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Case Report

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A Case of Primary Synovial Sarcoma of the Posterior Mediastinum with Aortic Wall Infiltration with Complete Imaging Pathway and Histological Diagnosis

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Abstract

Primary synovial mediastinal sarcomas are aggressive tumors with a very rare incidence. The few cases reported in the literature presented poor and incomplete imaging. In this report, we describe the case of a 72-year-old male patient hospitalized for dyspnoea and chest tightness. For these reasons, he underwent chest X-ray, contrast-enhanced CT, MRI, and PET, which revealed a large mass around the aortic walls. The patient underwent a CT-guided biopsy and the subsequent histopathological evaluation indicated features consistent with a soft tissue sarcoma and positive staining for vimentin. Infiltration of aorta walls was confirmed by an MRI study with cine-MRI sequences. The patient was treated with neoadjuvant chemotherapy, showing partial response. Unfortunately, during the following attempted surgical removal of the mediastinal mass, the patient deceased as a result of an aorta rupture. Our experience could help in diagnostic confidence and differential diagnosis, especially with benign tumors such as neurogenic tumors.

Keywords

Synovial sarcoma, Posterior mediastinum mass, Imaging, Mesenchymal sarcoma, Mesenchymal neoplasm

Abbreviations

AP: Antero-Posterior; **CE-CT:** Contrast-Enhanced Computed Tomography; **CE-MRI:** Contrast-Enhanced Magnetic Resonance Imaging; **CINE-MRI:** Cinematic Magnetic Resonance Imaging; **CT:** Computed Tomography; **GE:** Gradient Echo; **LL:** Later-Lateral; **NO-CE-MRI**: Non-Contrast-Enhancement Magnetic Resonance Imaging; **PET-CT:** Positron Emission Tomography – Computed Tomography; **RT:** Radiotherapy **SS:** Synovial Sarcoma; **STS:** Soft Tissue Sarcoma; **SUV:** Standardized Uptake Value; **TSE:** Turbo Spin Echo; **X-ray:** radiograph.

Introduction

Synovial sarcoma (SS) is the fourth most common soft tissue sarcoma (STS) (5-10%) [1]. Even though nowadays it is common knowledge that it can arise in any district of the body, the past observation of its extremity's location granted it the "synovial" definition [2, 3]. The incidence peak of SS is in children and young adults, mainly males (1.2/1), although it can occur at any age [2, 3]. The origin of the SS could be from the multipotent, neural, and myogenic mesenchymal

cells, so the histological picture appears very different in the various cases [4-7]. The clinical course of SS is largely unclear. SS is cytogenetically characterized by a t(X; 18)(p11; q11) translocation, which leads to the fusion of the SS18 Subunit Of the BAF Chromatin Remodeling Complex (*SYT*) gene [8]. Thoracic cases represent less than 5% of SS cases; less than 10% of thoracic cases are primary mediastinal SS cases [1, 2, 9]. Mediastinal SS has a worse prognosis than soft SS tissue, lung, and pleura [10]. About half of the patients die within 5 years, and even after surgery, local recurrence has been reported in over 80% of patients [10]. Therefore, early diagnosis and multimodal treatment are essential. Slow tumor growth and small size may lead to the misdiagnosis of benign tumors, especially neurogenic tumors, so it is important to consider SS in the differential diagnosis of a primary intrathoracic tumor [10]. However, only a few cases with incomplete imaging have been reported in the literature. Here, we report a case with complete imaging to increase diagnosis confidence and help in differential with benign tumors, especially neurogenic tumors.

Case Report

A 72-year-old man presenting dyspnoea, jugular turgor, and chest tightness underwent a chest X-ray and CT scan which showed a mass located in the posterior mediastinal region (Figures 1 and 2). Following these findings, a contrast-enhanced CT, a PET, and an MRI were performed confirming the presence of a vascularized posterior mediastinal huge mass with high metabolism without secondary lesions (Figures 2, 3, and 4). MRI was also performed with cardio-synchronized cine-MRI scans which confirmed infiltration of the aortic wall (Figure 3). The patient then underwent fine-needle aspiration that showed fragments of fibroconnective tissue infiltration of atypical, pleomorphic, voluminous cellular elements, immersed in a loose, oedematous, richly vascularized stroma, with large necrotic areas and widespread necrobiotic phenomena. Sparse mitosis was noted. Immunohistochemical characterization showed Vimentin+, CD34-, S100-, LCA-, TTF1-, P63-, EMA-, CKMNF116- with a growth fraction/Mib-1 equal to 20%. These findings were considered compatible with high-grade not otherwise specified (NOS) synovial neoplasia. The patient was treated with three cycles of neoadjuvant chemotherapy, with partial response. Considering the huge mass and its mass effect, the patient underwent chest surgery to resect part of the pathological tissue. Unfortunately, during the surgery, due to a rupture of the aortic wall with consequent hypovolemic shock, the patient deceased.

Discussion

SS owes its name to the initial hypothesis that the tumor originates from synovial cells, after a frequent association with joints, tendons, and bursal structures [10]. Cases not associated with these structures and the identification of variable epithelial differentiation led researchers to believe that synovial sarcomas are probably derived from pluripotent mesenchymal cells capable of divergent differentiation [10]. Usually, SS arises from the deep soft tissue of the extremities, accounting for 5-10% of all soft tissue sarcomas [10]. However synovial sarcomas have been found in most anatomical sites, including

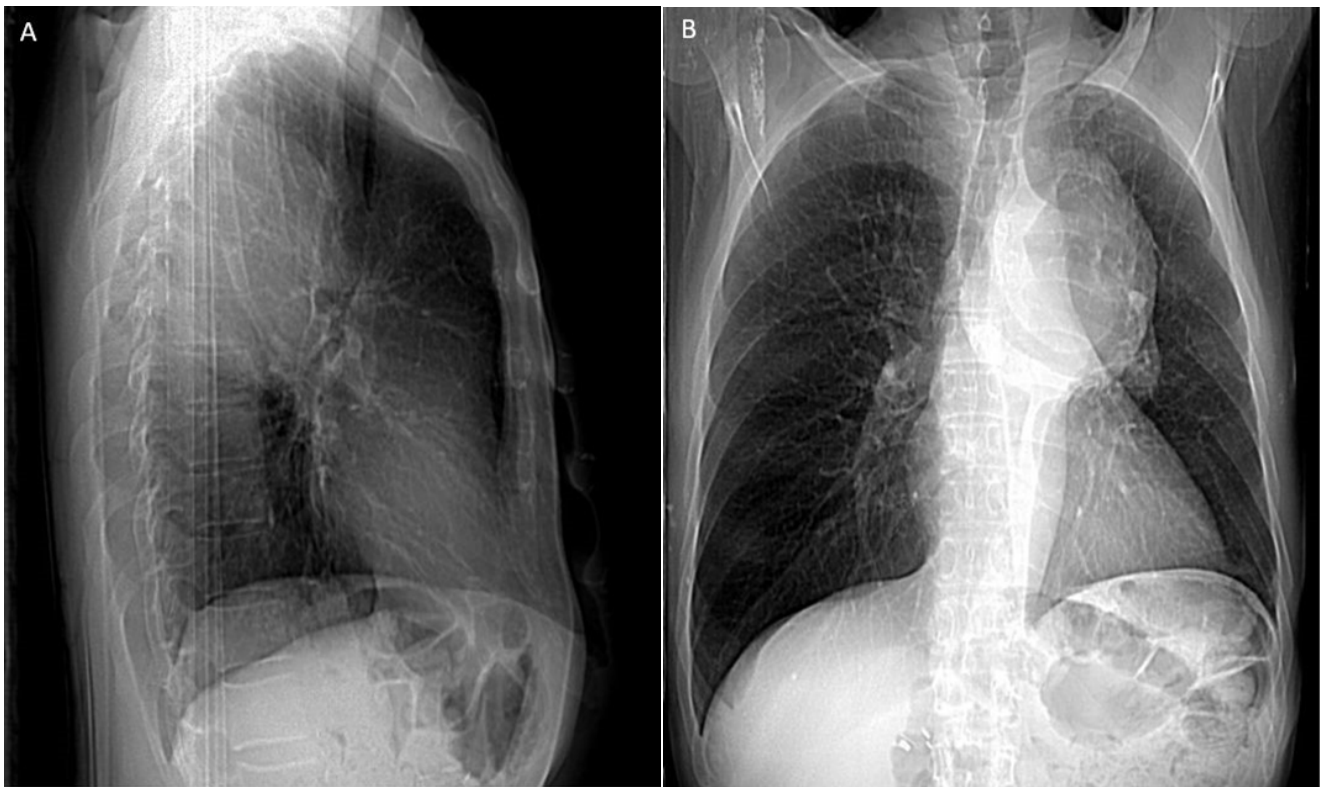


Figure 1: AP(b) e LL(a) view, showing a left posterior para-mediastinal/para-aortic round mass.
AP: Antero-Posterior; CT: Computed Tomography; LL: Later-Lateral.

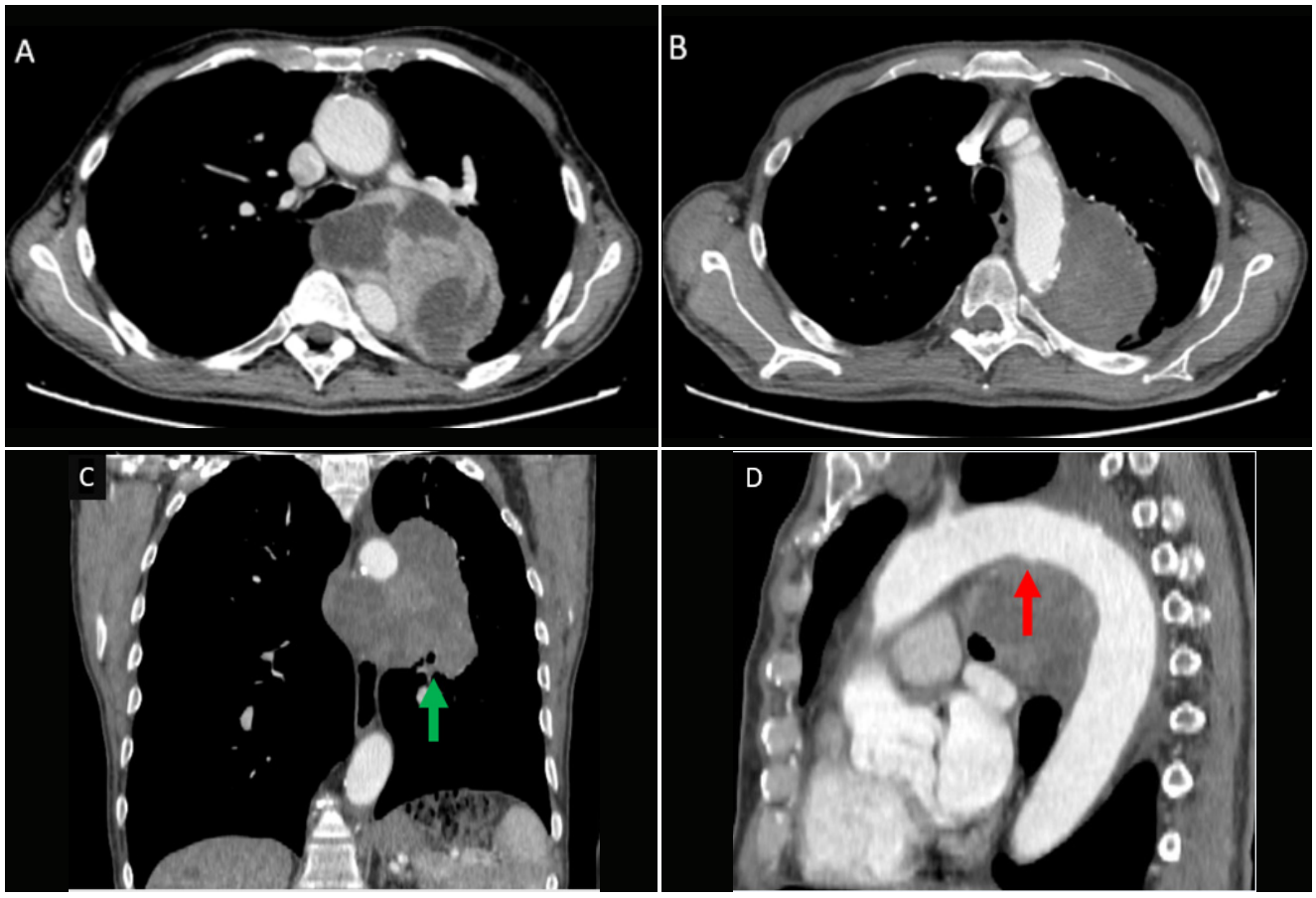


Figure 2: Multiplanar CE-CT reconstructions.
Figure 2a and 2b: arterial and venous phase axial CT, showing a large mass without early enhancement and indissociable from aortic walls, without clear signs of infiltration.
Figure2c: arterial phase coronal CT, showing clear proximity with the right main bronchus (green arrow)
Figure 2d: arterial phase parasagittal CT centered to the aortic arc, showing a tiny focal irregularity of the inferior aspect of the aortic wall, suspicious for infiltration (red arrow).
CE-CT: Contrast-Enhanced Computed Tomography; CT: Computed Tomography

the thoracic cavity. Most thoracic cases occur in the pleuropulmonary system while the mediastinum is an exceptionally rare site [10]. To date, in the largest series reporting 21 cases, the incidence of mediastinal synovial sarcomas has been estimated to be 11.2% of all thoracic synovial sarcomas [11].

The literature review showed few cases of mediastinal SS. Most of the cases were without a complete imaging path. Furthermore, a case with the involvement of the aortic walls is rare. We reported our experience in a patient with a bulky posterior mediastinal mass that surrounded the thoracic aorta, infiltrating it focally. MRI with the cardio-synchronized cine-MRI sequence was very useful to identify infiltration of the wall (Figure 3). In our case, the patient underwent neoadjuvant chemotherapy. Unfortunately, we do not have a clear therapy protocol available in our data.

STS treatment depends on the stage of the disease, resectability, and general status of the patient. Various therapeutic strategies could be used, including resection only, resection followed by adjuvant radiation and/or chemotherapy, and incomplete resection followed by chemoradiation [10, 11]. However, there is no definite consensus about the optimal treatment modality.

Patients with borderline resectable or unresectable localized disease can be treated with neoadjuvant chemotherapy to downstage the tumor and achieve an R0-resection [12, 13]. STS is sensitive to chemotherapy with reported response rates of 30–55%. Ifosfamide-based regimens are commonly used with or without doxorubicin and appear to be associated with the highest reported objective response rate [14].

Pre-operative and peri-operative radiotherapy (RT) and chemotherapy is the gold standard therapy for STS [14]. High-grade tumors usually respond with a greater degree of benefit. In the literature, there is no consensus about the timing of RT (preoperative and postoperative) [14]. In preoperative RT there is the benefit of delivering a lower total dose with a shorter course of treatment. Furthermore, the treatment field is smaller, leading to less radiation toxicity with an improvement in extremity functionality [14]. In the extremity, there is also potential downstaging of a borderline resectable sarcoma with an increased chance of preserving the limb. However, preoperative RT is associated with a higher rate of wound healing complications (35% for preoperative RT compared to 17% with postoperative RT) [14]. On the other hand, postoperative RT allows a definitive assessment of the tumor

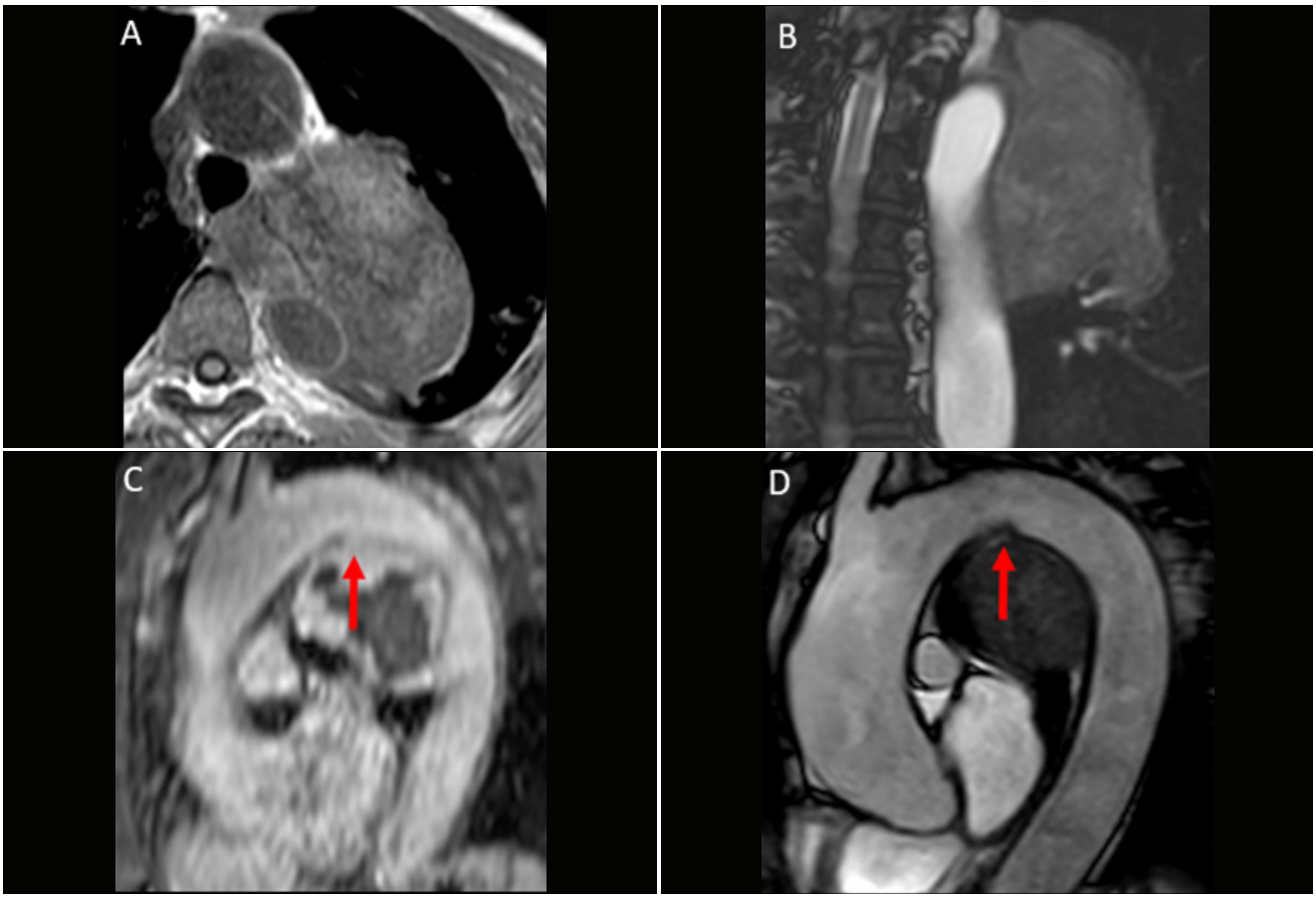


Figure 3: Multiplanar CE-MRI with cine-RM.
Figure 3a: axial T1 GE no-CE-MRI T1 sequence, showing an inhomogeneous hysointense mass indissociable from aortic walls.
Figure 3b: coronal TSE T2 no-CE-MRI sequence, showing an inhomogeneous iso/hyperintense mass suggestive of necrotic collections within the mass.
Figure 3c: parasagittal GE T1 CE-MRI sequence centered to the aortic arc, showing a tiny focal interruption of the inferior aspect of the aortic wall, suspicious of an infiltration (red arrow).
Figure 3d: cine-MRI in the same plane as fig.2c, confirming the tiny focal interruption of the inferior aspect of the aortic wall (red arrow).
CE-MRI: Contrast-Enhanced Magnetic Resonance Imaging; CINE-MRI: Cinematic Magnetic Resonance Imaging; GE: Gradient Echo; no-CE-MRI: Non-Contrast-Enhancement Magnetic Resonance Imaging; TSE: Turbo Spin Echo.

(grade, margin status, etc.) with a lower rate of postoperative wound healing complications [14]. However, it is associated with higher rates of fibrosis, edema, and joint stiffness. It is important to notice that although adding peri-operative RT improves the local outcomes, there is no evidence of any benefit in terms of overall survival or distant recurrence-free survival [14].

The use of neoadjuvant chemotherapy associated with RT is still a topic of debate [12]. Many chemotherapy regimens were tested. Ifosfamide emerged as the most effective drug and the treatment in the case of tumor necrosis presented the lowest rates of local recurrence [14]. Likewise, an escalating dose of gemcitabine and ifosfamide concurrently with preoperative RT (50 Gy) in patients with STS in the extremity/trunk, resulted in 5 years of local control, distant metastasis-free, and overall survival rates of 85%, 80%, and 86%, respectively [14]. However, despite encouraging results, neoadjuvant chemoradiation treatment remains experimental [14].

Surgery has to be done with a resection margin of more than 1 cm or an equivalent, like an intact fascia, considering the United Kingdom guidelines for STS [14]. Despite adding RT in patients with a positive margin after resection, the outcome is poor compared to those presenting a negative margin. Hence, the goal of surgery is to achieve a negative resection margin [14].

In our case, despite a clear imaging picture and partial chemotherapy response, a rupture of the aorta occurred during the surgery, resulting in hypovolemic shock that prevented the patient to survive the surgery. We believe that our imaging iconography can be useful to improve the radiological knowledge of this pathology. Indeed, when a mediastinal mass is found, consideration of a mediastinal SS in the differential diagnosis is important because, often, slow growth can mistakenly suggest a benign lesion, such as neurogenic tumors (neurinoma and paragangliomas). Mediastinal SS can arise in any mediastinal compartment, most commonly the anterior mediastinum, followed by the posterior mediastinum and middle and superior compartments [10]. The radiological appearance of mediastinal SS is non-specific, often precluding differential diagnosis from other mediastinal neoplasms. On X-ray, the tumors can appear as well-circumscribed neoplasms with sharply margined borders or as ill-defined infiltrative lesions. CT often reveals large tumor masses with a homogeneous or

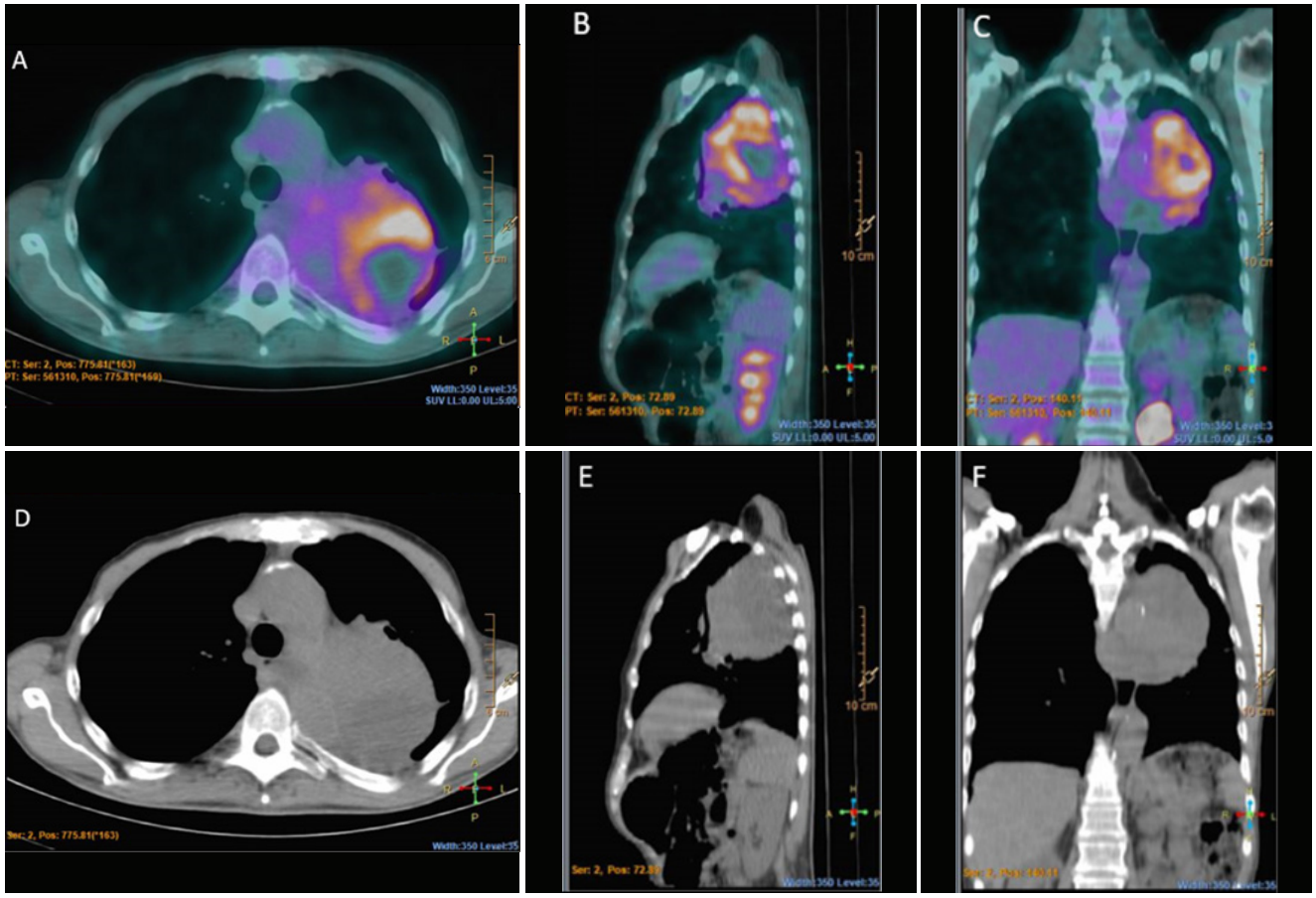


Figure 4: PET-CT.
Figure 4a, b, and c. Colorimetric axial (a), sagittal (b), and coronal (c) PET-CT images, showing an inhomogeneous active metabolism, due to wide ipocaptating necrotic areas within the mass (SUV max: 7,35).
Figure 4d, e and f. Corresponding Low-Dose axial (d), sagittal (e), and coronal (f) CT images.
PET-CT: Positron Emission Tomography – Computed Tomography; SUV: Standardized Uptake Value.

heterogeneous enhancement that show high uptake on PET. At MRI, the tumors typically demonstrate heterogeneous signal intensity on T1- and T2-weighted images and may contain fluid-fluid levels, due to hemorrhage or necrosis within cystic tumor components [15]. Cyst formation, areas of calcification, necrosis, and hemorrhage are common findings [10]. In our experience, heterogeneous enhancement, huge size, and infiltration of the aorta have been useful features to differentiate SS from benign tumors such as neurogenic tumors (usually with small size, homogeneous enhancement, and without aggressive infiltration). In this context, cardio-synchronized cine-MRI sequences, have been proved useful in loco-regional staging allowing better identification of aortic wall infiltration, and improving surgical planning (Figure 3).

Conclusion

In conclusion, SS originating in the mediastinum are rare neoplasms that are difficult to diagnose due to the unusual tumor location and the poor imaging experience [15]. In our opinion, although these tumors share many overlapping features with their soft tissue counterparts, it is useful to know that they are characterized by a slightly older age presentation, predominantly male, and usually have larger tumor sizes, heterogeneous enhancement, and may have aggressive infiltration [15]. Furthermore, we suggest considering cardio-synchronized cine-MRI to better define the tumor relationship with mediastinal structures, especially aorta and epi-aortic vessels, and to identify an aggressive infiltration useful in the differential diagnosis with benign neurogenic tumors.

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