

Case Resolution

Rodrigo Re,^{*} Maximiliano Negri^{**}

^{*}Diagnostic Imaging Service, Osteoarticular/Musculoskeletal Area - Interventionism, Sanatorio Allende, Córdoba, Argentina

^{**}Department of Pediatric Orthopedics and Traumatology, Sanatorio Allende, Córdoba, Argentina

See case presentation on page 3.

DIAGNOSIS: Alveolar rhabdomyosarcoma.

DISCUSSION

In the magnetic resonance, a lesion inside the epitrochlear muscular plane, with slightly defined edges, was visualized as hypointense in the T1 sequence (Figure 3A and B) and hyperintense in T2 and fat-suppression sequences (Figure 3C and D). It was not possible to identify bone involvement.



Figure 3. MRI of the right elbow, without contrast. **A.** Coronal plane in T1 sequence: expansive lesion at the level of the epitrochlear muscular plane, hypointense, with slightly tapered edges (arrows). **B.** Axial plane in T1 sequence. **C.** Coronal plane in STIR sequence: a markedly hyperintense lesion is visualized. **D.** Axial plane in STIR sequence: area of central necrosis (arrow).

Dr. RODRIGO RE • rodrigo_re@hotmail.com  <https://orcid.org/0000-0001-7382-9459>

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In the diffusion sequences, it was hyperintense, with low signal in the apparent diffusion coefficient in the peripheral sector (average value $0.7 \times 10^{-3} \text{ mm}^2/\text{s}$), raising the possibility of a lesion with high cellularity (Figure 3E).

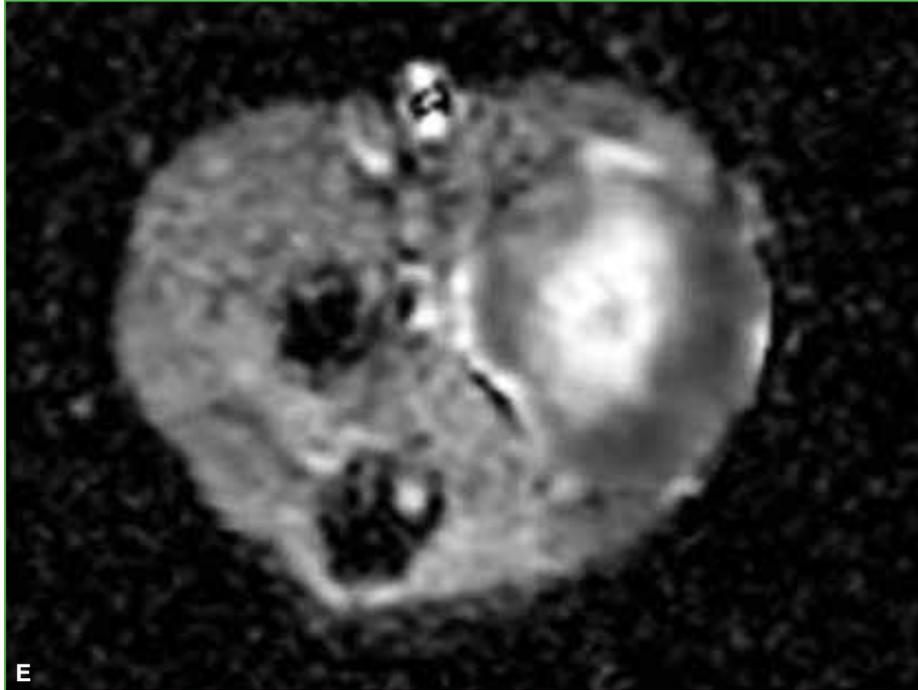


Figure 3. E. Axial plane showing the apparent diffusion coefficient (ADC) with low signal, possibly due to high cellularity.

Given these findings, a consultation was requested in the Hematology-Oncology Service and the slightly painful palpable mass was verified.

The patient underwent studies to establish the stage of the lesion. A CT scan of the chest showed no distant lesions (Figure 4). The bone scan was normal (Figure 5).

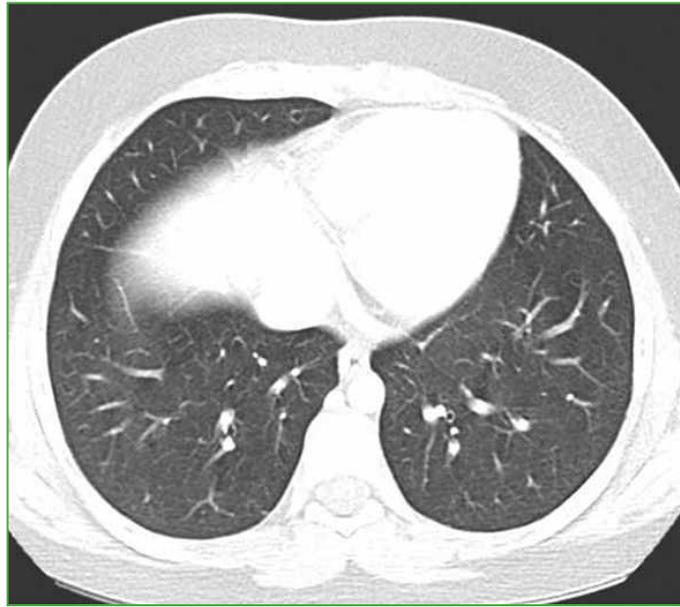


Figure 4. CT scan of the chest (lung window). Clear lung bases.

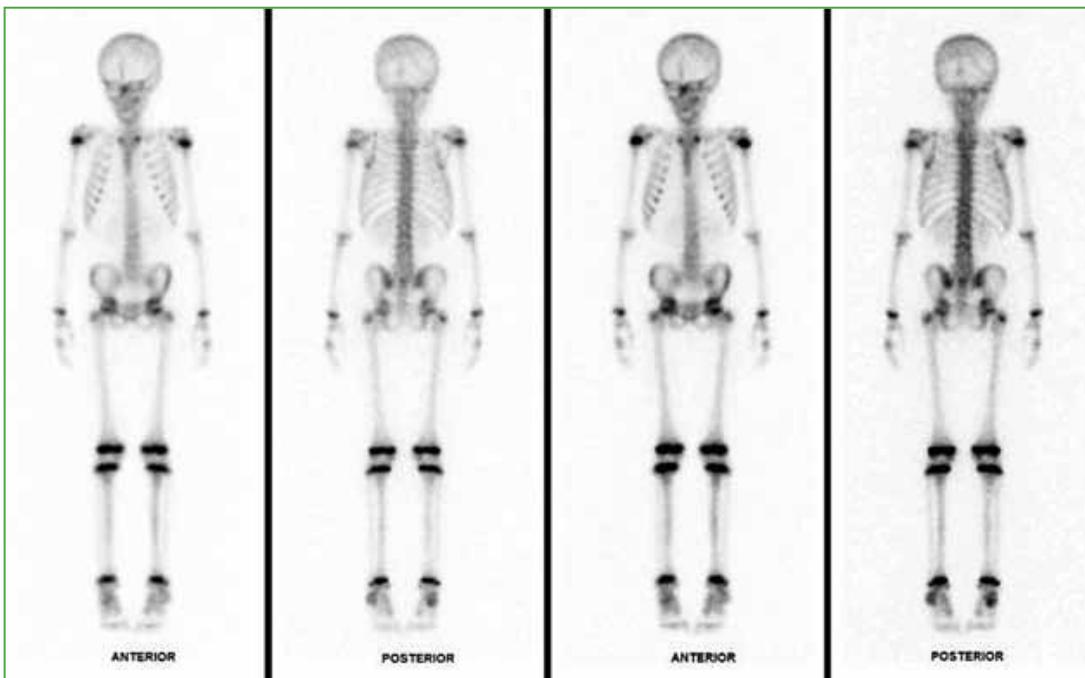


Figure 5. Bone scintigraphy reveals no distant lesions.

Subsequently, as it was a single lesion, the Tumor Committee decided to perform an ultrasound-guided biopsy (Figure 6). The pathological anatomy study reported a malignant mesenchymal neoplasm of small round blue cells, compatible with rhabdomyosarcoma. Immunolabeling yielded the following result: 1. Vimentin: Positive, 2. Desmin: Positive, 3. S100: Negative, 4. MYOD1: Positive, 5. Ki67: 60% proliferation, 6. P53: 40% positive cells.

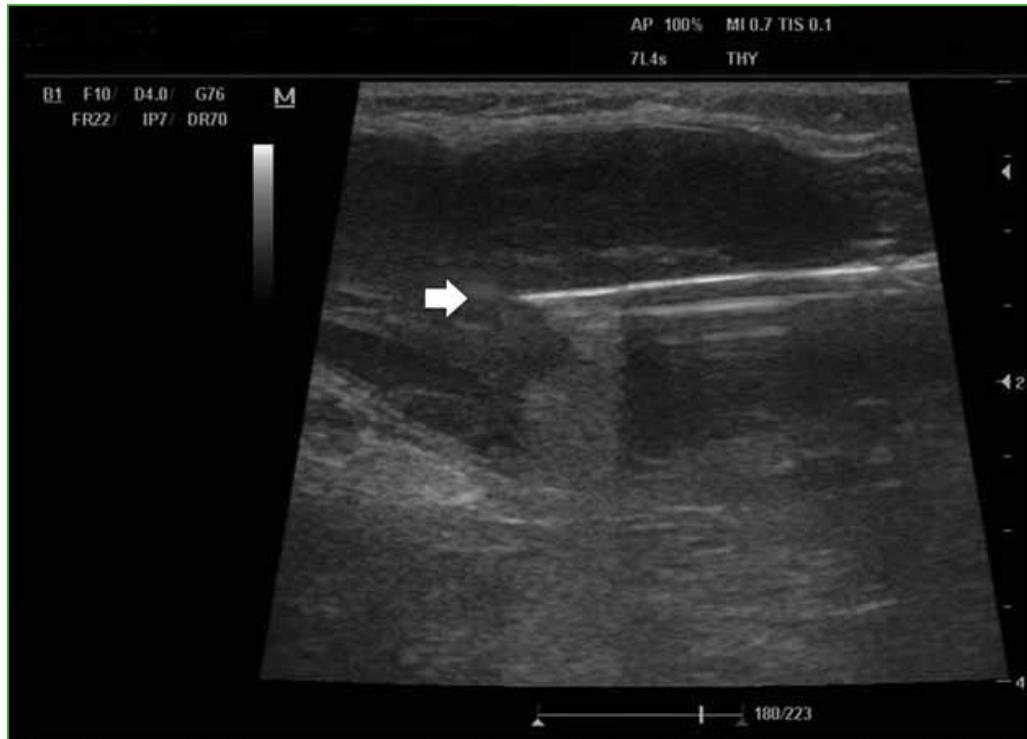


Figure 6. Ultrasound-guided biopsy of the soft tissue lesion with 14-10 G cutting needle, with the semi-automatic system. The tip of the needle inside the lesion (arrow) is displayed.

DIAGNOSIS

With all these findings, alveolar rhabdomyosarcoma was diagnosed.

Rhabdomyosarcoma is the most common malignant soft tissue tumor in pediatrics. It originates from immature mesenchymal cells with subsequent differentiation to striated muscle. It accounts for 20% of soft tissue sarcomas. It can affect the entire body, especially the head and neck, genitourinary tract, and extremities. At the time of diagnosis, it usually measures approximately between 3 and 4 cm. In 25% of cases, there may be bone involvement, with periosteal invasion and reaction.

Rhabdomyosarcoma has three variants: embryonic, alveolar, and pleomorphic. Embryonal rhabdomyosarcoma is the most common and predominantly affects children <5 years. The alveolar type affects children and young adults. The pleomorphic type is rare, exclusive to patients between 40 and 60 years old. Regarding its location, the embryonic type can be located in the head, neck, urogenital apparatus (botryoid variant), and retroperitoneum. The alveolar type involves the limbs, such as the forearm, hands, and feet. The pleomorphic type affects the limbs (thigh).

Clinically, it presents with a painless mass of rapid growth and the rest of the signs and symptoms depend on the anatomical location.

Regarding its evolution and prognosis, some factors are favorable, such as the presentation in lactation or childhood, the location in the orbit or the urogenital system, a size <5 cm, and the complete initial resection without metastases in the lymph nodes or at a distance.

Treatment consists of preoperative chemotherapy and subsequent excision with resection of the regional lymph nodes. Postoperative chemotherapy and radiation therapy can be performed if the excision was not complete.

This patient received four cycles of preoperative chemotherapy and then underwent surgery (Figure 7).

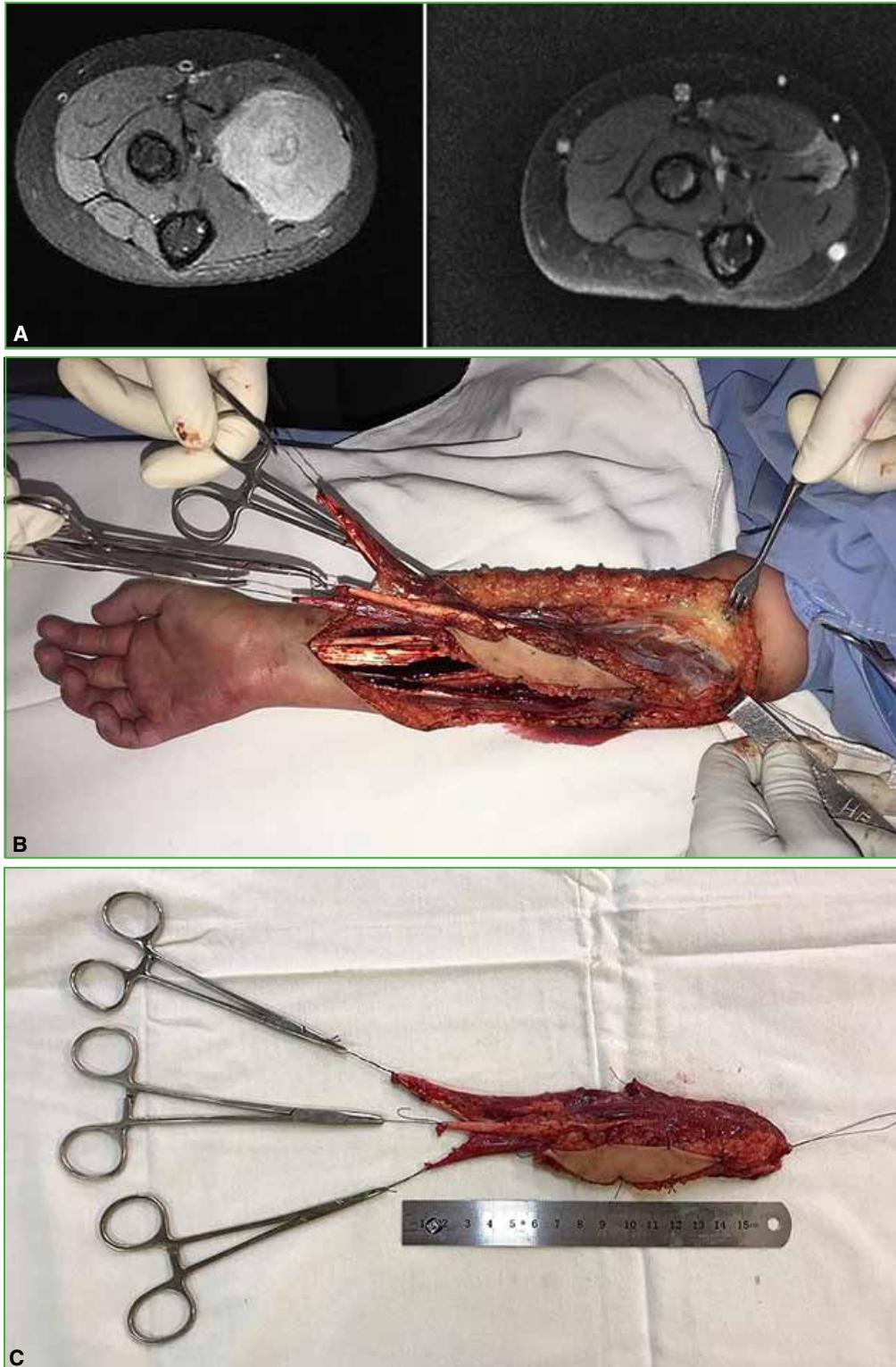


Figure 7. A. Axial MRI in comparative STIR sequence before chemotherapy (left) and after chemotherapy (right). B and C. Extensive tumor resection. Resection of epitrochlear, flexor digitorum superficialis, palmaris brevis, palmaris longus, flexor carpi ulnaris, and humeral belly of the pronator teres muscles with epineurotomy of the median and ulnar nerves under a microscope.

The most frequent differential diagnoses are other soft tissue sarcomas (fibrosarcoma or synovial sarcoma), soft tissue masses (ossifying myositis) (Figure 8), schwannoma (Figure 9), and lymphoma (Figure 10).

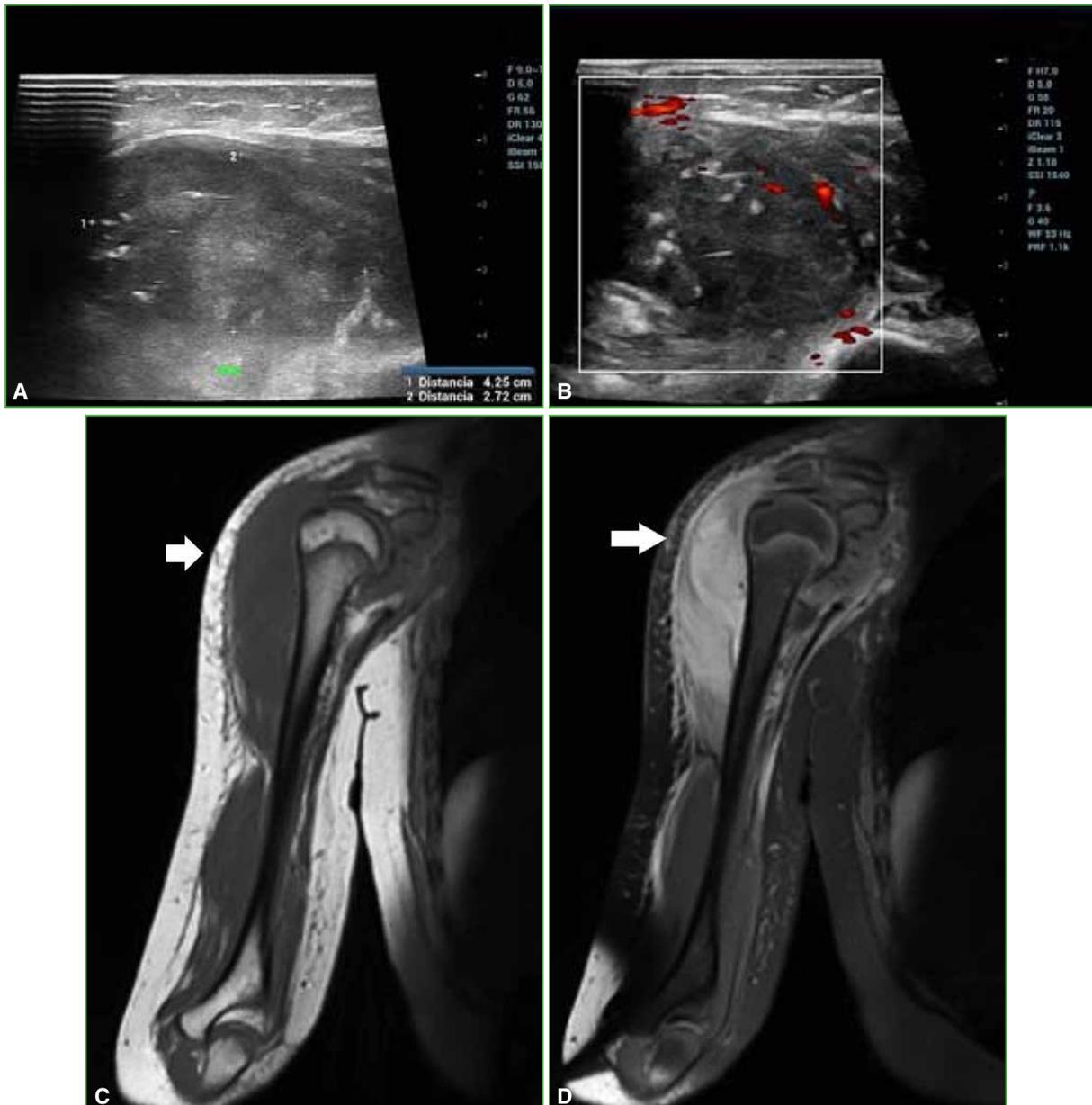


Figure 8. A 10-year-old patient with a painful tumor in the right shoulder of one month of evolution. **A.** Soft tissue ultrasound showing a hypoechoic tumor, with poorly defined edges, inside the muscular plane. **B.** Poor vascularization in the Doppler study. **C.** Coronal MRI in T1 sequence: increase in the size of the deltoid muscle (arrow). **D.** Coronal MRI in STIR sequence: diffuse lesion and significant edema of the rest of the deltoid (arrow). **E.** Axial MRI in T1 sequence: increase in the size of the deltoid without involvement of subcutaneous cell tissue (arrow).



Figure 8. **F.** Axial MRI in T2 sequence: better differentiation between injury and muscle edema (arrow). **G.** Axial MRI in STIR sequence: lesion inside the deltoid (arrow). **H.** Control radiograph 60 days after the original pain. A voluminous soft tissue calcification compatible with myositis ossificans is visualized.

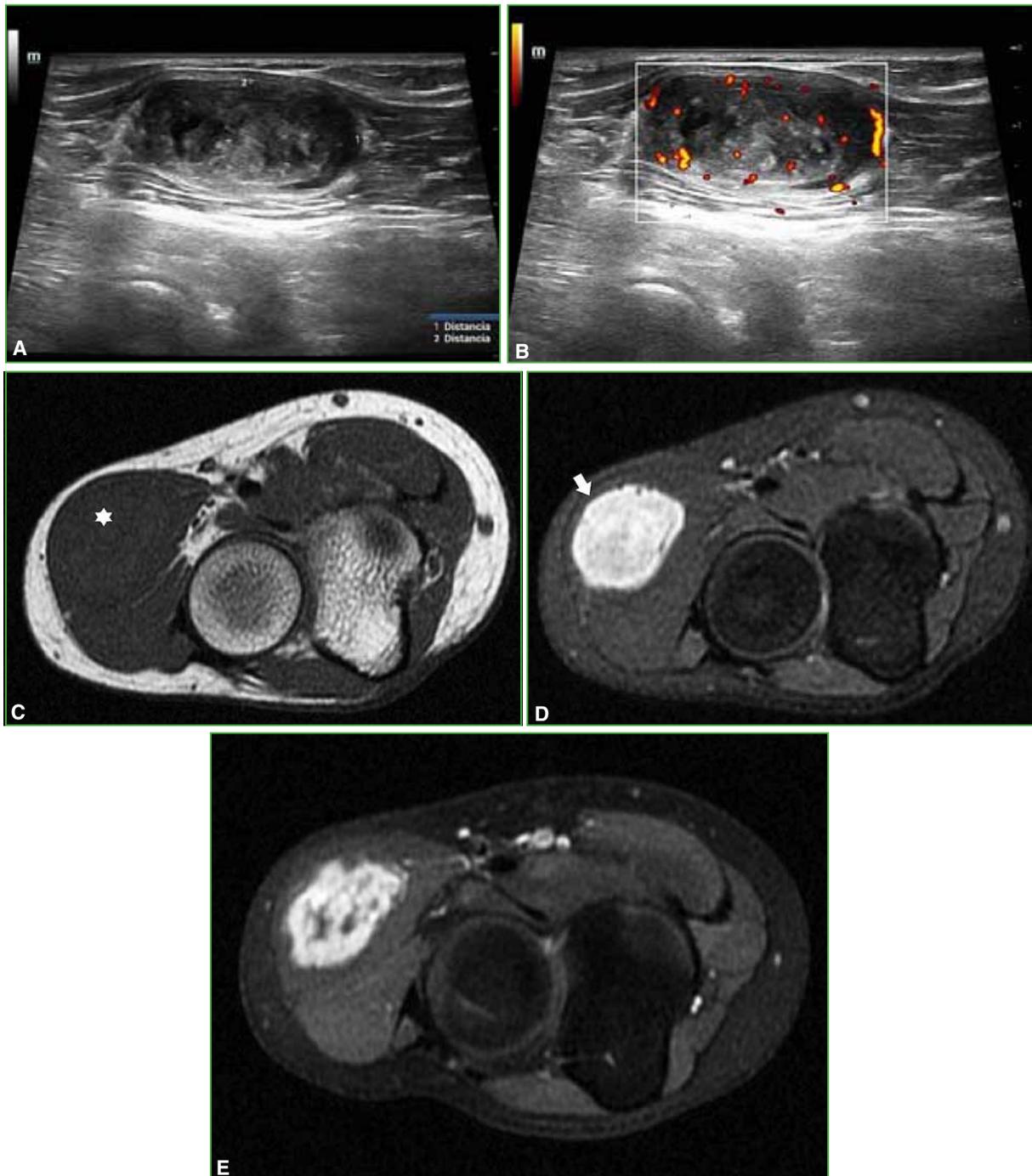


Figure 9. A 35-year-old patient with a painless tumor of years of evolution that has grown more in recent months. Upon physical examination, an elastic, mobile tumor is detected. **A.** Soft tissue ultrasound where a soft tissue mass with defined edges is observed inside the epicondylar muscles. **B.** Marked vascularization in the Doppler study. **C.** Axial MRI in T1 sequence: lesion in the muscle interior of the epicondylar group (asterisk). **D.** Axial MRI in STIR sequence: lesion of circumscribed edges (arrow). **E.** Axial MRI in T1 sequence with contrast injection and fat suppression (central necrosis impression). All of these findings are compatible with a schwannoma.



Figure 10. A 49-year-old patient with a tumor in the medial side of the forearm of months of evolution, painful on palpation. **A.** Soft tissue ultrasound showing a soft tissue mass with defined edges located in the subcutaneous cellular tissue. **B.** Marked vascularization in the Doppler study. **C and D.** Coronal MRI in STIR and T1 sequences. **E and F.** Axial MRI in STIR and T1 sequences. All of these findings were consistent with lymphoma.