

Primary synovial sarcoma of the abdominal wall: a case report and literature review

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Radiology Case. 2015 Jul; 9(7):47-52 :: DOI: 10.3941/jrcr.v9i7.1977

ABSTRACT

Synovial sarcoma (SS) is the fourth most common type of soft tissue sarcoma, following malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma. It usually occurs in the extremities near the large joints of middle-aged patients. We describe a case of synovial sarcoma of the anterior abdominal wall (SSAW) in an adolescent girl and undertake a review of the literature.

CASE REPORT

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A 12-year-old girl presented with a palpable painless mass at the left lower abdomen with a rapid increase in size over 1 month. The examination was negative except for an immobile firm mass of 10x15 cm arising from the left lower abdominal wall. Ultrasound (US) showed a heterogeneous hypoechoic mass with internal septation and honey combing appearance (Fig 1a). Increased vascularity in the solid parts of the mass was demonstrated on color Doppler ultrasound (Fig 1b). Computed Tomography (CT) scan of the abdomen revealed a heterogeneous enhancing mass of 5 x 9.8 x 9.8 cm originating from the left external oblique muscle (Fig 2a, 2b & 2c). Surgical removal discovered a well-defined round mass with hyper-vascularization and minimal invasion to the left external oblique muscle. The peritoneum and iliac bone were intact. The gross specimen, measuring about 13 x 12.5 x 6.5 cm, showed a grey white circumscribed rubbery mass with hemorrhage and necrosis at the muscular layer, and there were multiple mucinous cysts in the tumor (Fig 3). The histology was compatible with biphasic synovial sarcoma grade 3 and positive for vimentin, CD56+, epithelial membrane antigen

(EMA), and keratin; it was negative for desmin, smooth muscle actin (SMA), myogenin and mucin (Fig 4a & 4b). The patient received adjuvant chemotherapy and radiation therapy after surgical removal. Clinical and imaging follow-up (post-surgery to 9 months) revealed no evidence of local recurrence or metastasis.

DISCUSSION

Etiology & Demographics

Synovial sarcoma (SS) is an uncommon malignant mesenchymal tumor. The pathogenesis remains uncertain; however, it is believed to derive from primitive mesenchymal cells, which explains the extra-articular and diverse locations [1-5]. Although the common location for synovial sarcoma is close to large joints (not intra-articular), particularly in the knee (in the popliteal fossa), and has an association with joint capsules, tendon sheaths, bursae and fascial structures, rare unexpected sites can be found at the head and neck regions, including the pharynx, larynx and orbit. Retroperitoneum,

mediastinum, bones, nerves, and blood vessels, as well as visceral organs such as lungs, pleura, heart, prostate or kidneys, can also be involved. It is commonly found between 15 and 40 years; the incidence is 2.5 per 100,000 [1]. Only 47 cases of synovial sarcoma of the anterior abdominal wall (SSAW) have been reported in the English literature and only one of those cases was of an adolescent [1-8]. Although SSAW has a much greater frequency in females, SS in the extremities or head and neck regions tends to occur more frequently in males [6].

Clinical & Imaging findings

The tumors can present as slow growing palpable soft tissue masses, with or without pain, and are associated with the vague abdominal symptoms that occur more commonly in the lower abdomen. Metastases can present in 16-25% of cases (upon initial presentation), particularly to the lungs, and less commonly in lymph nodes and bones. The majority of metastases occur in the first 2-5 years of treatment [5]. Imaging findings of SSAW are not pathognomonic, although stipple calcifications are found in about 30%, and can be eccentric or peripheral. Necrotic and hemorrhagic areas are also commonly detected. The unusual but remarkable sonographic feature for SSAW, reported by Kishino et al, was a complex "honeycomb" echotexture that is distinct from previously reported SS at other sites [7]. Increased internal vascularity on Doppler imaging was also demonstrated in SSAW, which is similar to the SS arising from the extremities or head and neck regions. CT is more sensitive than both the plain radiograph and ultrasound for detecting calcifications. The most common CT appearance of SS is the heterogeneous soft tissue mass with attenuation equal or slightly less than that of muscle. The heterogeneous areas represent internal necrosis or hemorrhage. Magnetic Resonance Imaging (MRI) appears isointense to slightly hyperintense on T1-weighted images, and hyperintense to muscles on T2-weighted images. Marked heterogeneity and enhancement are highly suggestive of SS, likewise with the CT findings. Furthermore, MRI is better at delineating multi-lobulation on T2-weighted images. Cross sectional imaging is helpful for evaluating extensions, planning treatment, and monitoring responses to chemotherapy.

Treatment & Prognosis

A surgical wide excision is the treatment of choice with adjunctive radiation, chemotherapy or both; nevertheless, recurrence still ranges from 28%-36% [4]. A close follow-up of at least every 3 months for the first 2 years and then bi-annually for another 5 years is recommended due to high local recurrence [5]. The survival rate and prognosis are correlated to tumor size, histologic subtype, mitotic rate, percent glandularity, tumor necrosis, and vascular invasion. A tumor size larger > 5 cm with a presence of necrosis and a high mitotic rate are poor prognostic factors [9]. SSAW is staged using a modified tumor, node and metastases (TNM) classification, like other truncal sarcomas [5]. The three histologic subtypes are monophasic, biphasic, and the poorly differentiated type. The monophasic subtype predominantly consists of spindle cells resembling fibrosarcoma, while the biphasic subtype is composed of a bimorphic form, with mixed spindle and epithelioid cell components. The poorly differentiated subtype includes epithelioid cells with high

mitotic activity. Both monophasic and biphasic SS are usually of an intermediate grade (2/3). The poorly differentiated subtype is a high grade (3/3) [4]. Specific chromosome aberrations are the (X; 18) (p11.2;q11.2) translocation found in 90% of SS and the fusion of the SYT gene on chromosome 18 with either SSX1 (67% of cases) or SSX2 (33% of cases) on chromosome X [5]. All biphasic subtypes predominantly express SYT-SSX1 fusion transcription whereas monophasic tumors mostly carry SYT-SSX2 [3]. The most sensitive immune-histochemical staining for synovial sarcoma are positive EMA, cytokeratin AE1/AE3 and E-cadherin, in combination with negative CD 34[3].

Differential Diagnosis

Differential diagnosis of abdominal wall tumors in adolescents and young adults includes desmoid tumor (for benign tumors), mesenchymal chondrosarcoma, and rhabdomyosarcoma (for malignant tumors). Desmoid tumor is a well-defined lesion with variable echogenicity on ultrasonography. On CT scan or MRI, it appears as homogeneous or heterogeneous mass with a variable degree of contrast enhancement depending on its composition. Extraskelatal mesenchymal chondrosarcoma presents as a discrete lesion with ill-defined margin, heterogeneous hypoechogenicity compared with the surrounding soft tissue, and has scatter foci of calcifications and internal blood flow on ultrasonography. Multiple areas of fine and coarse calcifications (70%) and generalized enhancement can be found on CT scan. On MRI, this tumor appears isointense compared to muscle on T1-weighted images and hyperintense on T2-weighted images, reflecting cartilage or mesenchymal cellular component and inhomogeneous enhancement. Rhabdomyosarcoma has a slightly inhomogeneous hypoechoic appearance on ultrasonography. CT scan shows a circumscribed to infiltrative soft tissue mass and heterogeneous enhancement with or without necrosis or hemorrhage. Signal characteristics of rhabdomyosarcoma can present as mixed low and intermediate signal intense on T1-weighted images, intermediate to high signal intense on T2-weighted images, and strong heterogeneous enhancement. Furthermore, non-neoplastic lesions, such as a hematoma in the abdominal wall, should be considered in the differential diagnosis because the imaging characteristics of hematoma can mimic tumors. The hematoma has well-defined margins, variable and heterogeneous echogenicity, which represent a partial blood clot or organizing hematoma. On CT and MRI the attenuations and signal intensities vary depending on the stage of the hematoma. However, the hematoma has no significant contrast enhancement whereas the other tumors show homogeneous or heterogeneous enhancement. Repeat imaging study is helpful for excluding a true tumor because the hematoma evolves over time.

TEACHING POINT

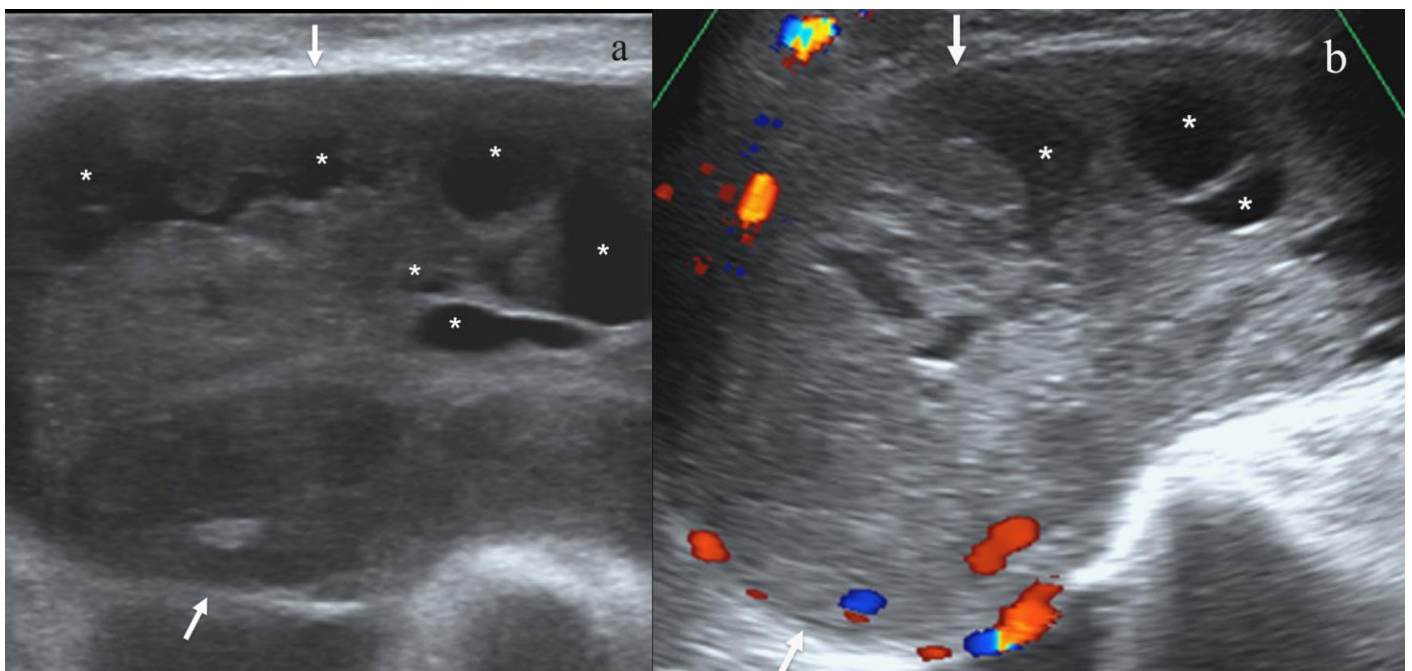
Primary synovial sarcoma is rarely found in the anterior abdominal wall. Nonetheless, it should be included in the differential diagnosis when an adolescent presents with stipple calcifications in an abdominal wall mass.

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FIGURES

Figure 1 (bottom): A 12-year-old girl with primary synovial sarcoma of the abdominal wall. (a) Gray-scale sonography (PLT-805AT, 12L5, 5-12MHz, linear transducer) shows a heterogeneous hypoechoic mass (arrows) with internal septation and a honey combing appearance (*). (b) Color Doppler ultrasound demonstrates increased peripheral vascularity in the solid parts.



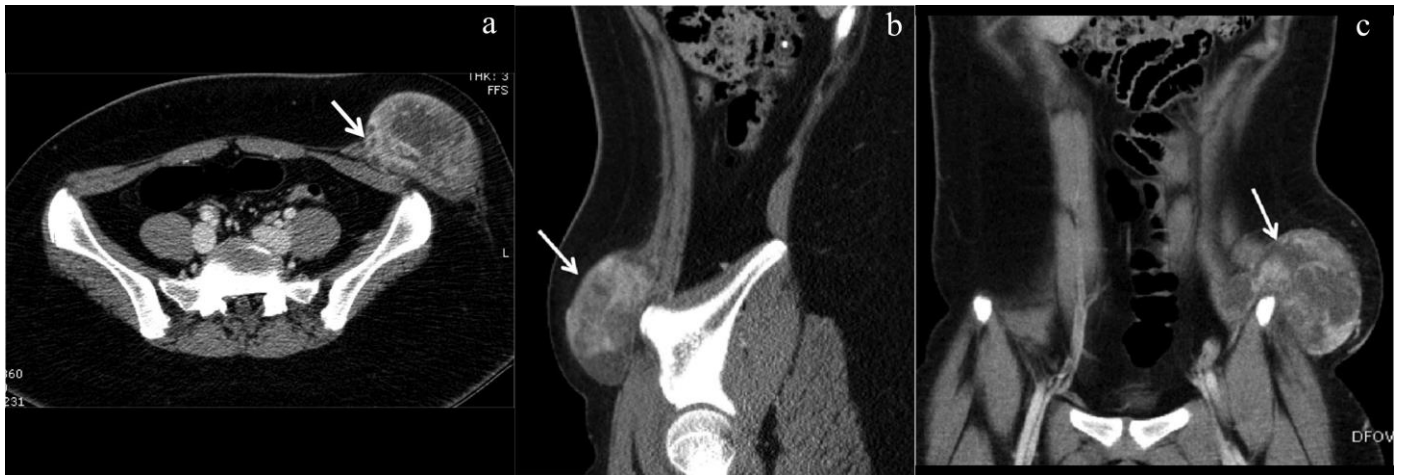


Figure 2: A 12-year-old girl with primary synovial sarcoma of the abdominal wall. (a) Enhanced axial venous phase of CT scan of the abdomen (Phillips Brilliance 64 slice, protocol: 180 mAs, 120 kVp, 3mm, 120 mL of Ultravist 370) reveals a heterogeneous enhancing mass about 5x9.8x9.8 cm originating from the left external oblique muscle (arrow). (b) Sagittal reformatted image of CT scan of the abdomen (Phillips Brilliance 64 slice, protocol: 180 mAs, 120 kVp, 3mm, 120 mL of Ultravist 370) reveals a heterogeneous enhancing mass about 5x9.8x9.8 cm originating from the left external oblique muscle (arrow). (c) Coronal reformatted image of CT scan of the abdomen (Phillips Brilliance 64 slice, protocol: 180 mAs, 120 kVp, 3mm, 120 mL of Ultravist 370) reveals a heterogeneous enhancing mass about 5x9.8x9.8 cm originating from the left external oblique muscle (arrow).



Figure 3: A 12-year-old girl with primary synovial sarcoma of the abdominal wall: The cut surface of the gross specimen measuring about 13x12.5x6.5 cm shows a grey white circumscribed rubbery mass with hemorrhage (arrow) and multiple mucinous cysts (*) in the tumor.

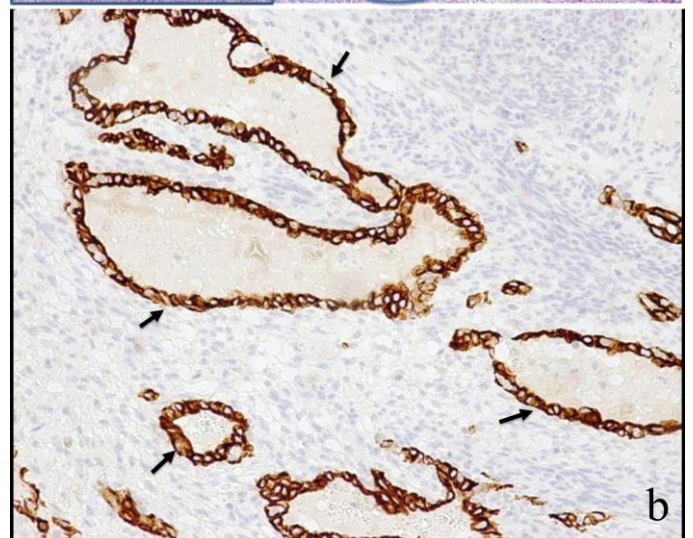
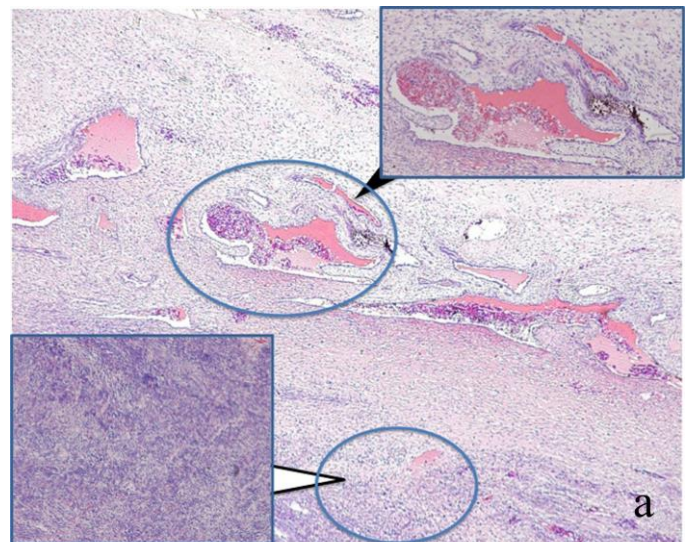


Figure 4 (right): A 12-year-old girl with primary synovial sarcoma of the abdominal wall. (a) This section reveals a biphasic synovial sarcoma with epithelial glandular and spindle cell component (hematoxylin and eosin staining, original magnification X100). Predominantly spindled tumor cells are uniform, ovoid nuclei arranged in fascicles and sheets (bottom left). The epithelial glandular component has ovoid nuclei, abundant cytoplasm, and form glandular structures (top right) (inserted panel, hematoxylin & eosin staining, magnification X150). (b) The glandular epithelial cells show positive staining for epithelial membrane antigen (EMA) (arrows) (original magnification X400).

Etiology	It is believed to derive from primitive mesenchymal cells.
Incidence	The incidence of synovial sarcoma is 2.5 per 100,000 [1]. Only 47 cases of synovial sarcoma (SS) of the anterior abdominal wall (SSAW) have been reported in the English literature [1-8].
Gender Ratio	SSAW has a much greater frequency in females while SS in the extremities or head and neck regions tends to occur more frequently in males [6].
Age Predilection	It is commonly found in adolescents and adults between 15 and 40 years of age.
Risk Factors	90% of cases have translocation of chromosome t(X; 18) (p11.2: q11.2) and fusion of SYT gene on chromosome 18 with either SSX1 or SSX2 on chromosome X [3, 5].
Treatment	Surgical wide excision is the treatment of choice followed by adjuvant chemotherapy and radiotherapy.
Prognosis	It is correlated to tumor size, histologic subtype, mitotic rate, percent glandularity, tumor necrosis and vascular invasion [3, 4, 5, 9].
Findings on Imaging	Radiograph shows a lobulated soft tissue density mass with underlying bone erosion; stipple calcifications can be found in 30% of cases. Remarkable sonographic feature is complex “honeycomb” appearance. On CT, the tumor appears as a heterogeneous soft tissue mass, representing necrosis and hemorrhage and its attenuation is equal or slightly less than that of muscle. On MRI, the mass usually presents as isointense to slightly hyperintense on T1- weighted images and hyperintense compared to muscle on T2-weighted images with markedly heterogeneous enhancement.

Table 1: Summary table for primary synovial sarcoma of the abdominal wall

Differential	Ultrasound	Computed Tomography	Magnetic Resonance Imaging
Synovial sarcoma of the abdominal wall (SSAW)	Well defined lesion with mixed heterogeneous echogenicity representing hemorrhage and necrosis. Increased internal vascularity on Doppler imaging. Reported unusual but remarkable feature of “Honeycomb” echotexture [7]	Heterogeneous soft tissue mass with attenuation equal or slightly less than that of muscle	Isointense to slightly hyperintense on T1-weighted images, hyperintense to muscles on T2-weighted image and marked heterogeneous enhancement
Desmoid tumor	Well defined lesion with variable echogenicity	Depending on its composition, homogeneous or heterogeneous hypodense-isodense or hyperdense compared to muscles, with a variable degree of contrast enhancement	Low signal intensity on T1-weighted images and heterogeneously hypointensity on T2-weighted images and variable contrast enhancement
Extraskelatal Mesenchymal Chondrosarcoma	Discrete lesion with ill-defined margin, heterogeneous hypo-echogenicity compared to surrounding soft tissue, scatter foci of calcifications and internal blood flow	A well-defined mass with multiple areas of fine and coarse calcifications (70%), generalized enhancement or peripheral enhancement possibly representing necrosis	Isointensity compared to muscle on T1- weighted images and hyperintensity on T2-weighted images reflecting cartilage or mesenchymal cellular component, and inhomogeneous enhancement
Rhabdomyosarcoma	A well-defined slightly hypoechoic inhomogeneous mass	A circumscribed to infiltrative soft tissue mass and heterogeneous enhancement +/- necrosis or hemorrhage	Mixed low and intermediate signal intensity on T1 weighted images, intermediate to high signal intensity on T2 weighted images, strong heterogeneous enhancement
Hematoma	Ovoid shaped lesion with variable echogenicity, internal heterogeneous hyper-echogenicity representing partially clotted blood or organizing hematoma	Acute hematoma appears hyper-attenuated in unenhanced images, and non- enhanced. Its density becomes lower and equal to serum (20-30H.U.) after 2-4 weeks.	The signal intensity varies according to degradation of RBC products. Acute hematoma appears high intense than muscle on T1-weighted images, and low intense on T2 weighted images. Marked hypointensity on T2-weighted images in evolving hematoma. No significant contrast enhancement.

Table 2: Differential table for US, CT and MRI findings of primary synovial sarcoma of the abdominal wall

ABBREVIATIONS

CT - Computed Tomography
EMA - Epithelial Membrane Antigen
MRI - Magnetic Resonance Imaging
SS - Synovial Sarcoma
SSAW - Synovial Sarcoma of the Abdominal Wall
US - Ultrasound

KEYWORDS

Synovial sarcoma; abdominal wall; adolescent; surgery; US;
CT; Computed Tomography

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