Primary intraabdominal synovial sarcoma

A 56-year-old woman presenting to her general practitioner with a palpable, slowly growing and sometimes painful inguinal mass. The mass had been gradually increasing over the last year.

Initially the patient underwent an ultrasound exam that showed a heterogeneous mass deeper to subcutaneous cell tissue with AP axis larger than the transverse axis. It was predominantly hypoechoic and had internal vascularization. We performed a core needle biopsy showing a mesenchymal neoplasia suggesting synovial sarcoma.

Subsequently, a thoracoabdominal CT was performed for initial staging that showed an intraabdominal but extraperitoneal lobulated well-defined heterogeneously enhancing mass with some hypodense areas probably necrotic or cystic component and solid pseudonodular enhancing component with some punctate hyperdensities that could be peripheral calcifications. A pelvic MRI and FDG PET-CT were performed for presurgical planning. The mass does not infiltrate adjacent anatomic structures, it is hypointense on T1 and shows the typical triple signal pattern on T2 and a solid nodular enhancing component, seen as well on the DWI and ADC maps. Another hypermetabolic lesion was seen in the right humerus, likely a metastasis.

Synovial sarcoma is the fourth most common type of soft-tissue sarcoma, accounting for 2.5%–10.5% of all primary
soft-tissue malignancies worldwide [1]. Synovial sarcoma originates from primitive mesenchymal cells and therefore is a malignant mesenchymal biphasic or monophasic neoplasm most often affects the extremities (80%–95% of cases) [1], particularly the lower limb around the knee. Despite its name, the lesion does not commonly arise in an intraarticular location but usually occurs near tendons ? tendon sheaths and next to joint capsules. Synovial sarcoma also arises in areas with no obvious synovial or periarticular structures and has been described in almost all parts of the body thanks to the immunohistochemistry and demonstration of the t(X;18) chromosomal translocation or resulting SYT / SSX fusion gene transcripts, which is very specific for synovial sarcoma [1, 2]. It affects young adults with no gender predilection and manifests itself as a palpable and slowly growing mass that usually is > 5cm at the initial diagnosis. Primary intra-abdominal synovial sarcoma is rare, with less than 100 reported examples [3].

Histologic subtypes include three main variants: a classic biphasic type that has both a mesenchymal spindle cell component and epithelial component, a monophasic type (the most common) in which the spindle cells predominate and a poorly differentiated type [1]. Synovial sarcoma could present with calcifications, cystic changes and necrosis.

The imaging features of synovial sarcoma could be summarized as follows:

- **CT**: for initial staging. Non-infiltrative, well-defined mass often with punctate peripheral calcifications.
- **MRI**: for presurgical planning.
  - T1: hypointense
  - T2: triple signal pattern (hypo, iso, hyper to fat)
  - Gadolinium enhanced: solid nodular enhancing component
  - FDG PET-CT: to assess the efficacy of chemotherapy

The prognosis is poor because of the delay in diagnosis, which leads to local recurrence and metastasis. Only tumor size >5 cm is consistently associated with a poor outcome. Up to 50% of all synovial sarcomas recur locally, usually within 2 years, but sometimes many years later [2]. In fact, our patient had bone metastasis at the time of the diagnosis, confirmed with core needle biopsy.

The current treatment of choice is surgery, wide local excision (removal of the tumor, its pseudocapsule, and a normal cuff of surrounding tissue) without or with adjuvant therapy (if the margins are affected). The role of adjuvant therapy in the treatment of synovial sarcoma remains controversial. Chemotherapy has been used to treat metastatic or residual disease.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Primary intraabdominal synovial sarcoma with bone metastasis, Carcinosarcoma, Wilms’ tumour, Extragonadal germ cell tumours, Mesotheliomas, Solitary fibrous tumour

**Final Diagnosis:** Primary intraabdominal synovial sarcoma with bone metastasis

**References:**


**Figure 1**

Description: Ultrasound and Doppler ultrasound exams show a heterogeneous mass deeper to subcutaneous cell tissue with an AP axis larger than the transverse axis. It is predominantly hypoechoic and has internal vascularization. **Origin:** Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain
Description: Show a lobulated well-defined heterogeneously enhancing mass with some low attenuation central areas probably resulting from necrotic or cystic component and thick nodular wall peripherally with some peripheral calcifications. It is located intraabdominal. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.
Description: Axial T1-weighted fast spin-echo MR image shows a hypo-isointense homogeneous mass. Axial T2-weighted fast spin-echo MR image shows a heterogeneously hyperintense lobulated intraabdominal but extraperitoneal mass. It has the characteristic triple signal. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.
Description: Axial, sagittal and coronal fat-suppressed T1-weighted MR images show the intraabdominal mass without fat suppression. We can see on the coronal section that the mass does not cross the deep inguinal ring. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.
Description: Axial postcontrast fat-suppressed T1-weighted MR images show prominent enhancing peripheral soft-tissue (solid) component and a central cystic/necrotic component. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.
Description: DWI (b=800) shows that the solid component has a restricted diffusion ADC map is consistent with the previous findings and shows a low diffusion coefficient of the solid component. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.
Description: Axial FDG PET-CT images show a distinct hypermetabolic component within the mass but also show an additional hypermetabolic lesion in the right humerus. Origin: Department of Radiology, General University Hospital Morales Meseguer, Murcia, Spain.