovial sarcoma treated at a single institution, Ferrari et al [5] reported the 5-year overall survival and the 5-year metastatic-free survival rate in 215 patients (tumors often occurred in close proximity to large joints of the extremities, particularly the knee in the popliteal fossa) with nonmetastatic surgically resected disease as 71% and 51%, respectively. The metastatic-free survival rate was 60% for patients who received chemotherapy compared to 48% for those who did not. Adult patients who received chemotherapy did better than those who did not, especially among those who had tumors measuring >5 cm (metastatic-free survival, 47% versus 27%). The investigators concluded that their retrospective findings suggest that all patients with tumors >5 cm should receive chemotherapy.

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