

Synovial Sarcoma in Upper Extremity in an Adolescent Boy

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Section: Musculoskeletal system

Area of Interest: Musculoskeletal soft tissue

Procedure: Diagnostic procedure

Imaging Technique: MR

Special Focus: Cancer Neoplasia Case Type: Clinical Cases

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Patient: 16 years, male

Clinical History:

A 16-years-old male with a history of non-Hodgkin lymphoma in remission for five years presented with a slowly growing and slightly painful mass in the posterior left arm.

Imaging Findings:

Grey-scale ultrasonography (US) demonstrated a well-defined, 5 cm solid mass in the triceps muscle with heterogeneous echotexture including isoechoic and hypoechoic areas. Colour flow Doppler US revealed mild vascularity within the mass (Figure 1).

The mass presented with the following imaging features on magnetic resonance imaging (MRI); isointense with skeletal muscle on T1-weighted (W); slightly heterogeneous and hyperintense on T2W images and almost homogenous enhancement in the delayed phase of the contrast administration (Fig.2, Fig.3). Dynamic contrast-enhanced (DCE) MRI revealed type III (rapid early enhancement followed by plateau/no significant wash-out) enhancement pattern an early enhancement noted within 7 s of contrast arrival into adjacent vascular structures (Fig.4).

In ADC maps, values varied between $0.9 \times 10^{-3} \text{mm}^2/\text{s}$ and $1.7 \times 10^{-3} \text{mm}^2/\text{s}$ (Figure 5).

Positron emission tomography revealed low uptake (SUV: 1) of ^{18}F -Fluorodeoxyglucose in the mass with no metastasis in other sites of the body (Figure 6).

Discussion:

Synovial sarcoma (SS) is a malignant tumor arising from mesenchymal cells and assumed to have resulted from the presence of a t(X:18) translocation with a fusion of the SYT (at 18q11) and SSX1 or SSX2 genes (at Xp11) which is present in 95% of tumours [1]. SS accounts for 8% to 10% of all soft tissue sarcomas being second-most prevalent soft tissue tumours after rhabdomyosarcoma in children and adolescents [2].

Patients with SSs usually present with slowly enlarging mass with or without pain [2]. Extremities are the most frequent site (80% to 95%) of involvement [2]. On US, SSs may present as round or lobulated, solid, hypoechoic mass resembling a benign tumour but some may show a heterogeneous and complex echotexture including hypoechoic and hyperechoic areas with irregular margins [3]. On CT, SSs most commonly present as well-circumscribed and non-infiltrative deeply seated mass with slightly lower attenuation compared to skeletal muscle [1].

On MRI, SS typically appears as a prominently heterogeneous multilobulated soft tissue mass with signal intensity similar to or slightly higher than that of skeletal muscle on T1-weighted (T1W) MR images. In T2W images, lesions mostly demonstrate heterogeneous high signal intensity [1]. T2W images may demonstrate characteristic 'triple signal intensity' that refers to the combination of high, intermediate, and low signals in the mass representing areas of necrosis, calcification, haemorrhage, and cyst formation respectively [1, 4]. SSs usually shows diffusion restriction on DWI, it has been shown that ADC values may range between $0.8 - 1.9 \times 10^{-3} \text{ mm}^2/\text{s}$ [5, 6]. In our patient, ADC values ranged between $0.96 - 1.68 \times 10^{-3} \text{ mm}^2/\text{s}$.

Synovial sarcomas manifest with early diffuse enhancement at dynamic contrast-enhanced MR imaging [8]. It has been shown that rather than peripheral enhancement and washout phase enhancement of tumour, within 7 s after arterial enhancement is the only sign that occurs consistently in synovial sarcoma patients as seen in our patient [8].

Treatment of SS may be accomplished with surgery with or without

chemotherapy and/or radiotherapy. Tumours with high grade or large size (> 5 cm) or where resection is felt to be difficult and margins will be close or positive may need neoadjuvant chemotherapy followed by surgical excision [1].

Take-Home Message / Teaching Points:

- A solid mass with a hypoechoic appearance in US even with well-defined contours should alert to exclude malignancy.
- Differentiation between SSs from other malignant and some benign soft tissue tumours are often not possible with conventional MRI features.
- Soft tissue mass adjacent to joint tendons, bursal surfaces in an adolescent patient with rapid contrast enhancement should always arise the possibility of SS.
- SS may present with low FDG uptake on PET-CT.

Differential Diagnosis List: Synovial sarcoma (monophasic type) in the triceps muscle, Benign peripheral nerve sheath tumor (PNST), malignant PNST, hemangioma, lymphoma

Final Diagnosis: Synovial sarcoma (monophasic type) in the triceps muscle

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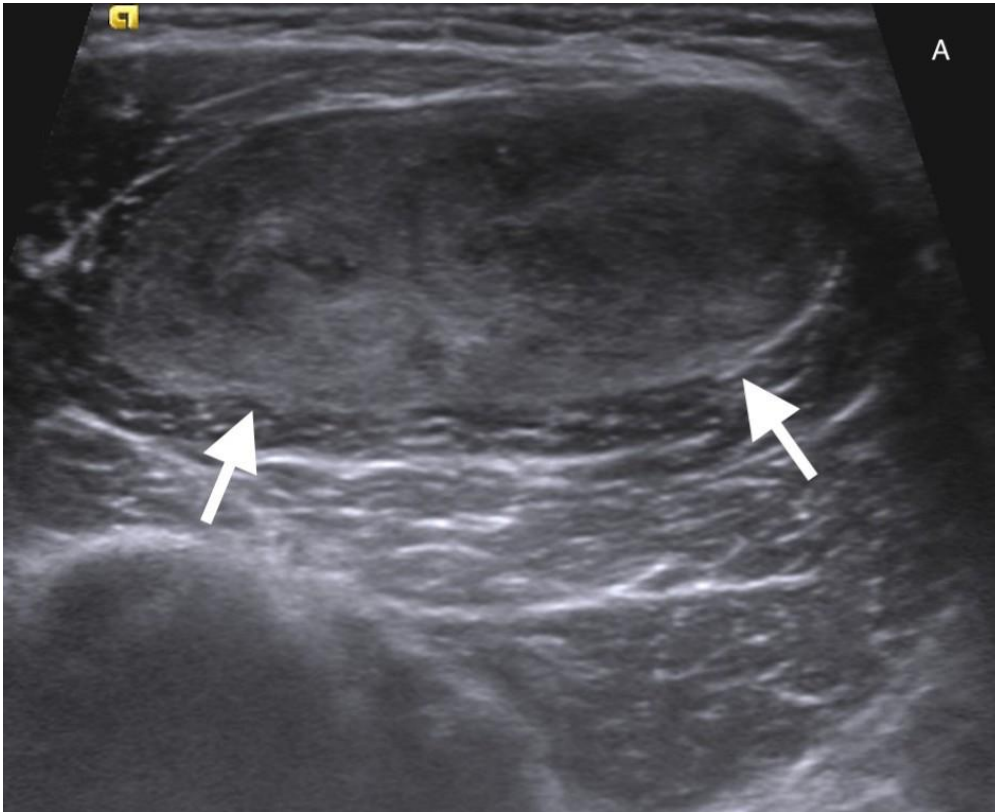
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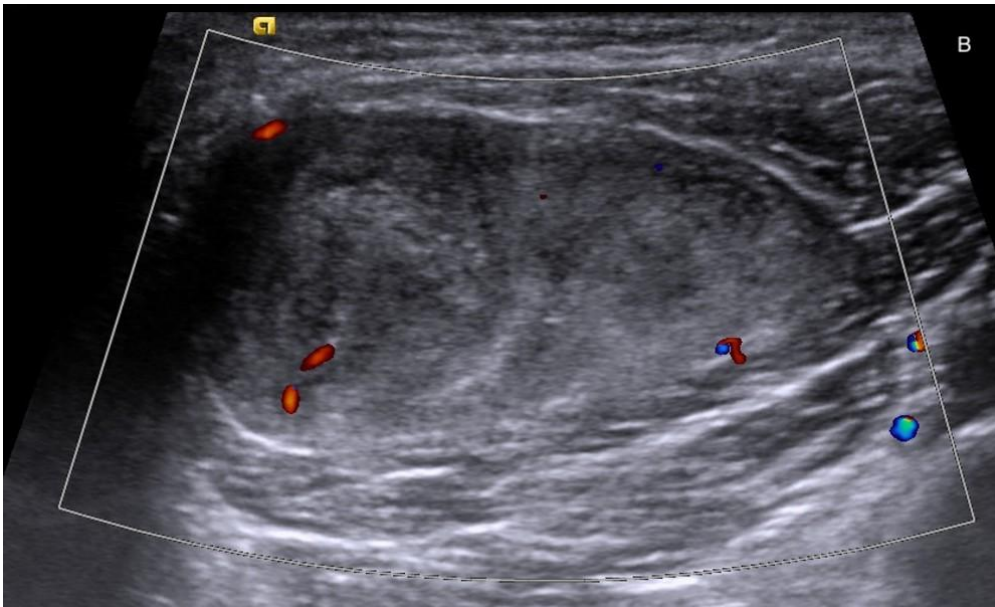
Figure 1

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Description: Gray-scale US shows a well-circumscribed mass with predominantly hypoechoic and mildly heterogeneous echotexture (arrows) in the medial head of the left triceps muscle. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

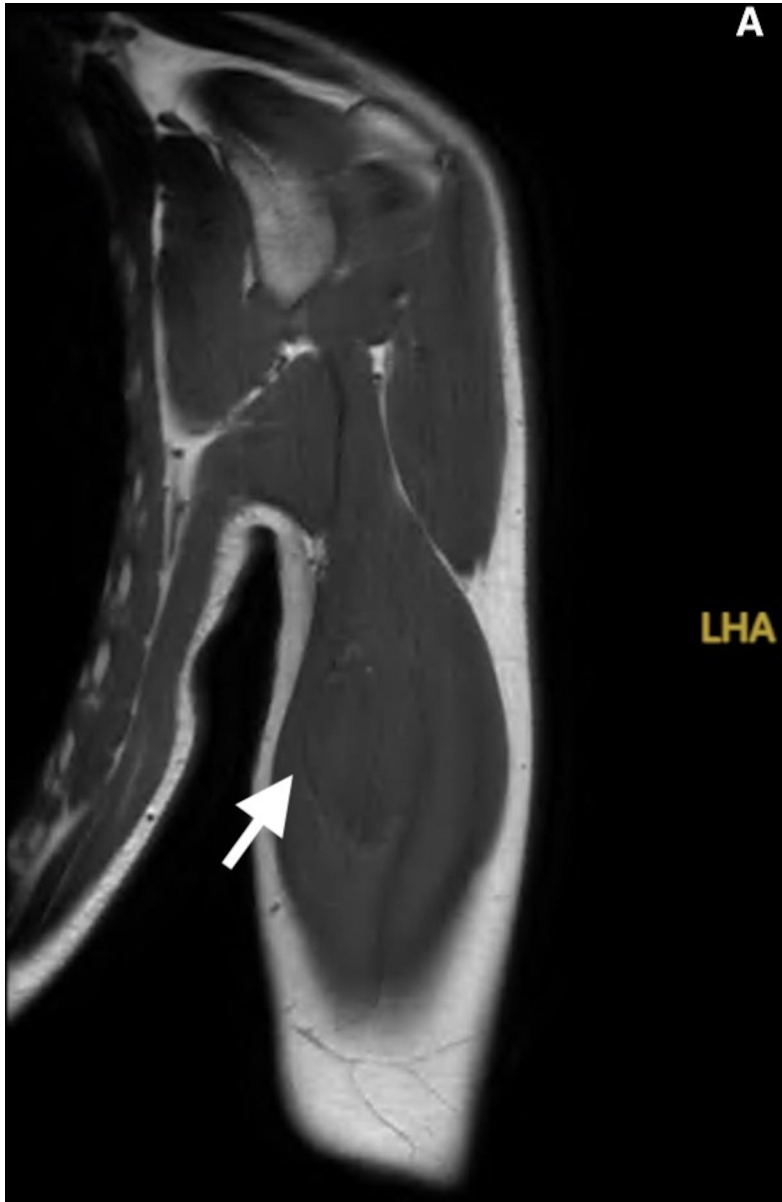
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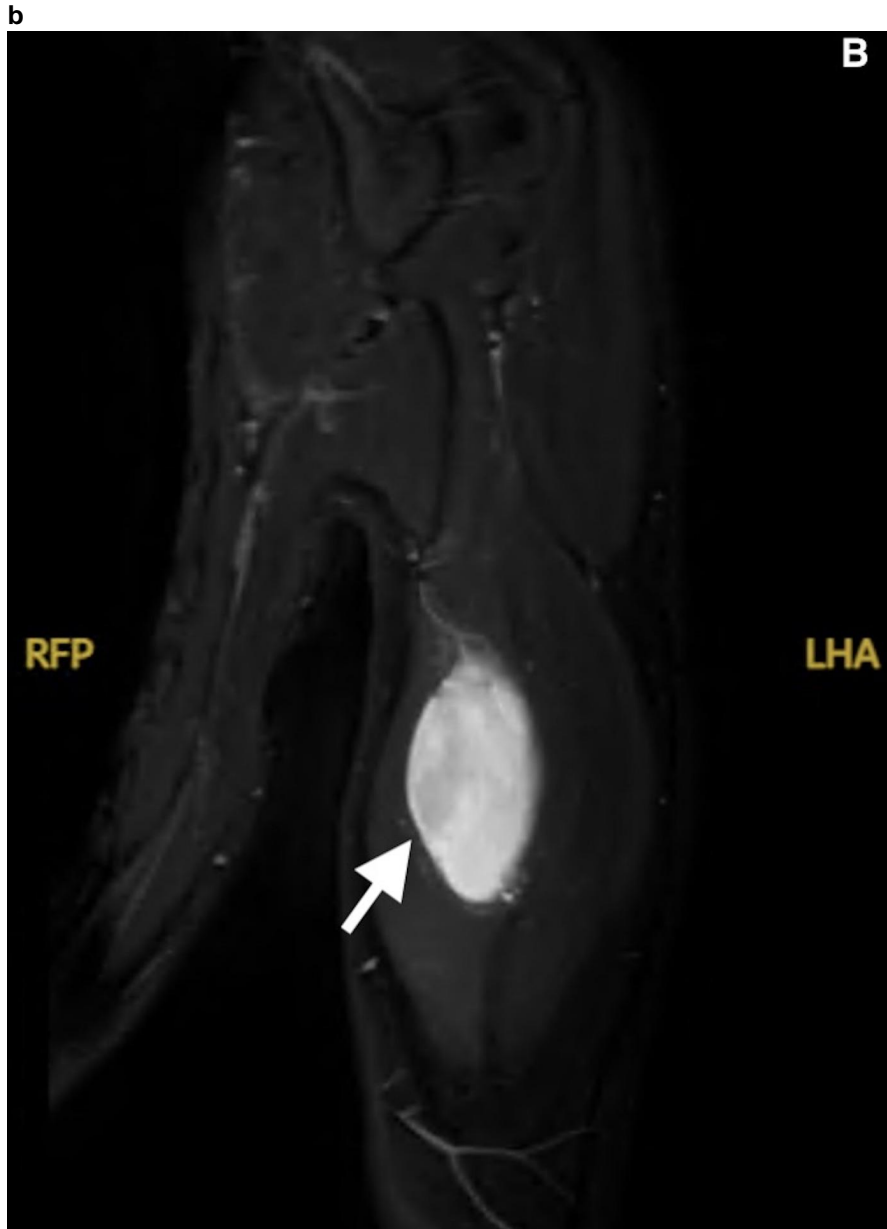
Description: Color-flow Doppler US demonstrates scant vascularity within the mass. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

Figure 2

a



Description: Coronal T1W (A), fat-saturated T2W (B), and post-contrast T1W images (C) demonstrate a 5 cm mass (arrows) with a similar signal intensity of the skeletal muscle on T1W, hyperintense heterogeneous signal on T2W image, and homogeneous enhancement (arrow) after intravenous contrast agent administration. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021



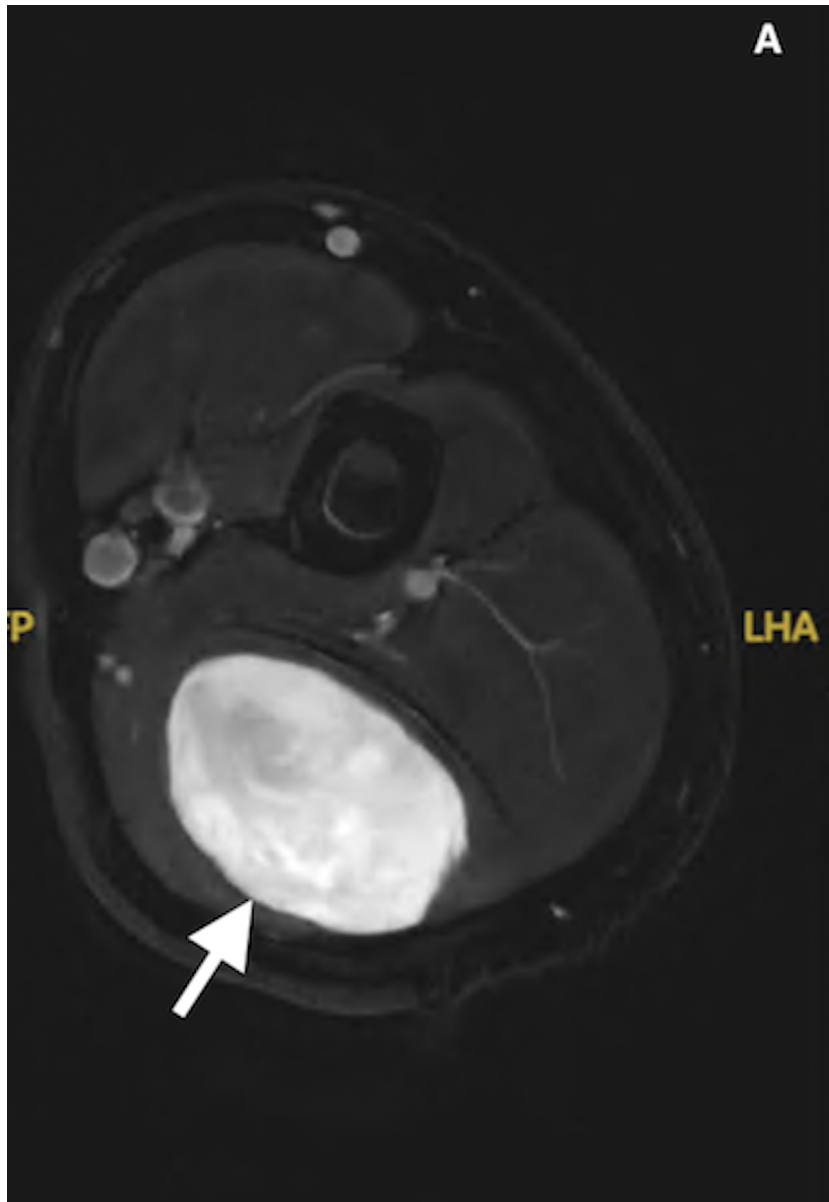
Description: Coronal T1W (A), fat-saturated T2W (B), and post-contrast T1W images (C) demonstrate a 5 cm mass (arrows) with a similar signal intensity of the skeletal muscle on T1W, hyperintense heterogeneous signal on T2W image, and homogeneous enhancement (arrow) after intravenous contrast agent administration. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021



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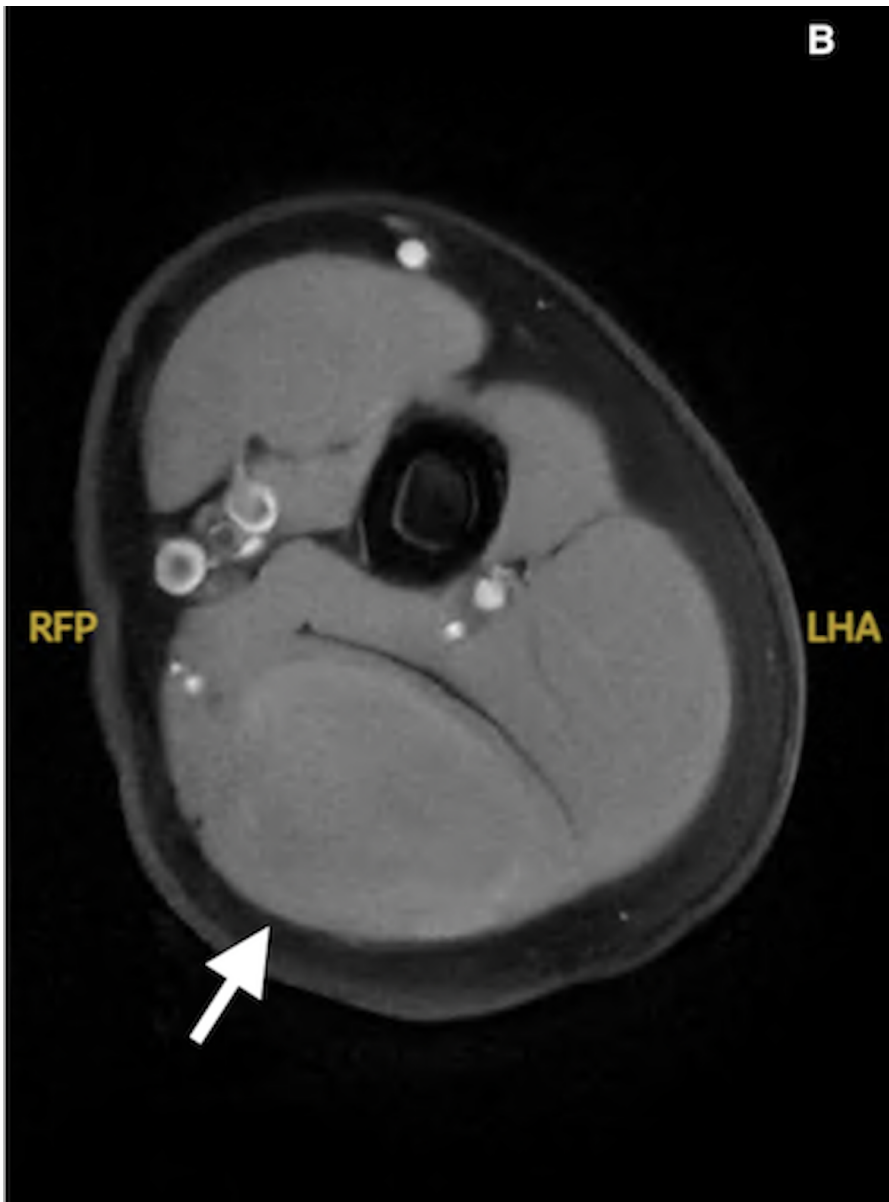
Figure 3

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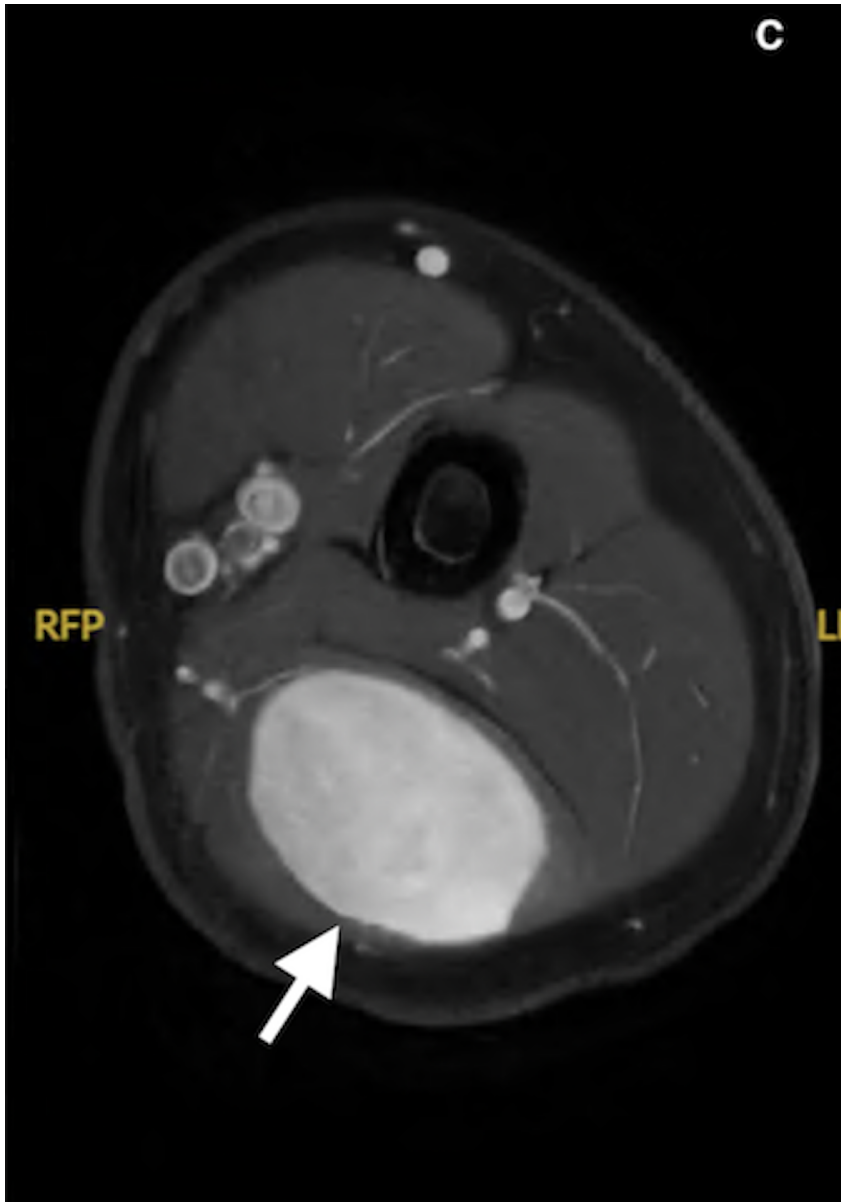
Description: (A) Transverse fat-saturated T2W MR image reveals heterogeneous T2 signal within the tumor (arrow) with well-defined margins. Pre and (B) post-contrast T1W (C) fat-saturated MR images demonstrate avid enhancement of the mass (arrows) without perilesional enhancement/edema. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

b



Description: (A) Transverse fat-saturated T2W MR image reveals heterogeneous T2 signal within the tumor (arrow) with well-defined margins. Pre and (B) post-contrast T1W (C) fat-saturated MR images demonstrate avid enhancement of the mass (arrows) without perilesional enhancement/edema. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

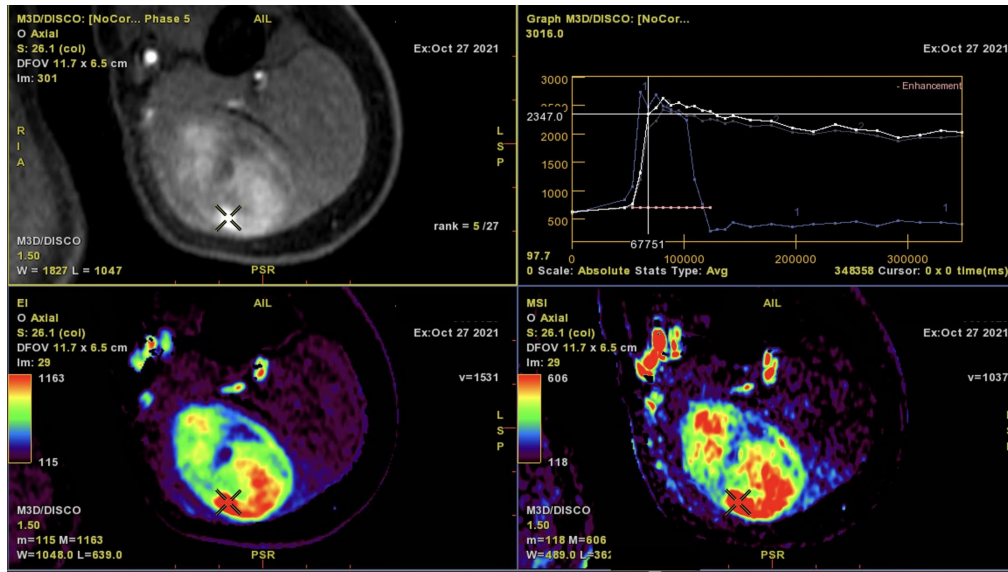
c



Description: (A) Transverse fat-saturated T2W MR image reveals heterogeneous T2 signal within the tumor (arrow) with well-defined margins. Pre and (B) post-contrast T1W (C) fat-saturated MR images demonstrate avid enhancement of the mass (arrows) without perilesional enhancement/edema. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

Figure 4

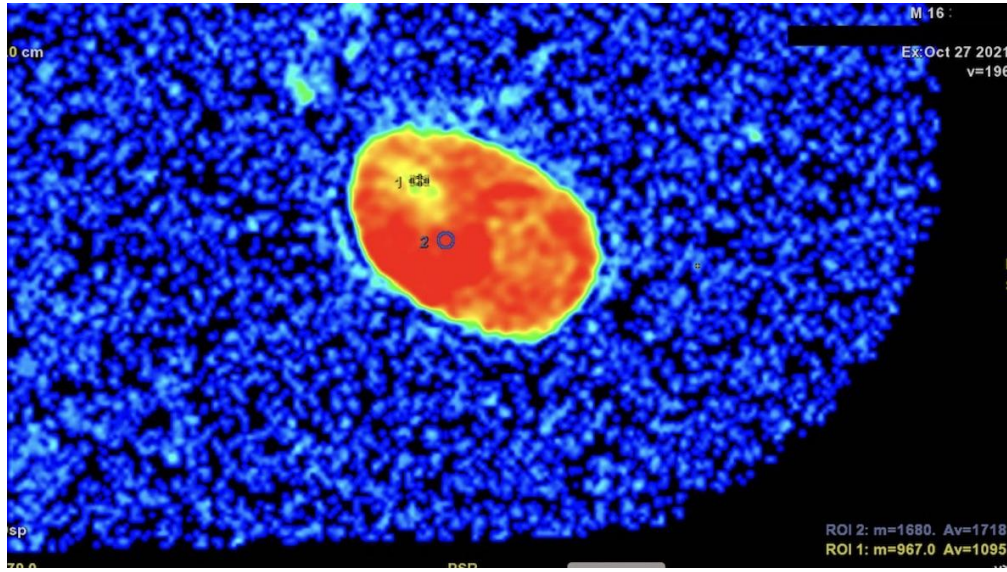
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Description: Dynamic contrast-enhanced MRI reveals type III (rapid early enhancement followed by plateau/no significant wash-out) contrast enhancement pattern. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

Figure 5

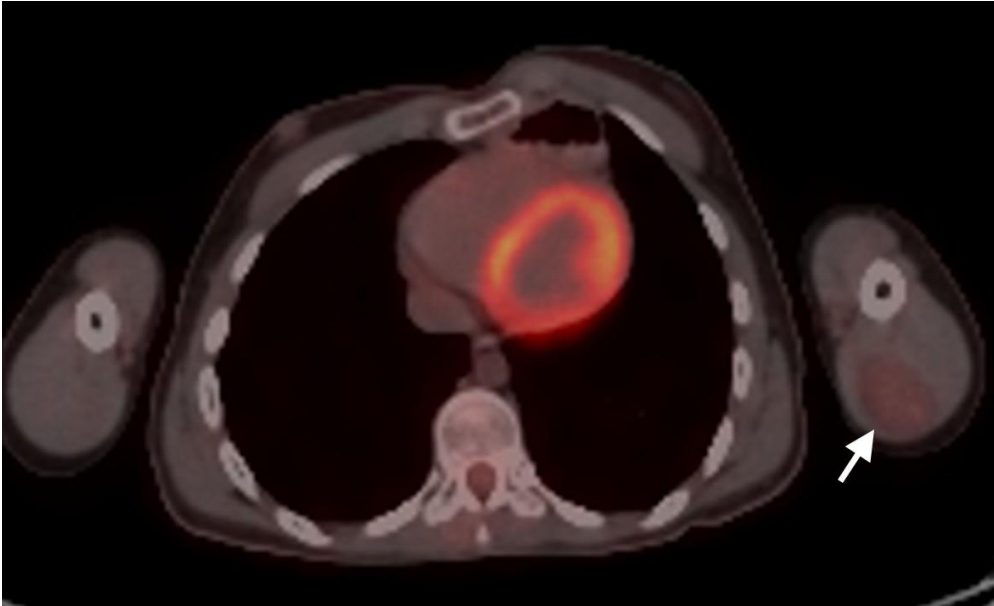
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Description: Colour-coded ADC map showed ADC values of $0.96 \times 10^{-3} \text{ mm}^2/\text{s}$ and $1.68 \times 10^{-3} \text{ mm}^2/\text{s}$ in two different areas within the mass. **Origin:** Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021

Figure 6

a



Description: 18F-Fluorodeoxyglucose PET/CT reveals a low FDG uptake within the mass (arrow).

Origin: Department of Radiology, Hacettepe University Medical Faculty Hospital, Ankara, Turkey, 2021