Revisiting the upper extremity synovial sarcomas

Poster No.:	C-2398
Congress:	ECR 2018
Type:	Educational Exhibit
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Keywords:	Musculoskeletal system, Oncology, MR, Digital radiography, Diagnostic procedure, Cancer, Neoplasia
DOI:	10.1594/ecr2018/C-2398

Learning objectives

To recognize the typical imaging findings of synovial sarcomas with emphasis on the upper limbs.

Background

Synovial sarcomas (SS) are tumors of mesenchymal and epithelial origin and is the fourth most common type of soft-tissue sarcoma, accounting for 8%-10.5% of all primary soft-tissue sarcomas. SS occur in any age, but mostly in adolescents and young adults (15-40 years of age). They are the most common sarcoma subtype in the adolescent population.

Despite its name, is not related to synovial tissue and does not commonly arise in an intraarticular location, but usually occurs near joints. SS can appear in almost any anatomical site. Often affects the extremities (80%-95% of cases): the most common being the knee, in the popliteal fossa. The large minority of cases are located in the upper extremities, but they present the same imaging features.

Patients usually present themselves with a progressive slow growing palpable mass or swelling. The average duration of symptoms is 2- 4 years, but can vary from weeks to decades.

Diagnosis is made mostly through histologic analysis. There are three main histologic subtypes of synovial sarcoma: biphasic, monophasic, and poorly differentiated. Biphasic tumors being those with both mesenchymal and epithelial components. It has been discovered that SS are marked by the presence of a pathognomonic translocation between chromosomes X and 18, t(X;18)(p11.2;q11.2), that appear in more than 95% of patients and represents an important diagnostic tool.

SS is generally considered a high-grade sarcoma, marked by a poor prognosis, with an expected 5-year survival in adult patients ranging from 50% to 60% and a 5-year metastatic disease-free survival (DFS) ranging from 40% to 60%. SS show high metastatic potential. Tumor size, young age, tumors not from the extremities and response to first-line chemotherapy are good prognostic indicators.

Treatment is usually done by resection of the tumor, with or without neoadjuvant chemotherapy, followed by an adjuvant chemotherapy regimen.

Normal 0 false false false PT-BR JA X-NONE

The role of imaging is planning the surgical procedure, initial staging of the disease, either locally and by identifying distant metastasis, as well as post-treatment follow-up.

Findings and procedure details

- · **Radiograph**

Radiopgraph is usually the first imaging exam performed when patients seek medical attention. They are normal in about 50% of cases, particularly when lesions arel small. Plain radiograph usually show a non-specific round to oval soft-tissue lesion, usually close to articulations. Calcification are identified in up to 30%, and are often eccentric or peripheral within the mass. Chondroid or osteoid mineralization is rare. Bone involvement is not rare, but as SS growth is usyally slow, when in contact with bones it most commonly determine superficial pressure erosions, periosteal reaction and osteoporosis.

- · **Computed Tomography**

CT typically shows a heterogeneous, non-infiltrative and well-circumscribed soft-tissue mass with attenuation similar or slightly lower than that of muscle, often with punctate or peripheral calcifications.

It is better than plain radiograph in identifying small lesions and subtle calcifications as well as in evaluating local bone involvement, such as erosion or marrow invasion particularly in areas of complex anatomy such as the pelvis, hip, or shoulder.

CECT is useful in differentiating SS from cystic lesions and hematomas as it most often shows heterogeneous enhancement. Areas of nodular enhancement may also be found in synovial sarcomas. Areas of internal necrosis or hemorrhage are also common.

CT is also the modality of choice to identify pulmonary metastases. Calcification can sometimes be found in lung nodules.

- · **Ultrasound**

The US appearance of synovial sarcoma has not been extensively studied. SS most often present themselves as a nodular, round or lobulated, solid mass, most usually hypoechoic, that suggests a more slow growing and less aggressive process. A complex lesion, with areas of necrosis inside a more echogenic tumor (indicating cellular areas) is the second most common form of appearance.

Doppler evaluation can demonstrate vascularity in the areas of viable tumor, and is helpful in evaluating biopsy sites.

- · **Magnetic Ressonance Imaging**

MRI has a superior contrast resolution and is the modality of choice for local staging as well as assessing the intrinsic features of synovial sarcomas. SS are usually non-specific heterogeneous soft-tissue masses, but some characteristics can help to differentiate them from other sarcomas. On T1-wheighted imaging SS present itself as a heterogeneous soft-tissue mass, usually well-defined with signal intensity equal to slightly higher than that of muscle. Areas of T1 hyperintensity can also be found and it is usually related to intralesional hemorrhage. On T2 lesions shows a marked heterogeneity, with predominant high signal intensity. The triple sign has been described in T2WI and it is represented by areas of low, intermediate, and high T2 signal intensity, which is thought to be the result of a mix of solid tumor (intermediate T2 intensity), hemorrhage or necrosis (hight T2 intensity) and calcified/fibrotic areas (low T2 intensity). Triple sign has been shown to be present in up to 57% of cases. Fluid levels are also often observed within SS and the combination of cystic areas and/or hemorhagic foci, creates a *bowl of grapes* appearance. Post-contrast imaging usually show heterogenous enhancement, with marked contrast impregnation in the solid parts of the tumor.

MRI is also useful in post-chemotherapy evaluation, in which increasing T2 signal intensity is seen as a mark of progressive necrosis. Tumor size reduction is also a sign of adequate treatment response. Edema surrounding the tumor is not commonly seen before therapy, but may develop after adjuvant chemotherapy.

We reviewed 6 cases of synovial sarcomas in the upper extremities from our institution, all of them with biopsy confirmation. As reviewed in this text, they all show the same imaging characteristics as synovial sarcomas of any other sites.

Images for this section:

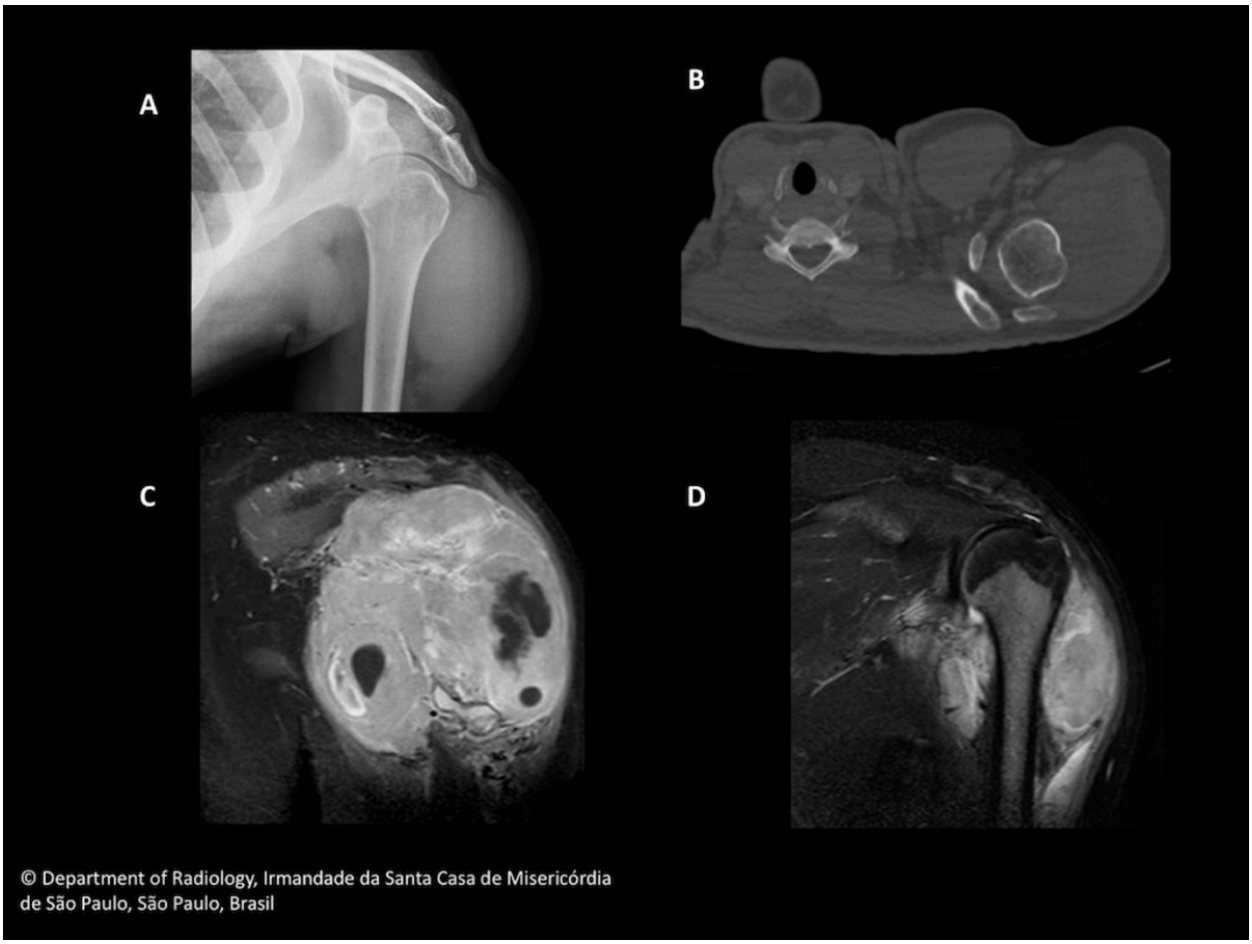

Fig. 1: 27 year old male with slow and progresive growth in the left shoulder for 6 months. A: left shoulder X-ray B: axial NECT C: Coronal T1WI-GD D: Coronal T2WI Lesion in the left shoulder invading all girdle muscles with cystic and hemorrhagic areas.

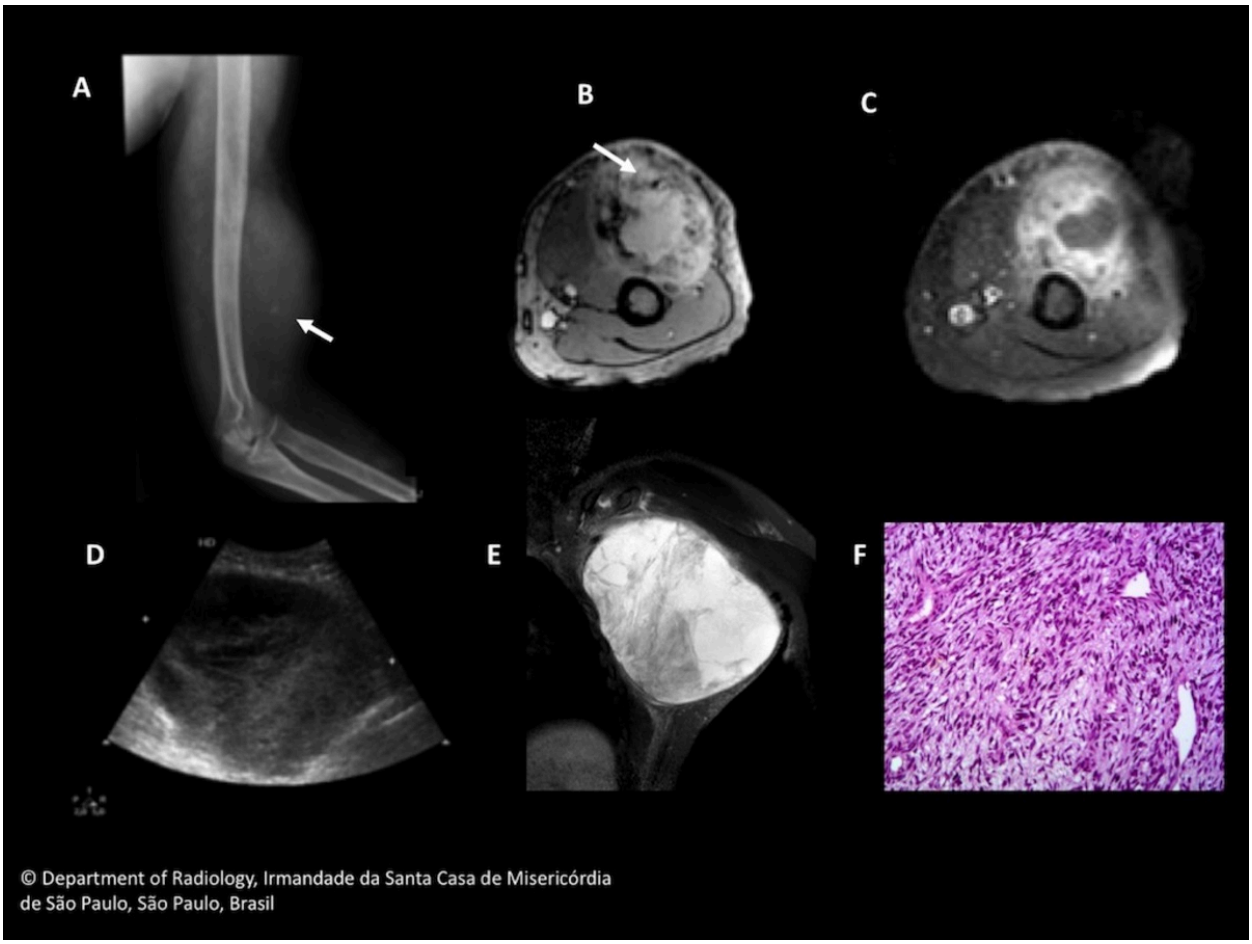

Fig. 2: 20 year old male with progressive growth in the left arm for 6 months submitted to ressection of the primary tumor, show up with an axillar lymphnode metasthasis after 3 months. A: left arm X-ray B: axial GRE MRI C: axial T1W-GD - shows lesion beetween the biceps and braquialis muscles D and E: US scan and STIR MRI of left axillar region. Arrows: Calcifications F: slide (10x), confirming the diagnosis

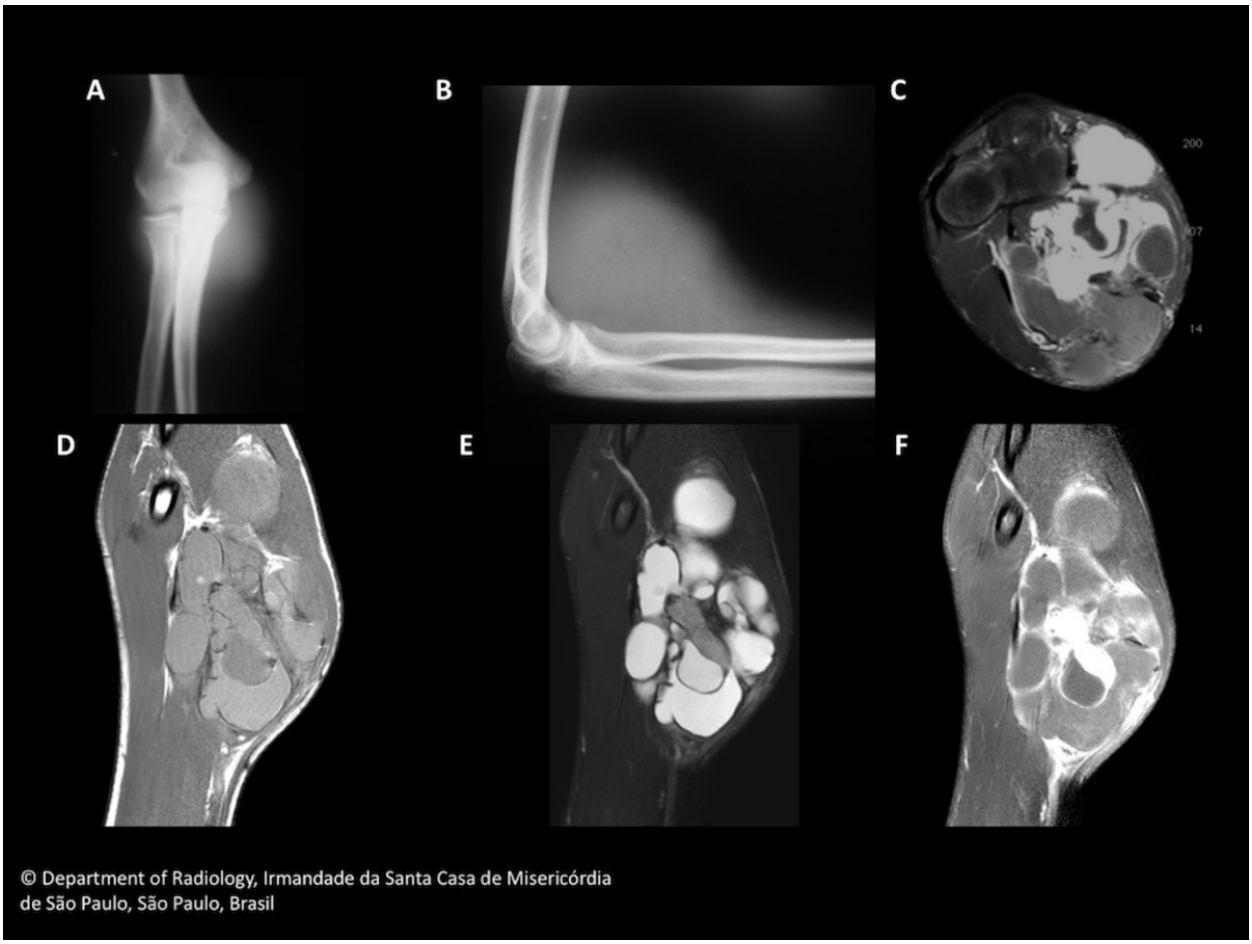

Fig. 3: 17 year old male with pain and swelling in the right elbow for 1 year A and B: right elbow X-ray C: Axial T1WI-GD D: Coronal T1W E: Coronal T2WI-fat sat F: Coronal T1WI-GD Anterior mass with cystic and hemorrhagic components invading the muscle of the anterior compartment of the elbow.

Fig. 4: 11 year old female with slow and progressive growth mass in the right elbow associated with movement impairment, A and D: right elbow X-ray B: axial T2WI C: axial T1WI E: Sagittal T2WI F: Sagittal T1-GD Shows lesion within the right elbow and forearm, that determines cortical erosion with signs of medular involvement of the proximal ulna

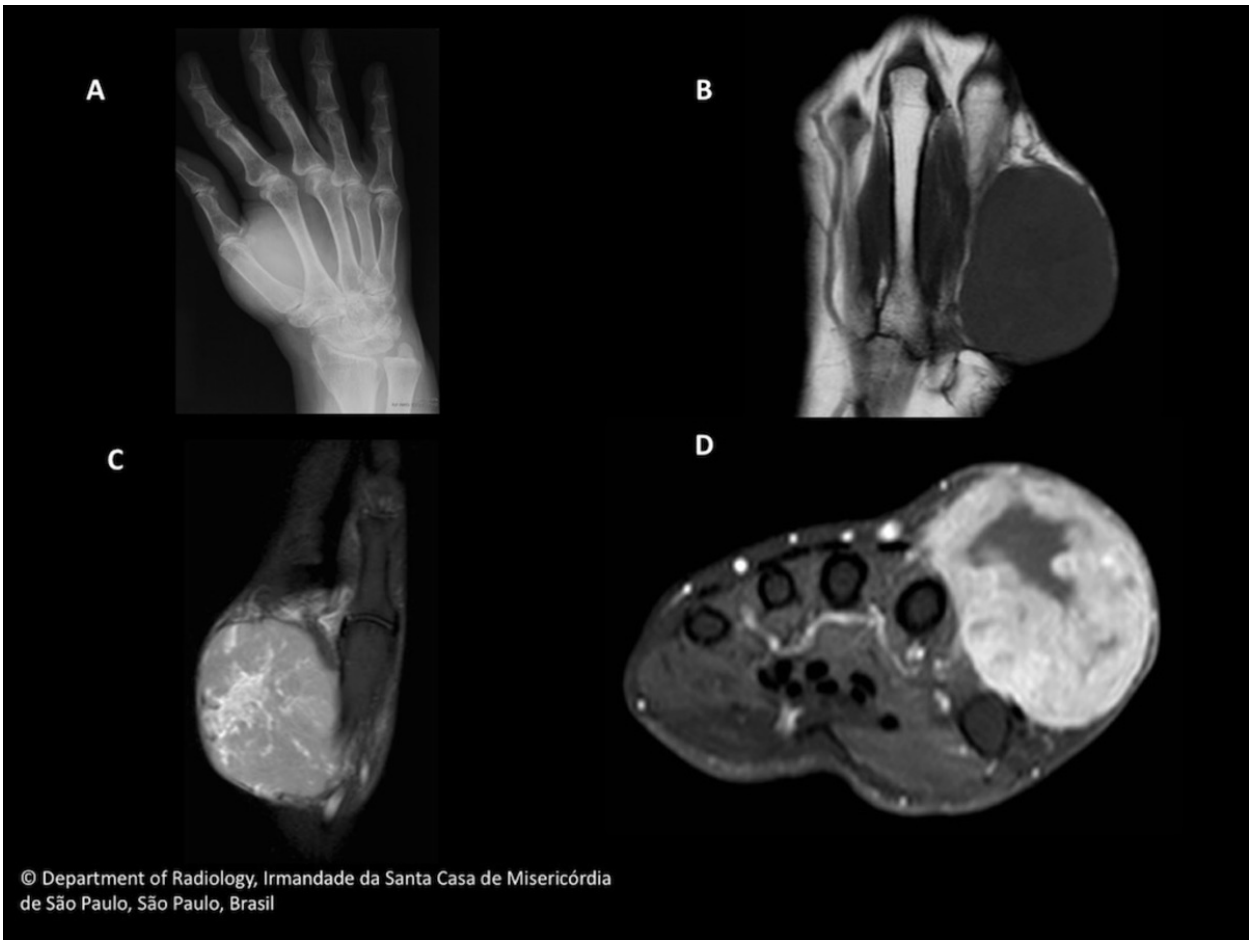

Fig. 5: 51 year old female with slow and progressive growth mass in the left hand region for 6 months. A: left hand X-ray ;B: Coronal T1WI; C: Sagittal T2WI fat-sat; D: Axial T1WI-GD Mass beetwen first and second fingers, invanding interosseous muscle, with central necrosis

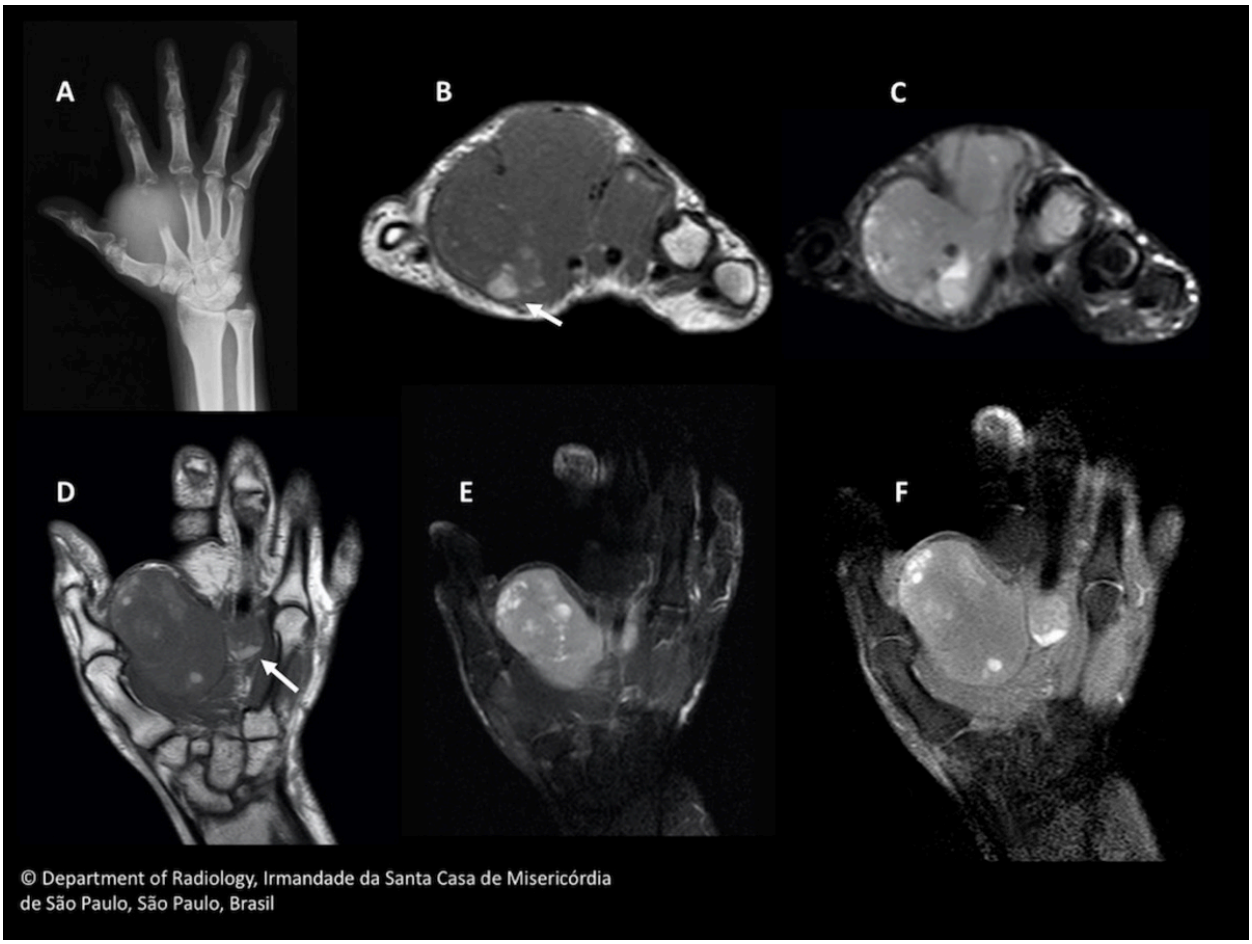

Fig. 6: 42 year old male with growing lesion in the right hand. A: right hand X-ray ; B: Axial T1WI; C: Axial T2WI fat-sat; D: Coronal T1WI; E: Coronal T2WI; F: Coronal T1-GD Lesion that promotes extensive erosion of the first and second methacarpal bones with involvement of the flexor and extensor tendons. Arrows: hemorrhagic areas inside the lesion with fluid level

Conclusion

Synovial sarcomas are rare and aggressive tumors. It should always be considered in the differential diagnosis of young adults presenting with a periarticular mass, particularly if it contains calcifications.

Although less common in the upper limbs, as we showed here, SS are a relevant differential diagnosis of lesions in these sites and its features should to be known by the radiologist.

The diagnosis of synovial sarcoma should also be considered when small periarticular masses are found and should be differentiated from other cystic lesions.

Finally, in addition to early metastasis, delayed metastasis is more typical of synovial sarcomas than other sarcomas and should be considered in determining the appropriate frequency and duration of follow-up imaging.

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