A Case of Primary Intra-abdominal Synovial Sarcoma

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Abstract

Synovial sarcomas are relatively common intermediate-to-high-grade malignant soft tissue tumors, often with an initial indolent course. And among the sarcomas primary intra-abdominal synovial sarcoma is a relatively rare entity that may present with an abdominal mass and diagnosis is usually confirmed by immunohistochemistry. The authors report a case of a 46-year-old man who presented with a large palpable abdominal mass. Contrast-enhanced computed tomography (CT) scan showed a large well-defined heterogeneous intra-abdominal mass filling the entire pelvis and extending upwards till the subhepatic region causing displacement of intra-abdominal organs. However, no other focal lesions were seen in the solid organs. The same findings were confirmed on surgery. Histopathology was suggestive of desmoid tumor/hemangiopericytoma. Immunohistochemistry showed positive markers corresponding to synovial sarcoma. The patient was advised to undergo chemotherapy which was refused and follow-up was lost. After 10 months, patient presented for follow-up CT, which showed marked increase in size of the lesion.

Keywords Synovial · Sarcoma · Abdominal

Introduction

Synovial sarcoma (SS) was first reported in 1893 and represents a relatively common type of primary soft tissue malignancy. Synovial sarcoma accounts for 2.5–10.5% of all primary malignant soft tissue neoplasms [1]. In spite of its name, synovial sarcoma is not related to the synovial tissues that are a part of the joints. The disease starts most commonly in the legs or arms, but it can appear in any part of the body. On pathology, synovial sarcoma may be classified in different subtypes depending on what it looks like under the microscope or what specific gene mutation is involved. Synovial sarcoma is a high-grade tumor. It spreads to distant sites in up to 50% of cases. There are no well-established risk factors for synovial sarcoma, but the disease is associated with the chromosomal translocation t(X;18) (p11;q11). This means that parts of chromosome 18 and chromosome X have switched places in synovial sarcoma tumor

cells. It is not known whether this mutation occurs randomly or follows a certain chain of events. Because of this translocation, synovial sarcoma cells become mutant. This mutant gene is thought to contribute to the development of the disease. Whereas benign neoplasms of soft tissues (i.e., lipoma, fibroma, leiomyoma, hemangioma) are relatively frequent and outnumber malignant cases by a factor of 50 (incidence of about 300 new cases per 100,000 persons) [2], malignant soft tissue tumors are rare, with an annual incidence of about 6 per 100,000 persons [2]. Soft tissue sarcoma (STS) occurs throughout childhood and adulthood; they account for about 1.5% of all malignant tumors in adults (and 2% of all cancer-related deaths), but represent about 7.4% of all pediatric malignancies (although their absolute number is lower than in adults) [2]. According to previous reports, only 5–10% of synovial sarcomas are in the head and neck, mediastinum, abdominal wall, esophagus, and retroperitoneum [3]. To date, synovial sarcomas have also been found to occur in several unusual locations, including skin, blood vessels, nerves, mediastinum, pleural cavity, prostate, and kidney [3]. Primary intra-abdominal synovial sarcoma has been mentioned as a rare case in several reports. Usually, primary imaging via computed tomography (CT) or magnetic resonance imaging (MRI) helps in identifying the mass followed by biopsy which is essential for establishing the diagnosis. Here, we discuss a case of a 46-year-old male patient who presented with intra-abdominal mass to our hospital that eventually turned out to be primary intra-abdominal synovial sarcoma following biopsy.

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Case Report

Here, we present a case of a 46-year-old man who came to our hospital with complaints of gradually increasing painless swelling in the abdomen for the past 2 years. Clinical examination revealed a large mobile mass in lower abdomen. A contrast-enhanced CT was performed which showed a large heterodense, multilobulated lesion occupying the entire pelvis and extending up to subhepatic region. No discernible fat plane was noted between the lesion and the rectus muscle in right iliac fossa. The lesion was seen displacing the right iliac vessels medially in the pelvis and extending upwards in the abdomen encasing the superior mesenteric vessels with an arc of contact $< 90^\circ$. The right ureter and kidney were closely abutting the lesion but there was no evidence of locoregional invasion into the adjacent viscera. There was no evidence of any visible distant metastasis from the mass. Hence, as per contrast-enhanced CT, the lesion was deemed operable to a large extent. Subsequently, a contrast-enhanced MRI was done, which showed a heterogeneously enhancing lesion extending from the pelvis with mixed signal intensity.

So report was given as an aggressive lesion arising from the pelvis probably desmoid tumor from muscle. Operation of the mass was planned. Preoperative lab investigations were unremarkable with normal routine blood and hemogram. At surgery, a large lobulated well-circumscribed mass was noted, which was adherent to the mesentery and small bowel. Approximately 95% of the lesion was removed since intra-operatively complete removal was not possible due to close adhesion between the mass and surrounding mesentery as described. Histopathology report was suggestive of desmoid tumor/hemangiopericytoma.

For confirmation, immunohistochemistry (IHC) was performed. Markers, EMA and CK7, were positive correlating with that of synovial sarcoma.

Patient was referred for chemotherapy. After that, follow-up was lost. Ten months later, patient came back.

He did not undergo chemotherapy in the interim. A repeat CT was performed. Compared with previous CT, the lesion showed marked increase in size.

Case Discussion

Synovial sarcoma is a misnomer because the tumor does not arise from the synovium; it only resembles synovial tissue at light microscopy. It appears to arise from as yet unknown multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures and lack synovial differentiation [4]. Synovial sarcoma occurs most commonly in the young, representing about 8% of all soft tissue sarcomas but about 15–20% of the cases occur in adolescents and young adults [5].

SS can arise anywhere in the soft part of the body, generally as a progressively expanding mass. The most common clinical presentation is a slow-growing mass in the soft tissues of the lower extremity, especially around the knee and the ankle. The tumor is often near a joint, tendon, or bursa. Head and neck region, abdominal wall, retroperitoneum, mediastinum, pleura, lungs, and other organs are less common locations.

The most common presentation is usually a painless mass. However, the symptoms depend on the site of the tumor/mass. Difficulty in swallowing and breathing, or alteration of voice, for example, might be associated with SS of the head and neck region. Pain may be related to the involvement of nerves.

The gross appearance depends on the rate of growth and site of the tumor. The tumors that are slow growing are well-circumscribed, firm, round, or multinodular lesion. Cystic changes may be prominent. Rapidly growing tumors are poorly circumscribed with a variegated, friable appearance. There may be areas of hemorrhage and necrosis within the tumor.

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 as seen at light microscopy [1]. Monophasic synovial sarcoma represents 50–60% (the most common subtype) of all lesions, and in this subtype, the mesenchymal spindle cell component predominates [1]. Poorly differentiated synovial sarcomas are generally epithelioid in morphology and have high mitotic activity (usually 15–20/10 high-power field) with geographic necrosis. This subtype represents up to 15–25% of all synovial sarcomas [1].

The presence of keratin (epithelial marker) positivity (approximately 90% of cases), measured with immunohistochemical staining in correlation with the histologic appearance, is diagnostic for synovial sarcoma. Both the glandular component (diffusely) and the spindle cell component (focally) demonstrate single cells (monophasic and poorly differentiated subtypes) or clusters of cells (biphasic subtype) that are positive for epithelial markers, most notably pankeratins, EMA, and CK7 [1].

Radiographs appear normal in approximately 50% of cases of synovial sarcoma, particularly those with small lesions [6]. Synovial sarcomas detected at radiography typically appear as nonspecific, round to oval juxtaarticular soft tissue masses. Calcification is identified in up to 30% of synovial sarcomas at radiography [6]. The extremities are the most common primary site of synovial sarcoma. The lower extremities account for approximately 70% of cases [7]. Most synovial sarcomas occur in the popliteal fossa, but this tumor also affects the foot and ankle, upper extremity, hand and wrist, and proximal limb girdle. CT typically shows a noninfiltrative, well-circumscribed mass, often with punctate, peripheral calcifications [7].

MRI is the modality of choice for the diagnosis and initial staging of synovial sarcoma because of the information provided by intrinsic signal characteristics and superior soft tissue contrast. Typical MRI findings include a mass larger than 5 cm near a joint, deep in the tissue, and in contact with bone. Aggressive features such as involvement of adjacent bone, neurovascular encasement, and muscular invasion can be optimally assessed with MRI. A previous study of dynamic gadolinium-enhanced MRI of synovial sarcoma showed that the only feature highly associated with malignancy was early enhancement, that is, within 7 s after arterial enhancement [7]. Fluoro-deoxy-glucose positron emission tomography (FDG PET) may be helpful for prognosis because pretreatment FDG avidity greater than 4.4 is reportedly associated with increased risk of local recurrence and metastatic disease. Like other soft tissue sarcomas, synovial sarcoma metastasizes mainly to the lung. Unlike most other soft tissue sarcomas, however, synovial sarcoma carries a small risk of spread to lymph nodes. Soft tissue sarcomas originating from extremities rarely metastasize to the liver [7]. Metastatic lesions also show evidence of calcification like primary lesions. Retroperitoneal organs can also be sites of primary disease [8]. Primary pericardial synovial sarcoma is rare, previously reported to account for approximately 5% of primary pericardial neoplasms [9].

Primary intraabdominal synovial sarcoma is rare, retroperitoneal lesions accounting for fewer than 1% of all cases of primary synovial sarcoma [7]. CT is valuable in detecting aggressive tumors, predicting resectability, detecting distant metastases, and evaluating response to treatment. Tumor masses appear large, irregular, and necrotic with heterogeneous contrast enhancement. MR imaging of synovial sarcomas reveals a mosaic of mixed low, intermediate, and high signal intensity on T2-weighted images as a result of the admixture of solid components, cystic degeneration, hemorrhage, myxoid stroma, and fibrous tissue. The main differentials for intra-abdominal synovial sarcoma include fibrosarcomas, malignant schwannomas, malignant fibrous histiocytomas and, in rare instances, carcinomas such as adenocarcinomas. The most common CT appearance of synovial sarcoma is mixed soft tissue with attenuation equal to or slightly less than that of muscle. The heterogeneous areas may represent necrosis or hemorrhage of the tumor. MRI images appear isointense to slightly hyperintense on T1-weighted images, and hyperintense to muscles on T2-weighted images. Marked heterogeneity and enhancement are highly suggestive of synovial sarcoma on both CT and MRI [10]. Hence, in our case report as well as in previous literature, imaging was not seen to provide any definitive diagnostic feature for intra-abdominal synovial sarcoma and the final diagnosis was always dependent on histopathology correlation.

Conclusion

Primary intra-abdominal synovial sarcoma is a rare entity and is known for high recurrence rate and mortality. Hence, this should be kept in the differential diagnosis for aggressive intra-abdominal masses since early detection can have a significant impact on the treatment.

References

1. Murphey MD, Gibson MS, Jennings BT, Crespo-Rodríguez AM, Fanburg-Smith J, Gajewski DA (2006) Imaging of synovial sarcoma with radiologic-pathologic correlation. *Radiographics* 26(5): 1543–1565
2. Ferrari A, Sultan I, Huang TT, Rodriguez-Galindo C, Shehadeh A, Meazza C, Ness KK, Casanova M, Spunt SL (2011) Soft tissue sarcoma across the age spectrum: a population-based study from the surveillance epidemiology and end results database. *Pediatr Blood Cancer* 57(6):943–949
3. Wang YJ, Wen SC, Chien ST, Sheu JW, Hsueh CW, Feng NH (2006) Primary intra-abdominal synovial sarcoma. *J Chin Med Assoc* 69(10):492–495
4. Braham E, Aloui S, Aouadi S, Drira I, Kilani T, El Mezni F (2013) Synovial sarcoma of the chest wall: a case report and literature review. *Ann Transl Med* 1(1)
5. Weiss SW, Goldblum JR (2001) *Enzinger and Weiss's soft tissue tumors*. Mosby, Inc. A Harcourt Health Sciences Company, St Louis
6. Horowitz A, Resnick D, Caird Watson R (1973) The roentgen features of synovial sarcomas. *Clin Radiol* 24(4):481–484
7. Bakri A, Shinagare AB, Krajewski KM, Howard SA, Jagannathan JP, Hornick JL, Ramaiya NH (2012) Synovial sarcoma: imaging features of common and uncommon primary sites, metastatic patterns, and treatment response. *Am J Roentgenol* 199(2):W208–W215
8. Miyashita T, Imamura T, Ishikawa Y, Okinaga K, Kunii O, Miyashita H (1994) Primary retroperitoneal synovial sarcoma. *Intern Med* 33(11):692–696
9. Cina SJ, Smialek JE, Burke AP, Virmani R, Hutchins GM (1996) Primary cardiac tumors causing sudden death: a review of the literature. *Am J Forensic Med Pathol* 17(4):271–281
10. Kritsaneepaiboon S, Sangkhathat S, Mitarnun W (2015) Primary synovial sarcoma of the abdominal wall: a case report and literature review. *J Radiol Case Rep* 9(7):47–52