Case Report

Synovial sarcoma of the abdominal wall: An unusual presentation

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ABSTRACT

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm which commonly occurs in the extremities in close association with tendon sheaths, bursae, joint capsules, and fascial structures. Rarely, SS may be present in unexpected location such as the abdominal wall. Surgical resection with wide margins is the initial standard treatment; however, a multimodal approach including radiotherapy and chemotherapy is often favored. Here, we present a case of SS of the anterior abdominal wall in a 14-year-old patient with a right upper abdominal lump. He underwent wide surgical excision and has received adjuvant chemotherapy. He is doing well on follow-up of six months.

Key words: Anterior abdominal wall, soft tissue sarcoma, synovial sarcoma

INTRODUCTION

The term "synovial sarcoma" (SS) was coined to denominate tumors arising near the tendon sheaths and the joint capsules. Despite its name, SS does not arise from the synovial membrane but rather from a yet unknown multipotent stem cells that are capable of differentiating into mesenchymal and/or epithelial structures and lack synovial differentiation.[1] SS is a rare tumor, accounting for 5% to 10% of soft tissue sarcomas. This tumor rarely arises from an intra-articular location, although it usually arises in an extremity or limb girdle. Rarely, SS may be present in unexpected location such as the abdominal wall.[2] Here, we report a case of SS of anterior abdominal wall in a 14-year-old patient who underwent complete surgical excision and is doing well on follow-up of six months.

CASE REPORT

A 14-year-old male had presented to our hospital with complaints of low-grade diffuse pain in the right upper abdomen since six months, with an increase in the pain intensity since 15 days. He had also noticed a lump in the right upper abdomen since one month which was gradually increasing in size. He did not have any associated complaints of fever, vomiting, jaundice, urinary, or bowel defects. On examination, a lump of approximately 12 cm in diameter, mildly tender, firm in consistency, having an irregular surface was palpable in the right upper quadrant of the abdomen. Clinical examination suggested that this mass was fixed to the muscles of the abdominal wall.

All hematological and biochemical investigations were within normal limits. On ultrasonography of the abdomen, a 12 cm × 11.5 cm sized homogenous solid mass was seen related to the medial surface of the right lobe of liver, posteriorly compressing the pancreatic head and the inferior vena cava, and hence, suggesting a possibility of a pancreatic mass. Computed tomography scans of the abdomen [Figure 1] revealed an 11.3 cm × 10 cm sized hypodense lesion having heterogeneous contrast enhancement, probably arising from the porta hepatis and involving the anterior abdominal wall. There was also a resultant intrahepatic biliary radicle dilatation with compression of the stomach and gall bladder without involving them. Origin of the mass from the liver or the abdominal wall was unclear.

In view of the above investigations, the patient was posted

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for exploratory laparotomy. On laparotomy, the mass was seen arising from the rectus abdominis involving the perichondrium of the 12th rib and compressing the inferior surface of the liver without involving the liver parenchyma or the biliary ducts. The mass was excised with a wide margin (including partial excision of the right rectus abdominus, 12th rib). During intraoperative dissection of the mass, a rent occurred in the right hemidiaphragm with breach of the right pleura. The diaphragmatic rent was repaired, and a right-sided intercostal drain was kept. The abdominal defect was closed with a prolene mesh. The postoperative recovery was uneventful.

Histopathological examination of the specimen showed oval to spindle pleomorphic cells in a loose myxoid stroma suggesting a high-grade spindle cell sarcoma. On immunohistochemistry analysis, the tumor was positive for immunohistochemistry markers such as EMA, pancytokeratin [Figure 2a], Bcl-2 [Figure 2b], MIC-2 [Figure 2c], and Ki-67 and focally positive for CD34 marker and negative for smooth muscle actin, which suggested of SS.

The patient has received six cycles of adjuvant chemotherapy (ifosfamide and adriamycin) and is asymptomatic on follow-up of six months.

DISCUSSION

SS is the fourth most common soft tissue sarcoma after malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma.[3] About 85% to 90% of the tumors arise from the extremities.[2,3] The most common location of origin is near the knee in the popliteal fossa. The tumors occur most frequently in adolescents and young adults, with 30% of SS presenting in the first two decades of life with the median age of presentation being 13 years.[4,5] SS of the abdominal wall is more common in females, while SS in the extremities and the neck are more common in males.[2] No race or ethnic predilection have been reported.[4]

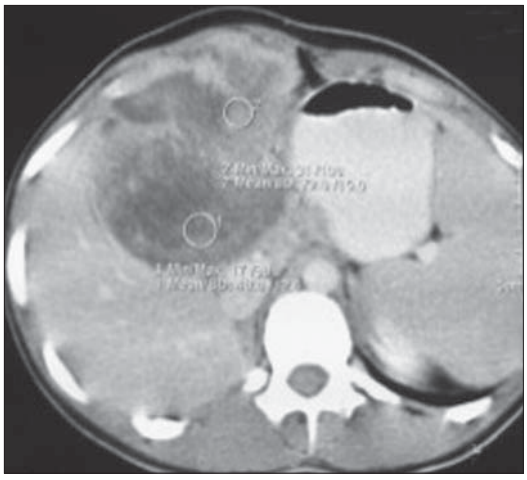

Figure 1: CT scan of the abdomen showing a hypodense lesion anteriorly involving the anterior abdominal wall and posteriorly probably arising from the liver

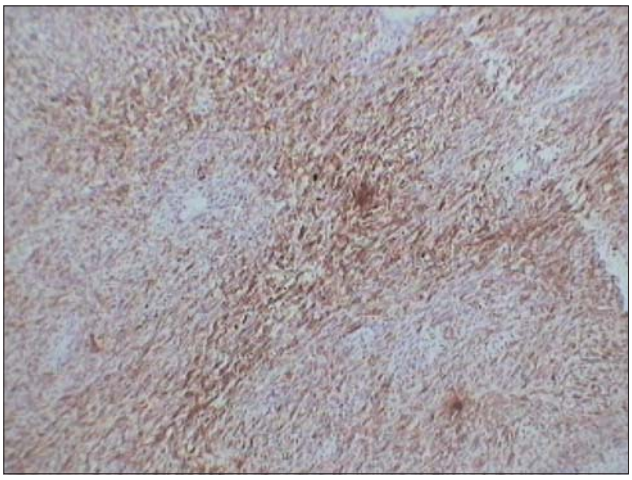

Figure 2a: Immunohistochemistry: Tumor cells positive for pancytokeratin (×10)

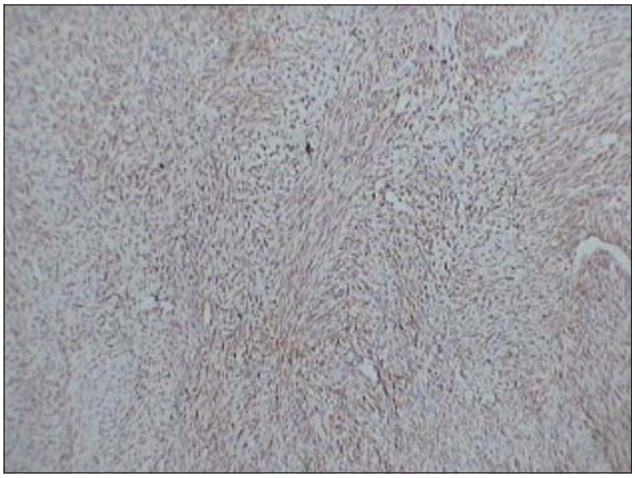

Figure 2b: Immunohistochemistry: Tumor cells positive for Bcl-2 (×10)

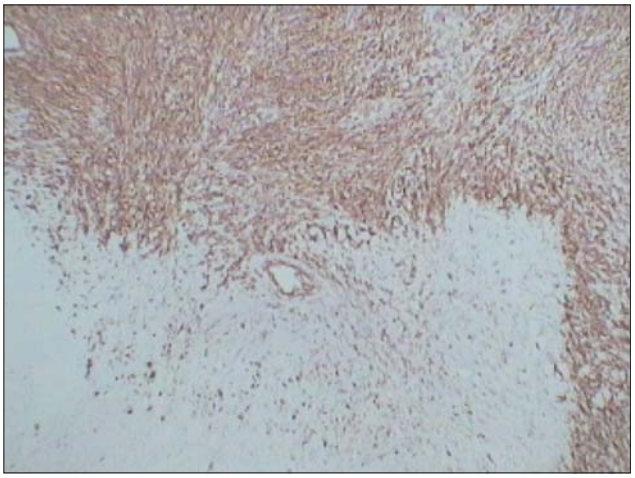

Figure 2c: Immunohistochemistry: Tumor cells positive for MIC-2 (×10)

Although the etiology of SS is unknown, almost all SS cells are characterized by a translocation fusing of two genes (*SYT*) located on the chromosome 19q11, and *SSX1*, *SSX2*, or *SXX4* located on the chromosome Xp11 breakpoint. Tumors expressing the *SYT-SSX1* fusion proteins more often exhibit biphasic histology, a higher proliferation rate, and are associated with a poor outcome. Monophasic tumors express either *SYT-SSX1* or *SYT-SSX2* genetic rearrangement.[2,4,6]

SS usually presents as a slow growing, palpable soft tissue swelling. Occasionally, the swelling may be associated with pain or tenderness and possible minor limitation to range of motion. Constitutional symptoms such as weight loss are rare. Abdominal SSs may present with vague intestinal symptoms along with abdominal pain or lump. These tumors usually occur equally on both sides of the abdomen and are more common in the lower half of the abdomen. 16% to 25% of patients at their initial presentation have metastasis to the lungs (the most frequent site), while others being the bone, the bone marrow, and the lymph nodes.[4,7]

There are three main histologic subtypes of SS: (1) biphasic, (2) monophasic, and (3) poorly differentiated. The biphasic type represents 20% to 30% of lesions and has both mesenchymal spindle cell components and an obvious epithelial component usually forming glands. The monophasic type is the most common (50-60%) in which the spindle cell component predominates. The poorly differentiated SS are epithelioid in morphology and have high mitotic activity. This type has the poorest prognosis.[2,4,7]

In addition to the microscopic evaluation of these tumors, a recent immunohistochemical profile suggests that EMA, cytokeratin AE1/AE3 and E-cadherin, in combination with CD34 negativity, are the most useful and sensitive markers for diagnosing SS.[4,8] CD34 may rarely be positive and Bcl-2 and MIC-2(CD 99) usually positive in SS as was seen in our case.[9]

SS, on X-ray films appear as round or oval lobulated swellings or masses of moderate density. Although, half of the X-rays are normal, calcification is seen in up to 30% of the cases. Calcification is usually eccentric or peripheral within the soft tissue mass, represented by multiple small spotty radiopacities. Extensive calcification is associated with an improved prognosis.[2,4,10] SS appear as a heterogeneous soft tissue mass with attenuation, similar or slightly less than that of the muscle, as seen on computed tomography scans. Area of necrosis or hemorrhage presenting as lower density areas are common.[2] Magnetic resonance imaging is the optimal radiological modality for assessing the characteristics and the extent of SS. Intermixed areas of low, intermediate, and high signal intensities on long repetition time images have been marked as the triple sign apparently resulting in a mixture of solid cellular elements, hemorrhage or necrosis, and calcified or fibrotic regions with hemorrhage, fluid levels, and septa creating the bowl of grapes sign. This triple sign has been described to be occurring in 35% to 57% tumors. However, the triple sign is also seen in other soft tissue tumors; therefore, this finding alone lacks a high degree of specificity.[4,11] Angiographic studies of the SS frequently reveal a prominent vascularity of primary lesions and metastatic disease.[4]

The treatment of choice for SS is a wide local excision with negative surgical margins to minimize the chances of recurrence.[2,4,5] Radiotherapy is useful in local control of diseases, especially in gross residual tumor. Adjuvant chemotherapy mostly consists of regimens with combination of alkylating agents (cyclophosphamide or ifosphamide) and anthracyclines. Although chemotherapy is useful in disease control, their overall benefit in survival rate is controversial.[5] The 5-year survival rate for intermediate to high-grade sarcoma is 36% to 76%.[4] The clinical course of SS is characterized by a high rate of local recurrence (30-50%) and metastatic disease. The majority of metastases occur within the first 2 years to 5 years after treatment.[2,4]

Various adverse prognostic features in SS include metastasis, invasiveness, positive surgical margins, high histological grade, and poor differentiation. While, SYT-SSX2 gene fusion, monophasic SS, size less than 5 cm, less than 15 years of age, and distal extremity location have better prognostic outcomes; the significance of these findings is still debated.[2,4,5]

Our case presentation shows an atypical SS location within the abdominal wall. It should be considered as one of the differential diagnosis in anterior abdominal wall tumors.

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