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Imaging features of primary and recurrent intrathoracic synovial sarcoma: a single-institute experience☆

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ABSTRACT

Objective: The objective was to study the imaging features of primary and metastatic intrathoracic synovial sarcoma (ISS).
Materials and methods: We reviewed the imaging of 42 patients with ISS (31 pleural, 7 intrapulmonary, 4 mediastinal), with baseline imaging of 19 patients and follow-up imaging in all.
Results: Primary tumors ($n=19$) were well circumscribed (15/19), homogeneous or heterogeneously enhancing, with mean size 9.1 cm (range: 1.8–25 cm). Recurrent/metastatic disease developed in 28/42 patients (67%). Lung was the most common site of metastases (21/28), with most of them predominantly pleural (11/21). On pathology, 30 patients had monophasic tumors, and 6 each had biphasic tumors and poorly differentiated tumors.
Conclusion: Intrathoracic SS most commonly presents as a large heterogeneous pleural mass without associated adenopathy. Lung metastases are often pleural based and ipsilateral.

1. Introduction

Synovial sarcoma (SS) is a relatively common high-grade sarcoma, accounting for 5%–10% of all soft tissue sarcomas [1,2]. Its name is a misnomer; and it is believed to arise from primitive mesenchymal cells which undergo epithelial (and not synovial) differentiation. Histologically, it consists of three subtypes: monophasic (most common subtype; consisting of spindle cells), biphasic (consisting of spindle and epithelial cells), and poorly differentiated, with poorly differentiated SS having the worst prognosis. SS is characterized by the presence of the t(X;18)(p11;q11) translocation involving SS18 (SYT) and SSX1, SSX2 or SSX4 in about 95% of tumors [1,3–5].

SS most commonly affects the extremities (in about 80% of cases) but may affect almost any region or organ [6,7]. Intrathoracic SS has poor prognosis as compared to extremity SS, with a high rate of recurrent disease and worse survival [5,8].

Certain typical magnetic resonance imaging (MRI) signs of soft tissue SS have been well described in literature. Tumor may demonstrate T2 hyperintense, intermediate and hypointense components (triple sign) due to cystic, necrotic, or hemorrhagic elements (hyperintense), cellular areas (intermediate), and calcified or fibrotic regions or hemosiderin staining (hypointense). The presence of sedimented blood products may give rise to fluid–fluid levels. A 'bowl-of-grapes' appearance may be visualized in the form of T2 hyperintense areas with intervening T2 hypointense septa [9–12]. A few small radiology series (up to 12 cases) have previously described the imaging features of primary intrathoracic synovial sarcoma [13–15]. However, they described the imaging findings of the primary tumor without focusing on the imaging of recurrent disease. We present the cross-sectional imaging features of 42 patients of primary and recurrent intrathoracic synovial sarcoma with clinicopathological correlation.

2. Materials and methods

2.1. Subjects

This was a Health Insurance Portability and Accountability Act-compliant retrospective study approved by the Institutional Review Board with waiver of informed consent. Between 2002 and 2013, 374 consecutive patients with histopathologically confirmed SS involving the entire body were identified from the pathology database of our institute. Of these, 157 patients had imaging studies of the primary and/or recurrent tumor available for review. Forty-two of these patients had primary intrathoracic SS and were included in the present study.

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2.2. Clinical and histopathologic data

Detailed clinical and histopathologic information was extracted from the electronic medical records for all the 42 patients, including demographic data, date of diagnosis of primary tumor, clinical features at presentation, date of diagnosis of metastases, management of the primary tumor and metastases, duration of follow-up, and final outcome (alive, dead, or lost to follow-up). The histopathology of all the cases was reviewed at the pathology department of our institute. Molecular testing for the presence or absence of t(X;18) was performed in all cases.

2.3. Imaging and image analysis

Baseline imaging of the treatment-naive primary tumor was available in 19 patients [computed tomography (CT) in 17, MRI in 6, and positron emission tomography (PET)/CT in 7], and follow-up imaging was available in all 42 patients. A systematic review of all pretreatment and follow-up imaging studies was performed in consensus by two oncoradiology fellowship-trained radiologists (A.D.B and S.H.T) with 5 and 8 years of experience. A total of 278 CTs of the chest, abdomen, and pelvis; 55 thoracic MRIs; and 36 whole-body PET/CTs were reviewed (mean, 4; range, 2–39).

Contrast-enhanced multidetector CT (MDCT) images were acquired on 4-slice (GE Health Care, Barrington, IL, USA), 16-slice (Siemens Medical Solutions, Forchheim, Germany), and 64-slice MDCT (Aquilion 64; Toshiba America Medical Systems, Tustin, CA, USA) scanners with 5-mm-thickness axial images and 4-mm coronal reconstructions. Iopromide (300 mg I/ml; Ultravist 300; Bayer HealthCare Pharmaceuticals, San Francisco, CA, USA) was administered as intravenous contrast using an automated injector (Stellant; Medrad, Warrendale, PA, USA) at a rate of 2–3 ml/s with a scan delay of 60 s. The MRI examinations were performed on a 1.5-T MRI (GE Healthcare, Milwaukee, WI, USA) or 3-T MRI (Siemens Medical Solutions, Forchheim, Germany) scanners with gadolinium administration at doses of 0.1 mmol/kg body weight up to a maximum dose of 20 ml. Sequences acquired included axial T2-weighted [repetition time (TR)/echo time (TE), 1869/78]; coronal T2-weighted fat-suppressed (TR/TE, 2020/95); in- and out-of-phase T1-weighted, diffusion-weighted sequences (*b*: 0, 500, and 800), and nonenhanced and dynamic contrast-enhanced fat-suppressed T1-weighted gradient-recalled echo (TR/TE, 5.4/2.3). The images were reviewed on Centricity PACS RA1000 (GE Health Care, Barrington, IL, USA) workstation. PET was performed from the base of the skull through the thighs (Discovery LS; GE Healthcare, Waukesha, WI, USA; or Biograph 16-HiRes, Siemens), with noncontrast helical CT imaging (5-mm-thickness axial images) performed over the same range without breath-hold for attenuation correction of PET images and anatomic correlation. The PET/CT images were reviewed on HERMES GOLD (Hermes Medical Solutions Inc., Greenville, NC, USA) workstation.

Imaging features of the primary tumor that were recorded included the site of origin (intrapulmonary, pleural, or mediastinal), largest dimension, margin (well defined or ill defined), outline (smooth, lobulated, or irregular), tumor heterogeneity, enhancement (compared to skeletal muscle), fluorodeoxyglucose uptake (where applicable), and presence of calcification or hemorrhage. Pleural-based and pericardial-based tumors were considered in the pleural group. The presence of metastases at presentation and of development of local recurrence or metastases during follow-up, their site, number (whether single or multiple), and imaging features were also noted. In the case of patients with resected pleural SS, lesions were considered locally recurrent if they recurred in the surgical bed and were considered metastatic if they recurred at other sites. Metastatic lesions were confirmed either at histopathology (8 patients) or by the presence of unequivocal progression or response to treatment on serial follow-up imaging (20 patients).

3. Results

The study consisted of 16 men and 26 women with a mean age of 45 years (range, 15–81 years). Thirty-one patients had pleural SS, 7 had pulmonary SS, and 4 had mediastinal tumors. The results are tabulated in Table 1.

3.1. Clinical presentation

Majority of the patients were symptomatic at presentation (33/42, 79%), with the most common presentations being chest pain in 20/42 (48%), shortness of breath in 8/42 (19%), cough in 5/42 (12%, including 2 patients with hemoptysis), and a painless lump in 4/42 (9%). Two patients presented with a spontaneous pneumothorax and two with spontaneous hemothorax, while one patient presented with pneumonia. The remaining 9/42 (21%) patients were asymptomatic and detected incidentally on imaging for other causes.

3.2. Imaging of the primary tumor ($n=19$)

Baseline imaging of the primary was available in 19 patients, while the rest had imaging of treated/recurrent disease. Thirteen out of 19 patients had pleural tumors, 4/19 had intrapulmonary tumors, while 2/19 had mediastinal tumors. The imaging features are tabulated in Table 2. The mean size of the primary tumor on imaging was 9.1 cm (range, 1.8–25 cm). Margins were well circumscribed in 15/19 (79%) patients and ill defined and infiltrative in 4/19 (21%) patients, with the ill-defined tumors having associated areas of lung atelectasis/consolidation or pleural fluid masking the tumor margins. Five patients had ipsilateral effusion at presentation.

On CT ($n=17$), 3/17 tumors showed homogeneous solid enhancement compared to skeletal muscles (2 intrapulmonary tumors and a mediastinal tumor), and 2 had diffuse pleural involvement without a focal mass seen on CT. The remaining 12 lesions demonstrated heterogeneous enhancement, with 9 having cystic/necrotic components. Calcifications were present in two tumors.

On MRI ($n=6$), tumors were hyperintense on T2-weighted images relative to the chest wall musculature and demonstrated low to intermediate signal intensity on T1-weighted images with T1 hyperintense hemorrhagic foci in three patients. Hemorrhage was seen in three of six patients; fluid–fluid levels and 'bowl of grapes' appearance were each present in two of six tumors; and the triple sign consisting of T2 hypo, intermediate, and hyperintense foci was seen in one tumor.

PET/CT ($n=7$) revealed similar imaging features on CT, with the tumor being non-FDG avid in two patients, mildly avid in two other (SUVmax 2 and 4), and strongly avid in the remaining three patients (mean SUVmax, 8.2).

Two patients had mediastinal tumors, one having a homogeneously enhancing superior mediastinal mass and the other having

Table 1
Summary of clinicopathological findings

Characteristic	Findings ($n=42$)
Male vs. female	16 vs. 26
Mean age	45 years (range, 15–81 years)
Symptomatic at presentation	33 (79%)
Tumor site: pleural based vs. intrapulmonary vs. mediastinal	31 vs. 7 vs. 4
Metastases at presentation	8 (19%)
Overall metastases/recurrence	28 (67%)
Monophasic vs. biphasic vs. poorly differentiated on pathology	30 vs. 6 vs. 6
Median follow-up	30 months (range, 2–158 months)
Overall median survival	35 months (range, 5–158 months)
Median survival for patients with metastases at presentation	20 months (range, 5–50 months)