

Alveolar soft part sarcoma of the scapula

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Section: Musculoskeletal system

Area of Interest: Musculoskeletal system Lung

Procedure: Diagnostic procedure

Imaging Technique: Conventional radiography

Imaging Technique: CT

Imaging Technique: MR

Special Focus: Neoplasia Metastases Case Type:

Clinical Cases

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Patient: 33 years, male

Clinical History:

A 33-year-old man with no relevant pathological history presented with pain and swelling on the right scapular region and reduced range of motion of the right shoulder. The patient noticed those symptoms a few weeks before.

Imaging Findings:

A radiographic study of the right scapula was conducted showing bone expansion and destruction. These changes are suggestive of the presence of an aggressive lesion.

A chest CT study was performed, which showed the presence of a bulky mass involving the right scapula, measuring about 9 x 14 cm of greater antero-posterior and longitudinal axes. It causes destruction and bone expansion of the body and spine of the scapula, the distal portion of the acromion is spared. Multiple metastatic nodules are also identified in both lungs.

MRI was performed, the solid lesion has smooth lobulated borders, slight high signal comparing with the muscle on T1WI and T2WI. It has an internal nodule appearance with smaller masses separated by thin bands with low intensity on T1WI and T2WI. Some large-caliber vascular structures are identified within the mass.

An ultrasound-guided biopsy was performed and revealed the mass to be an alveolar soft part sarcoma.

Discussion:

Alveolar soft part sarcoma is a rare form of sarcoma, with distinct histology, molecular characteristics and clinical evolution [1]. It accounts for less than 1% of soft tissue tumors, and is characterized by a pseudoalveolar or organoid arrangement of polygonal tumor cells separated by fibrovascular septa and delicate vascular channels of capillary size [1]. It predominantly affects people of a young age group, namely children, adolescents and young adults, with a slight female predominance. This type of sarcoma usually occurs in deep tissues of the buttock and thigh in adults [1]. In children commonly occurs in the head and neck, predominantly in the orbit and tongue. It usually has an indolent evolution, being noticed when it reaches large dimensions, being metastasized at presentation in up to 65% of the adults and 30% of the children [1,2]. The most common metastatic organs are lung, bone, central nervous system and liver. Unlike other subtypes, alveolar soft part sarcoma often metastasizes to the brain. Surgery is the therapy of choice for localized disease and may be associated with adjuvant radiotherapy. Chemotherapy is usually ineffective. In disseminated disease the prognosis is worse. Features associated with

better prognosis include younger age at diagnosis, small tumor size and not disseminated disease. In imaging studies these tumors are usually well defined, lobulated, showing large vessels surrounding and inside the mass, emerging from the poles [2]. It usually shows strong enhancement after contrast administration on CT and MRI, mainly peripheral rim enhancement. It may have areas of necrosis. On MRI it is characterized by having iso or high signal comparing to muscle on T1WI. It may show a nodular pattern with several smaller nodules separated by thin bands of low signal intensity on T1WI and T2WI. On T2WI it can present a target morphology, with low signal hypointense at periphery and higher signal in the center. The most typical imaging features are the presence of peri and intra-tumoral vessels [2].

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Alveolar soft-part sarcoma, Other sarcoma subtypes, Metastasis

Final Diagnosis: Alveolar soft-part sarcoma

References:

- A L Folpe, A T Deyrup (2006) Alveolar soft-part sarcoma: a review and update. Journal of Clinical Pathology (PMID: [17071801](#))
- M. Beth McCarville, et al. (2014) Imaging Features of Alveolar Soft-Part Sarcoma: A Report From Children's Oncology Group Study ARST0332. Am J Roentgenol (PMID: [25415714](#))

Figure 1

a



Description: Front view radiogram showing expansion of the body and spine of the right scapula.

Origin: Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

Figure 2

a



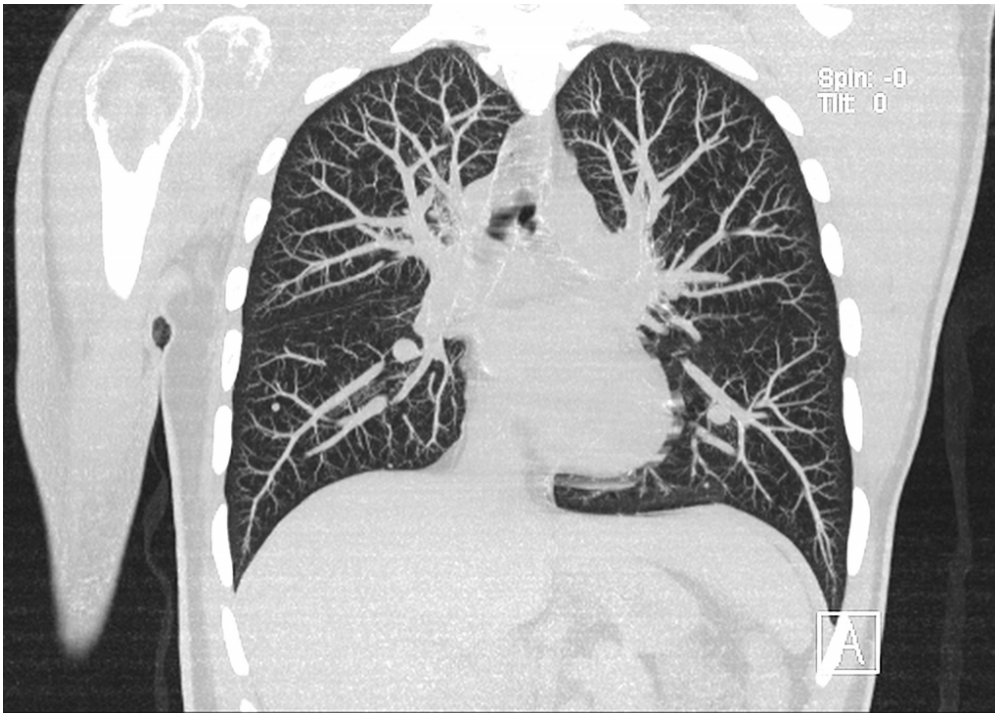
Description: Axial image showing a soft tissue mass involving the right scapula, with bone expansion and cortical interruption. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

b



Description: Sagittal reformat showing a bulky mass centered to the body of the right scapula. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

c

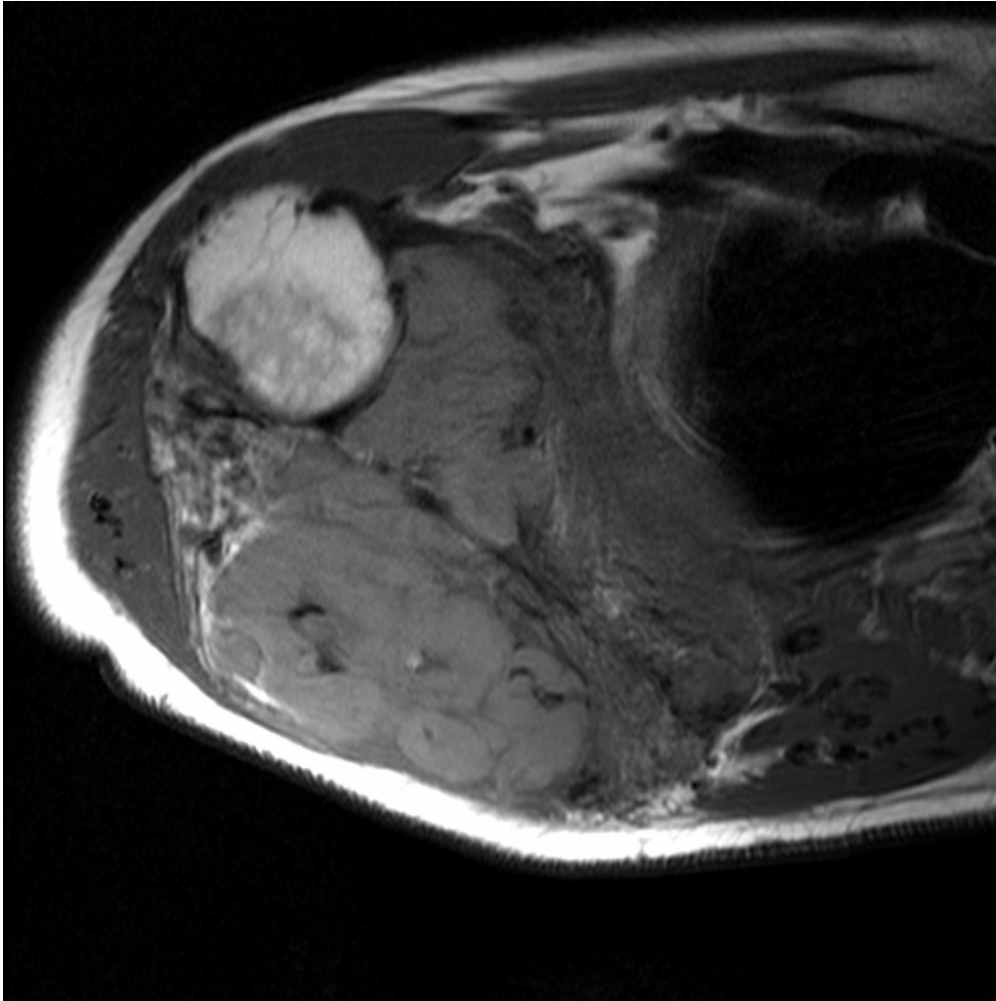


Description: MIP coronal reconstruction with lung algorithm showing many nodules in both lungs.

Origin: Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

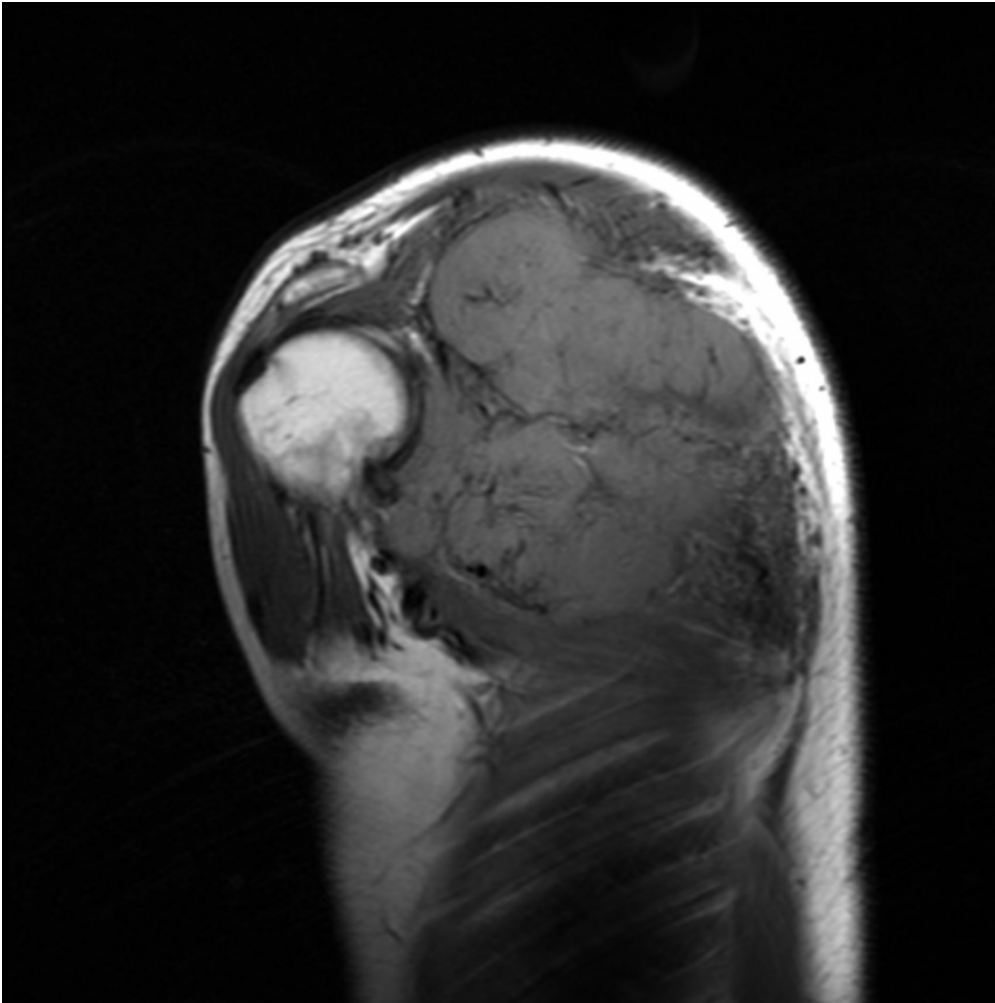
Figure 3

a



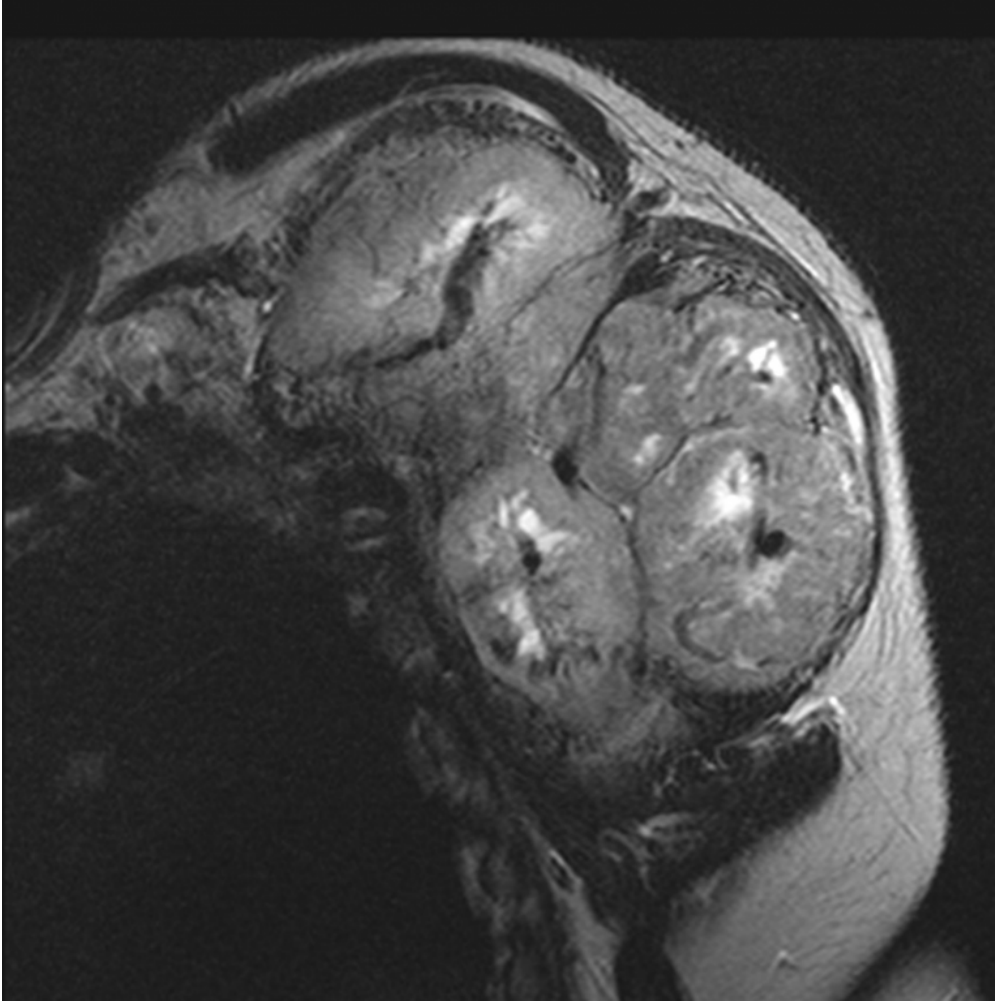
Description: Axial T1WI image showing a bulky high signal (comparing to the muscle) mass involving the right scapula. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

b



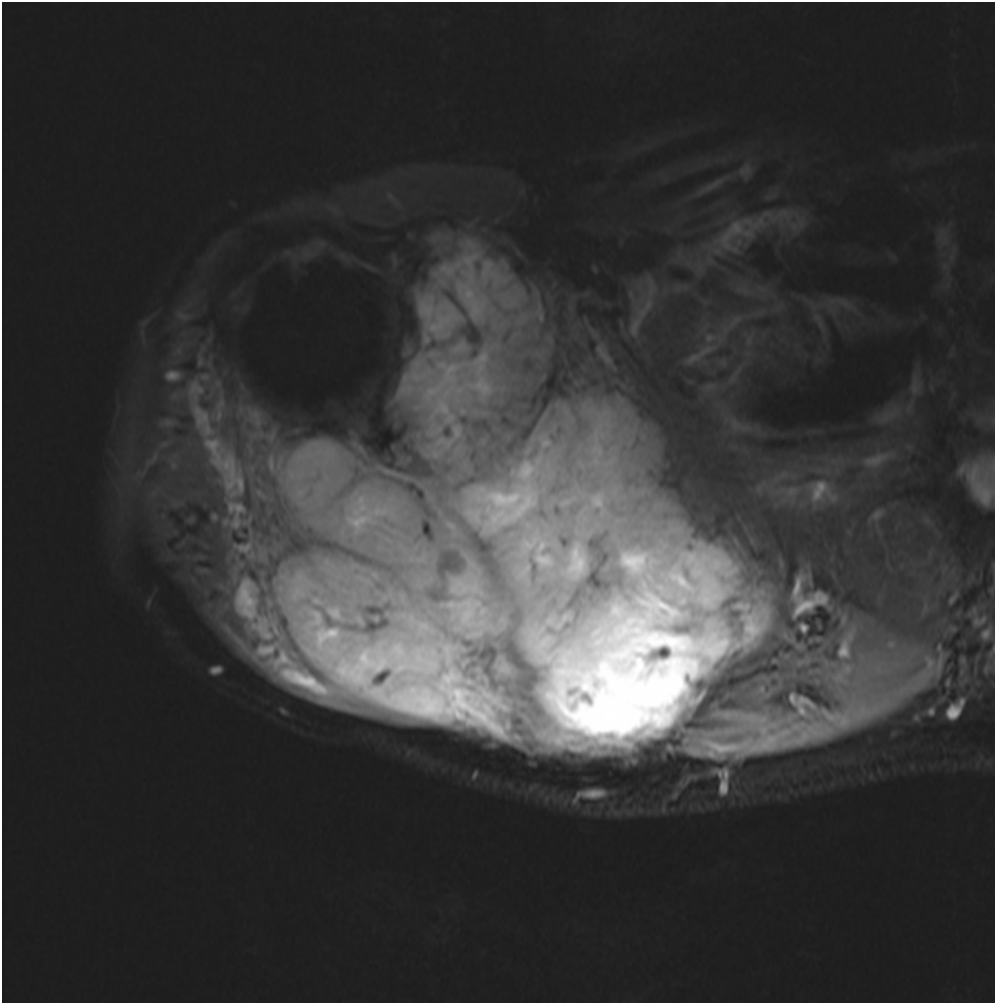
Description: Sagital T1WI image. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

c



Description: Sagittal T2WI image showing a bulky lobulated intermediate signal involving the scapula. The signal is inhomogeneous with an internal nodular appearance. There are also some large caliber vessels within the mass. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal

d



Description: Axial T2WI with fat suppression, the mass has high inhomogeneous signal. **Origin:** Centro Hospitalar e Universitário de Coimbra, Coimbra, Portugal