

# Leiomyosarcoma of Extremity: Are Clinical Findings and Imaging Modalities Helpful For Differential Diagnosis? A Case Report

---

Mehmet Cengiz ÇOLAK<sup>\*</sup>, Hasan KOCATÜRK<sup>\*\*</sup>,  
Ednan BAYRAM<sup>\*\*</sup>, Hikmet KOÇAK<sup>\*\*\*</sup>, Cemal GÜNDOĞDU<sup>\*\*\*\*</sup>

## ÖZET

*Soft tissue sarcomas account for approximately 5-10% of all mesenchymal malignancies and can arise at any site throughout the body. A 31-year-old man admitted to our hospital for further evaluation of swelling in the posterior region of the right upper extremity. He had been good health until six weeks previously, when he developed a swelling in the right upper extremity after heavy exercise. Clinical findings and imaging modalities including Magnetic resonance imaging (MRI) were suggestive of a diagnosis of intramuscular deep-seated hemangioma. Right arm axillary angiogram revealed diffuse tumor vascularization. The tumor was removed surgically and interestingly leiomyosarcoma was detected upon pathologic examination. Despite the fact that MRI remains gold standard for noninvasive assessment of soft-tissue structures, the present case demonstrates that imaging studies, especially MRI, can not differentiate benign soft tissue structures from the malign ones.*

**Anahtar Kelimeler:** Hemangioma, diffusion magnetic resonance imaging, leiomyosarcoma.

---

<sup>\*</sup> Department of Cardiovascular Surgery, Medical Faculty, İnönü University, Malatya.

<sup>\*\*</sup> Department of Cardiology, Sifa Medical Center, Erzurum.

<sup>\*\*\*</sup> Department of Cardiovascular Surgery, Medical Faculty, Atatürk University, Erzurum.

<sup>\*\*\*\*</sup> Department of Pathology, Medical Faculty, Ataturk University, Erzurum.

## Ekstremitenin Leiomyosarkomu: Klinik Bulgular ve Görüntüleme Yaklaşımları Ayırıcı Teşhiste faydalı mıdır? Bir Olgu Sunumu

### ABSTRACT

*Yumuşak doku sarkomları bütün mezenkimal tümörlerin yaklaşık % 5-10'unu oluşturur ve vücudun herhangi bir bölgesinden gelişebilir. 31 yaşında erkek hasta sağ üst ekstremitenin arka bölgesindeki şişliğin daha ileri incelemesi için hastanemize başvurdu. 6 hafta öncesine kadar sağlıklı olan hastamızın, ağır bir egzersiz sonrası sağ üst ekstremitesinde bir şişliği gelişmişti. Klinik bulgular ve manyetik rezonans incelemesi derin yerleşimli kas içi hemanjiom olabileceğini düşündürdü. Sağ kol aksiler anjiyografisi yaygın damarsal kitle olduğunu gösterdi. Kitle cerrahi olarak çıkarıldı ve patolojik incelemede leiomyosarkom tanısı konuldu. Yumuşak doku kitlelerinin değerlendirilmesinde manyetik rezonans görüntülemesi altın standart olmasına rağmen şimdiki vakada olduğu gibi manyetik rezonans görüntülemesi kötü huylu yumuşak doku kitlelerini iyi huylu yumuşak doku kitlelerinden ayıramayabilir.*

**Key Words:** Hemanjiom, difüzyon manyetik rezonans görüntüleme, leiomyosarkom.

### INTRODUCTION

Soft tissue sarcomas account for approximately 5-10% of all mesenchymal malignancies (Mann et al., 2006) and can arise at any site throughout the body. Almost 45% of all soft tissue sarcomas are located in the extremities, especially in the lower limb (Gomez ve Morcuende, 2004). Leiomyosarcoma represents approximately 7% of all soft tissue sarcomas and are primarily a disease of middle-aged persons, presenting between the fifth and seventh decades of life (Gomez ve Morcuende, 2004). Sarcomas are typically very vascular, with large intra- and extra-tumoral blood vessels (Chu et al., 2003).

Hemangiomas are common neoplasms that account for 7% of all benign soft-tissue tumors. Most patients with deep-seated hemangiomas present with pain or intermittent swelling. Imaging studies or biopsy is required in some deep-seated hemangioma cases to distinguish malignant soft tissue tumors (Teo et al., 2000).

There are some instances in which the patient presents after moderate trauma to the extremity masking the underlying tumor on imaging studies (Suh et al., 1994). We describe a case of right upper extremity leiomyosarcoma after preoperative misdiagnosis of hemangioma on the basis of clinical findings and imaging modalities.

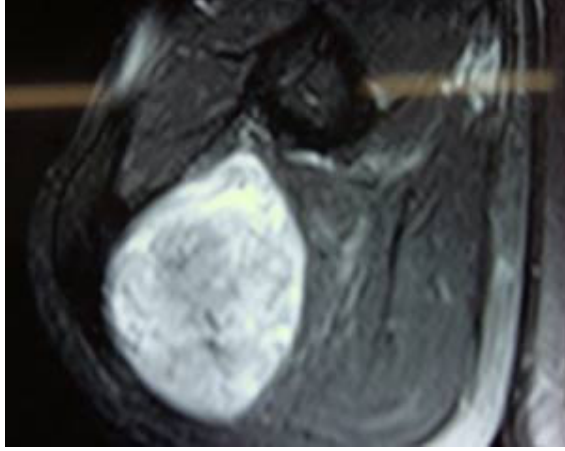
## **CASE PRESENTATION**

A 31-year-old man admitted to our hospital for further evaluation of swelling in the posterior region of the right upper extremity. He had been good health until six weeks previously, when he developed a swelling in the right upper extremity after heavy exercise. He had been evaluated previously elsewhere and was diagnosed as having traumatic soft tissue injury occurring after exercise. A course of analgesic was prescribed however; he had been seen regularly by a physician regularly. About one week before the current admission he experienced recurrent pain by movements in the right arm. His medical and family history was unremarkable. Physical examination revealed a firm, tender and slightly mobile mass of 4 x 5 x 6.5 cm size with smooth borders in the posterior region of the right upper extremity (Figure 1).



**Fig 1.** A swelling in the arm

There was no fluctuance, erythema, ecchymosis, increased warmth, or lymphadenopathy. The elbow and shoulder had full range of motion. Distal pulses were equal in both upper extremities. Findings on the rest of the physical examination were normal. Laboratory and plain radiographs data yielded normal results. Ultrasound (USG) examination revealed a heterogeneous mass of mixed echo texture with smooth borders and vascularity with solid character suggestive of an intramuscular hemangioma. Magnetic Resonance Imaging (MRI) demonstrated a cylindrically shaped mass of 37x 48 x 65 mm size within the right triceps muscle without bone involvement (Figure 2).



**Fig 2.** MRI revealed a cylindrically shaped, no bony involvement, and 37 x 48 x 65 mm mass within the right triceps muscle.

The density of the mass was slightly lower than that of the muscles, with a low signal in T1, and a high signal in T2. The radiologists interpreted the findings of USG and MRI as a hemangioma. Angiogram consistently showed evidence of vessels supplying the lesion (Figure 3).



**Fig 3.** Angiogram showed evidence of vessels supplying the lesion.

Although preoperative diagnosis was consistent with hemangioma, it would be appropriate to suspect a soft tissue sarcoma in the presence of a deep seated and considerably large mass. Therefore, the wide excision was done under general anesthesia (Figure 4,5).



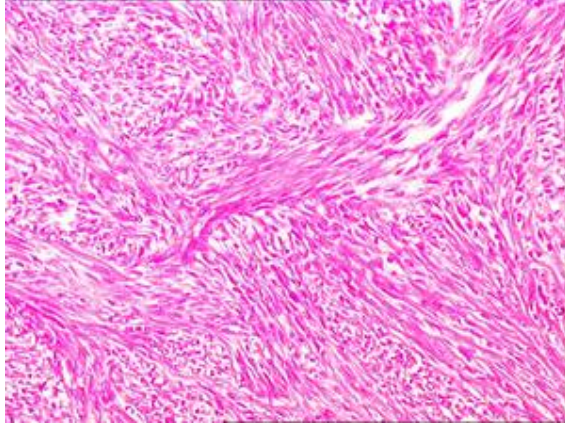
**Fig 4.** Intraoperative appearance of the mass.



**Fig 5.** Macroscopic appearance of the mass after it had been cut open.

Histopathologically, the tumor consisted of spindled cells with abundant, fibrillary, eosinophilic cytoplasm, symmetric, “cigar”-shaped nuclei and atypical nuclear features. Mitosis was counted in-between 10 and 15 cellular and mitotically active sites per 10 high power field (HPF). Immunohistochemistry findings revealed positivity for vimentin, smooth muscle actin and desmin; primary antibodies for markers such as S100, HMB 45, EMA were negative. Resection margins were free of tumor.

Histopathological examinations disclosed the diagnosis of leiomyosarcoma in accordance with preoperative suspicion (Figure 6). The patient’s postoperative course was uneventful and referred to oncologists for further therapy.



**Fig 6.** Intersecting bundles of smooth muscle fibers in longitudinal and cross-sections.(H&E x100).

## **DISCUSSION**

Differentiating between malignant and benign soft tissue lesions has proven to be a difficult task even with the advantage of MRI. MRI remains the gold standard for evaluation of most soft-tissues lesions. However, the sensitivity for diagnosis and grading remains controversial in the literature. MRI is not able to predict malignancy, and the findings commonly associated with malignant lesions frequently overlap with those seen in benign tumors (Gomez ve Morcuende, 2004). MRI also performs poorly in the histological classification of soft-tissue tumors (Kransdorf, 1995). This is because MRI

provides only indirect information about tumor histology by showing signal intensities related to some physicochemical properties of the tumor components and consequently reflect gross morphology of the lesion rather than underlying histology. The time-dependent changes of the tumors (as a consequence of intratumoral necrosis and/or bleeding), makes the differentiation process even more difficult. Soft-tissue tumors grow in a centrifugal manner until resistance is met. The barriers in soft tissues consist of major fibrous septa and the origins and insertions of muscles. Growth tends to occur in the plane of least resistance. The host responds to tumor growth by creating a reactive fibrovascular tissue that forms a limiting capsule in benign lesions (Teo et al., 2000). High-grade malignant soft-tissue tumors may show low to intermediate signal intensity on T2-weighted images because of an increased nucleocytoplasmic index and an altered cellular and interstitial components proportion, both resulting in a decrease of intra and extra-cellular water (De Schepper, 1997). Hermann et al. reported that changes in homogeneity (from homogeneous on T1-weighted images to heterogeneous on T2-weighted images) and the presence of low-signal intra-tumoral septations have a sensitivity of 72 and 80 percent and a specificity of 87 and 91 percent, respectively, in predicting malignancy (Lewis, 1992). T1-weighted images show no reliable distinguishing features. Both hemangiomas and soft-tissue tumors are low in signal intensity. The frequency of lobulation, septation, and central low-intensity dots on T2-weighted images are much higher for hemangiomas than for malignant soft-tissue tumors (Suh et al., 1994).

Other signs related to malignancy include the presence of tumor necrosis, bone or neurovascular involvement, mean diameter of more than 66 millimeters, and irregular or partially irregular margins (Gomez ve Morcuende, 2004).

Angiographic studies also show numerous enlarged vessels, arterio-venous shunt, and delayed washout of contrast medium from the tumor (Suh et al., 2000). The presence of a large soft tissue mass associated with large peritumoral vessels is strongly suggestive of sarcoma.

Hemangiomas constitute 7% of all benign tumors, and are most commonly superficial and easy to diagnose clinically. However, deep-seated hemangiomas such as intramuscular hemangiomas are relatively uncommon (Engelstad et al., 1980) and often pose diagnostic difficulties. Age of occurrence of intramuscular hemangioma is most common in the third and fourth decades, but they occasionally may present earlier (Kim et al., 1999). Although

intramuscular hemangioma has a wide anatomical distribution, it usually occurs in the extremities and all patients present with a growing palpable mass, which may or may not be accompanied by pain or may present with pain and without a mass (Kim et al., 1999). For the diagnosis of intramuscular hemangioma, plain radiographs, MRI, angiography, and positron emission tomography (PET) may be helpful. Plain radiographs show phleboliths, particularly associated with a soft-tissue mass and cortical or periosteal reaction adjacent to a soft tissue hemangioma (Sung et al., 1998). MRI is useful in defining the location and extent of intramuscular hemangiomas and is now routinely used to differentiate intramuscular hemangiomas. Angiography of a peripheral hemangioma is especially useful for evaluating the extent and degree of vascularity, and vascular supply. <sup>18</sup>F-fluoro-2-deoxy-D-glucose (FDG)- PET and fluorine-18 alpha-methyltryosine (FMT)-PET may be useful in the diagnostic workup of hemangioma as well as for the differentiation from malignancy (Hatayama et al., 2003).

Hemangioma has slightly hyperintense signal intensity on T1-W images as compared to muscle and is hyperintense on T2-W images, also. There is moderate to strong diffuse enhancement with intravenous contrast medium. Curvilinear flow voids, representing blood vessels, are typically present within the mass. The presence of intra-tumoral flow voids is regarded as useful in differentiating a hemangioma from a more aggressive lesion. Tumor does not usually contain large vessels (Chu et al., 2003). However, our case shows that, similar radiological features are present in sarcoma, which makes it indistinguishable from a benign hemangioma by both USG and MRI.

## **CONCLUSION**

Clinical suspicion should rise when imaging studies suggest hemangiomas in cases where atypical age of presentation, smooth borders, evidence of vessels supplying the lesion, large mean diameter and unusual clinical course are present.

### **Abbreviations**

USG, ultrasound; MRI, magnetic resonance imaging; HPF, high power field; FDG, fluoro-deoxy-D-glucose; FMT, fluorine-alpha-methyltryosine.

### **Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### **Competing interests**

The authors declare that they have no competing interests.

### **Authors' contributions**

MCC is the corresponding author and responsible for the surgery team. CG performed the histological examination of the mass. The others are responsible for the editing of the text and data collector.

### **REFERENCES**

- Chu, W.C., Howard, R.G., Roebuck, D.J., Chik, K.W., Li, C.K. (2003). "Periorbital alveolar soft part sarcoma with radiologic features mimicking hemangioma", **Med Pediatr Oncol.**, 41:145-146.
- De Schepper, A. (1997). "Grading and characterization of soft tissue tumors. Imaging of soft tissue tumors", (ed), De Schepper A, Berlin Heidelberg New York: Springer, 127-139.
- Engelstad, B.L., Gilula, L.A., Kyriakos, M. (1980). "Ossified skeletal muscle hemangioma: radiologic and pathologic features", **Skeletal Radiol**, 5: 35-40.
- Gomez, P. (2004). "Morcuende J. High-grade sarcomas mimicking traumatic intramuscular hematomas: a report of three cases", **Iowa Orthop J.**, 24:106-110.
- Hatayama, K., Watanabe, H., Ahmed, A.R., Yanagawa, T., Shinozaki, T., Oriuchi, N., et al. (2003). "Evaluation of hemangioma by positron emission tomography: role in a multimodality approach", **J Comput Assist Tomogr**, 27: 70-77.
- Kim, E.Y., Ahn, J.M., Yoon, H.K., Suh, Y.L., Do, Y.S., Kim, S.H., et al. (1999). "Intramuscular vascular malformations of an extremity: findings on MR imaging and pathologic correlation", **Skeletal Radiol**, 28: 515-21.

Kransdorf, M. (1995). "Malignant soft tissue tumors in a large referral population distribution of specific diagnoses by age, sex and location", **Am J Roentgenol**, 164:129-134.

Lewis, M. (1992). "Tumour and tumour-like conditions of the soft tissue: magnetic resonance imaging features differentiating benign from malignant masses", **Br J Radiol**, 65: 14-20.

Mann, H.A., Hilton, A., Goddard, N.J., Smith, M.A., Holloway, B., Lee, C.A. (2006). "Synovial sarcoma mimicking haemophilic pseudotumour", **Sarcoma**, 2006: 27212.

Suh, J.S., Cho, J., Lee, S.H., Shin, K.H., Yang, W.I., Lee, J.H., et al. (2000). "Alveolar soft part sarcoma: MR and angiographic findings", **Skeletal Radiol**, 29: 680-689.

Suh JS, Hwang G, Hahn SB. (1994). "Soft-tissue hemangiomas: MR manifestations in 23 patients", **Skeletal Radiol**, 23: 621-625

Sung, M.S., Kang, H.S., Lee, H.G. (1998). "Regional bone changes in deep soft tissue hemangiomas: radiographic and MR features", **Skeletal Radiol**, 27: 205-210.

Teo, E.L., Strouse, P.J., Hernandez, R.J. (2000). "MR imaging differentiation of soft-tissue hemangiomas from malignant soft-tissue masses", **AJR Am J Roentgenol.**, 174:1623-1628.