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Radiologic Findings in Primary Thoracic Synovial Sarcoma Differ from Soft Tissue Synovial Sarcoma

Paul H Hartel[1,2] and John E Parker[2]*

[1]Consultant Histopathologist, Sligo University Hospital, Ireland

[2]Department of Medicine, West Virginia University, School of Medicine, USA

Abstract

Radiologic imaging is an important tool in diagnosis, staging and management of synovial sarcoma. Soft tissue synovial sarcoma has some characteristic findings on imaging studies, i.e., high vascularity, tumour calcification and intermuscular growth. We compared known imaging characteristics of soft tissue synovial sarcoma with those of a set of primary thoracic synovial sarcomas to add to the literature of this rare sarcoma. 61 cases diagnosed as primary thoracic synovial sarcoma, from 1981 to 2020, were retrieved from tissue archives. Chest radiographs, computed tomography, magnetic resonance imaging, or positron emission tomography/computed tomography studies were available and reviewed in 14 cases. Radiologically, compared with soft tissue synovial sarcoma, primary thoracic synovial sarcoma shows less vascularity in general, no cortical bone destruction, no tumour calcification or HPC-like vasculature and no invasion into chest wall or intermuscular growth. Additionally, in contrast to soft tissue synovial sarcoma, lymphadenopathy suggestive of metastasis was not seen. Lack of PET-avidity of suspicious lesions should not discourage close clinical follow-up. Awareness of the lack of classic soft tissue synovial sarcoma imaging findings will aid radiologic diagnosis of primary thoracic synovial sarcoma.

Introduction

Synovial sarcoma is a rare and aggressive neoplasm conventionally involving knee and ankle of young adults and children [1]. However, it is now known to affect older adults and involve numerous sites including soft tissue around other joints, retropharynx, oral cavity, salivary glands, abdomen, retroperitoneum, blood vessels, GI tract, kidneys, prostate, vulva and lung and mediastinum [2]. Grossly it is a well-circumscribed firm tan-pink-grey lesion with dense spindle cells microscopically. Well-formed epithelial components can be seen with biphasic tumors as can poorly differentiated tumors showing round, or rarely, rhabdoid cells. Stromal hyalinization, hemangiopericytoma-like vasculature and focal myxoid areas are also common. Tumors are typically immunoreactive with bcl-2 and CD99 as well as at least focally with at least one epithelial marker [3]. The t(x;18) translocation and SS 18/SSX fusion products are pathognomonic and present in greater than 90% of cases. Synovial sarcomas often recur locally and metastasize to lymph nodes, a usual feature of other soft tissue sarcomas, in 10% to 15% of cases [2]. Preferred treatment is surgical excision with clear margins and adjunctive high dose radiation therapy. Five-year survival is around 50%, although younger patients, and patients with distal tumour sites, smaller tumors (<5 cm) and heavily calcified lesions do better.

Radiologic imaging is an important tool in diagnosis, staging and management of synovial sarcoma. Soft tissue synovial sarcoma has some characteristic findings on imaging studies, i.e., high vascularity, tumour calcification and intermuscular growth [4]. We compared known imaging characteristics of soft tissue synovial sarcoma with those of a set of primary thoracic synovial sarcomas to add to the literature of this rare sarcoma.

Materials and Methods

Sixty-one cases diagnosed as primary thoracic synovial sarcoma, from 1981 to 2020, were retrieved from tissue archives. Hematoxylin and eosin-stained sections were reviewed. Tumors were sub typed as monophasic or biphasic according to World Health Organization criteria [3]. Grading by tumor cell differentiation, mitotic rate, and necrosis was performed following the French Federation of Cancer Centers (FNCLCC) scheme. Cases that failed to meet the World Health Organization histologic and/or immunohistochemical criteria for synovial sarcoma were excluded.

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Correspondence:
Paul H Hartel, Department of Medicine, West Virginia University, School of Medicine, USA,
E-mail: phartel16@gmail.com

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Table 1: Radiologic imaging characteristics of thoracic synovial sarcoma.

Imaging modality and findings	Chest x-ray	CT Scan	MRI	PET
Well-circumscribed	8 (80%)	9 (75%)	3 (100%)	-
Mediastinal shift	1 (10%)	0	0	-
Cavitation	0	0	0	-
Calcification	0	0	0	-
Lymphadenopathy	0	0	0	0
Pleural based	-	9 (75%)	-	-
Pleural effusion	0	8 (67%)	-	-
Enhanced vessels	-	1 (8%)	-	-
HPC-like vessels	-	0		
Chest wall invasion	-	0	0	-
Bone destruction	-	0	0	-
Triple signal	-	-	2 (67%)	-
Peripheral rim of enhancement	-	-	3 (100%)	-
Uptake of fludeoxyglucose F18	-	-	-	1 (50%)
Total cases	10	12	3	2

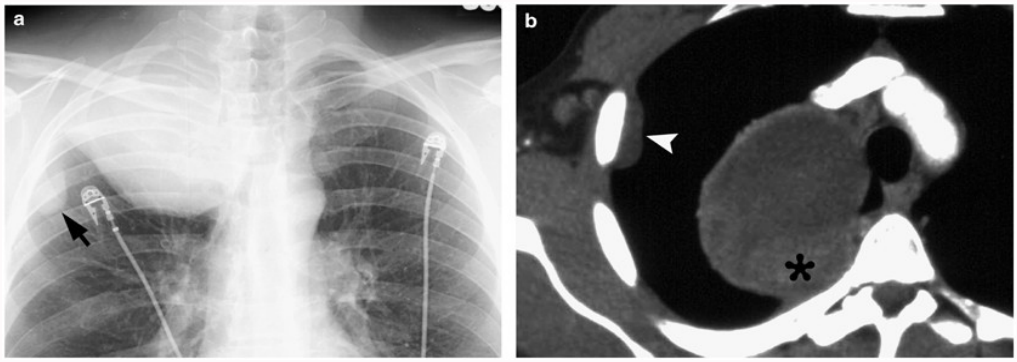

Figure 1: Synovial sarcoma in a young man with shortness of breath. Chest radiograph (a) demonstrates right apical mass. A pleural-based nodule (arrow) lies inferiorly. Contrast-enhanced CT scan (b) demonstrates a cystic mass with thickened wall (asterisk) and a metastatic nodule (arrowhead) along lateral pleuron.

Chest radiographs, computed tomography, magnetic resonance imaging, or positron emission tomography/computed tomography studies were available and reviewed in 14 cases. These included 10 chest radiographs, 12 computed tomography studies, three magnetic resonance imaging studies, and two cases with fludeoxyglucose F18 positron emission tomography/computed tomography images. Follow-up data were obtained from patient records.

Results and Discussion

Patients were 8 females and 6 males ranging in age from 17 to 68 years (m=38). The most common symptoms were shortness of breath and chest pain. Tumors were in lung (10) and pleura (4) and ranged from 5 cm to 16 cm in size (m=10). Tumors were diagnosed by a pulmonary pathologist using World Health Organization criteria and immunohistochemistry and graded using the FNCLCC grading scheme for sarcomas. Tumors were monophasic (11) and biphasic (3) and eight tumors were grade 2 and 6 were grade 3 (poorly differentiated). No additional tissue was available for t(x;18) testing. Tumors were treated with surgical excision including pneumonectomy (3), lobectomy (8) and excision of tumour mass (3). Known adjuvant treatment included chemotherapy in two patients and radiation therapy in one patient. Patients had no evidence of metastases at time of diagnosis. Clinical follow-up ranged from 12 to 21 months (m=17) and patients at that time were dead of disease (3), alive with disease (3) or had no evidence of disease (5). Three patients were lost to follow-up.

Table 1 summarizes radiologic findings across modalities. Tumors were well-circumscribed with sharp borders and homogeneous with rare contralateral mediastinal shift on chest radiographs. On computed tomography, tumors showed well-defined homogeneous or heterogeneous enhancement and appeared pleural based with ipsilateral pleural effusion (Figure 1). Enhanced tumor vessels were rare. Calcification, bone destruction or chest wall invasion was not identified. No cavitation, calcification, or lymphadenopathy was seen. Magnetic resonance T1-weighted and T2-weighted images showed well-circumscribed masses without satellite nodules. Intermediate signal intensity nodules and high signal intensity cystic areas were consistent with myxoid areas, hemorrhage and necrosis (Figure 2). Magnetic resonance imaging also revealed more striking tumor heterogeneity than computed tomography. A peripheral rim of enhancement was demonstrated on magnetic resonance imaging following administration of gadolinium DTPA. Positron emission tomography/computed tomography studies showed increased uptake of fludeoxyglucose F18 by thoracic synovial sarcoma in one case, but was non-avid in a second case.

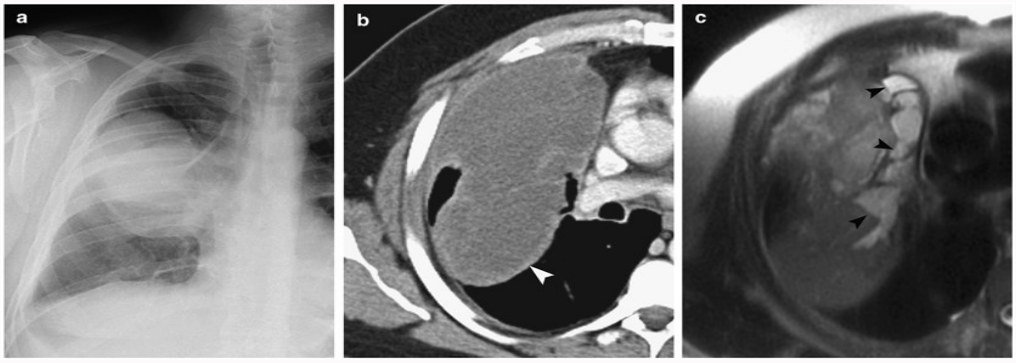

Figure 2: Synovial sarcoma in a young man with chest pain. Chest radiograph (a) of right lung demonstrates a well-circumscribed right hilar mass. Contrast-enhanced CT scan (b) demonstrates a large mass with low attenuation and rim enhancement (arrowhead). Axial T2-weighted MRI (c) shows contrast among internal components (triple-signal) with cysts (arrowheads).

In common with soft tissue synovial sarcoma, primary thoracic synovial sarcoma radiologically shows a similar "triple-signal" (bright, grey and dark) representing tumor, necrosis and hemorrhage [5]. Primary thoracic synovial sarcoma is also typically well-circumscribed with round or lobulated borders similar to soft tissue sarcoma [6-8]. Mediastinal shift is seen in patients with large thoracic tumors [9]. Computed tomography, in both soft tissue and thoracic synovial sarcomas, demonstrates homogeneous or heterogeneous enhancement with necrosis (Figure 1). Ipsilateral pleural effusion is common in thoracic tumors, while regional lymphadenopathy is rare [6-10]. Sarcomas in general arising in chest wall soft tissue, in contrast to thoracic synovial sarcomas, closely match imaging qualities of soft tissue synovial sarcoma including cortical bone destruction, tumour calcification and chest wall musculature invasion [6]. Soft tissue synovial sarcoma tends to be intermuscular and have hemangiopericytoma-like vascular pattern in around 30%, only rarely observed on chest imaging in our thoracic tumors [11,12]. One case was PET-negative and followed with serial chest radiographs but presented shortly thereafter with metastatic disease that was inoperable.

Conclusion

Radiologically, compared with soft tissue synovial sarcoma, primary thoracic synovial sarcoma shows less vascularity in general, no cortical bone destruction, no tumour calcification or HPC-like vasculature and no invasion into chest wall or intermuscular growth. Additionally, in contrast to soft tissue synovial sarcoma, lymphadenopathy suggestive of metastasis was not seen. Lack of PET-avidity of suspicious lesions should not discourage close clinical follow-up. Awareness of the lack of classic soft tissue synovial sarcoma imaging findings will aid radiologic diagnosis of primary thoracic synovial sarcoma.

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