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Case Report
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Findings of Alveolar Soft Part Sarcoma in Comparison with Other Soft Tissue Sarcomas

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Keywords

Sarcoma; Alveolar Soft Part Sarcoma; MRI

Case Presentation

A 32 year old male with past medical history of hypertension and sleep apnea and maternal family history of breast cancer presents to a local emergency department complaining of shortness of breath. Symptoms improved with breathing treatments and evaluation for deep venous thrombosis and pulmonary embolus was initiated. Lower extremity Doppler ultrasound (not shown) revealed an anterior right thigh mass. Upon further questioning, he did affirm several months of "nagging" thigh pain and tightness. He was admitted for work-up of the thigh mass. Contrast-enhanced MRI was obtained raising concern for sarcoma (Figure 1A-1C). Chest CT was performed demonstrating numerous lung nodules concerning for metastatic disease (Figure 1D).

Open biopsy of thigh mass and percutaneous biopsy of a pulmonary nodule confirmed sarcoma, not otherwise specified. Outside pathology report from biopsy of thigh mass describes "a poorly differentiated neoplasm composed of predominantly plump cells" with "prominent nucleoli" and a "vast arborizing intratumoral vascular pattern." Immunohistochemical staining demonstrated vimentin positivity; all others markers including but not limited to cytokeratins, smooth muscle actin, desmin, S-100, melan-A, and CD34 were negative. Rare mitoses (1 per 10 high power fields) were noted. He was referred to our institution for further histopathologic differentiation and treatment of sarcoma. Additional testing at our institution demonstrated positive staining PAS and TFE3 (transcription factor 3) confirming a diagnosis of alveolar soft part sarcoma.

Discussion

Soft tissue sarcomas are rare malignant mesenchymal neoplasms comprised of numerous histological types as defined by the World Health Organization [1]. Of these, alveolar soft part sarcoma (ASPS) represents less than 1% of soft tissue sarcomas [2]. While immunohistochemical analysis helps confirm definitive diagnosis, certain clinical and imaging findings shown in this case may allow for accurate prediction of histopathology at preoperative evaluation.

ASPS is typically an indolent, but malignant neoplasm with slow growth and low mitotic rate, though metastatic disease is common, most often to the lungs [3]. High rates of distant disease are thought to be secondary to marked tumoral vascularity with hematogenous spread of disease. ASPS most commonly affects young adults with a slight female predominance. Treatment consists of margin-negative surgical resection of the primary disease and systemic chemotherapy for distant metastases. Despite indolent course, the prognosis is generally poor due to common distant dissemination of disease [3].

The most common sites of disease include the thigh and buttocks and the vast majority are deep seated tumors [2]. Several typical imaging features seen at MRI suggest the diagnosis and help differentiate ASPS from other soft tissue sarcomas [4]. While most sarcomas appear isointense or hypointense to skeletal muscle on T1-weighted imaging, ASPS usually appears slightly hyperintense to muscle as shown in Figure 1A. Differential considerations for a T1-hyperintense mass include lipomatous tumors such as liposarcoma, hemorrhagic masses such as synovial sarcoma, and melanotic metastases. Numerous internal and peripheral flow voids due to neovascularity as seen in Figure 1B are often identified, also present in vascular tumors such as hemangiopericytoma and solitary fibrous tumor. ASPS typically demonstrates moderate to intense intravenous contrast enhancement related to increased vascularity with variable presence of internal necrosis as shown in Figure 1C. Most often, pulmonary metastases appear as multiple randomly distributed solid

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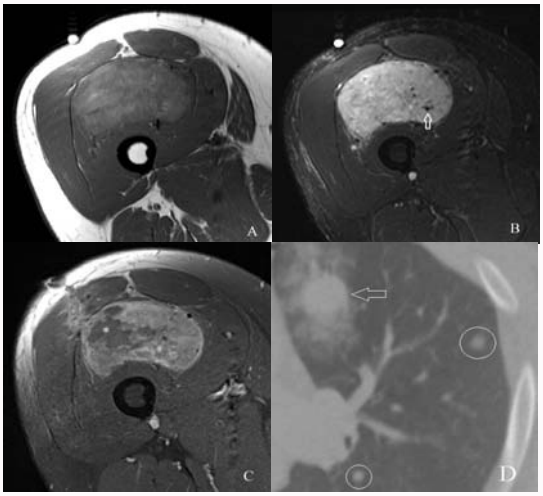

Figure 1: 32 year old with a right thigh mass and multiple pulmonary nodules. **A – C:** Axial (A) T1-weighted TSE, (B) fat-suppressed T2-weighted TSE, and (C) gadolinium-enhanced fat-suppressed T1-weighted magnetic resonance imaging of the right thigh demonstrates an anterior compartmental soft tissue mass with increased T1-weighted signal relative to skeletal muscle, hyperintense T2-weighted signal with several peripheral and internal flow voids (arrow in B), and moderate intravenous contrast enhancement with an area of necrosis (* in C).
D: Axial CT image of the left lung demonstrates a dominant solid lung nodule (arrow) with surrounding ground glass opacities and other small rounded solid pulmonary nodules (circles).

rounded lung nodules, though larger lesions not uncommonly present with surrounding ground glass opacities as shown in Figure 1D due to perilesional alveolar hemorrhage as often seen in other highly vascularized tumors such as angiosarcoma.

At histopathology, ASPS demonstrates notable consistency within the same tumor and across different patients, a distinct difference when compared with most soft tissue sarcomas which tend to demonstrate marked internal heterogeneity and often several distinct subtypes of disease [2]. ASPS consist of nests of epitheloid like cells lined by thin fibrous septae with abundant vascular channels. Tumor cells are large and rounded with granular eosinophilic cytoplasm. Nests of cells are typically detached resulting in a pseudoalveolar pattern and mitoses are typically rare. Immunohistochemical evaluation is most often negative for epithelial, neuroendocrine, and melanoma markers, while demonstrating variable positivity to vimentin, smooth and skeletal muscle markers [5]. A more recent discovery of a specific chromosomal abnormality involving the ASPL gene and the TFE3 gene has proven highly sensitive and specific for ASPS [5].

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