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CLINICAL IMAGING

Case report

MRI of epithelioid sarcoma of the thigh

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Abstract

A 31-year-old woman with epithelioid sarcoma of the thigh is presented. The patient had had a progressively growing mass in her left thigh for 4 months. Magnetic resonance imaging (MRI) showed a subcutaneous mass with a central area of hypointensity and serpiginous hyperintensity in the peripheral portion. Such findings have not been reported before. Histologic findings were compatible with epithelioid sarcoma.

Keywords: Epithelioid sarcoma; MRI; Soft-tissue sarcoma

1. Introduction

Epithelioid sarcoma is a rare soft tissue tumor that commonly occurs in the distal portion of the upper extremity. Although magnetic resonance imaging (MRI) findings are nonspecific, epithelioid sarcoma commonly shows a multinodular configuration [1]. Epithelioid sarcoma with unusual radiological findings has also been reported [2–4]. We present a patient with epithelioid sarcoma in the thigh. The tumor showed a central area of hypointensity with serpiginous peripheral hyperintensity on MRI images; the findings have not been reported before.

2. Case report

A 31-year-old woman complained of a progressively growing mass over the upper portion of her left thigh for 4 months. Occasional sharp pain over the mass area was noted. The patient had no history of trauma. On physical examination, the tumor mass was hard, with induration, and measured 10 cm in diameter. Bloody discharge at drainage was noted.

MRI (1.5-T, Horizon LX, General Electric, WI) revealed an 11×6×5-cm subcutaneous mass in the upper lateral portion of left thigh (Fig. 1A–D). The tumor showed a central area of hypointensity with serpiginous peripheral hyperintensity on short tau inversion recovery (STIR) and gadolinium-enhanced T1-weighted images. No extension to surrounding muscles was noted.

The patient received radical resection of the tumor. At surgery, the tumor had an unclear margin and a necrotic and multinodular consistency. Gross specimen revealed a tumor with central hemorrhage and necrosis. Histologically, the tumor mass was composed of ephithelioid cells intermingled with spindle cells in vaguely nodular and diffuse patterns (Fig. 1E). The epithelioid cells were round to oval in shape with abundant eosinophilic cytoplasms. Extensive necrosis and hemorrhage were noted within the tumor (Fig. 1F). In the periphery, increased blood vessels were noted (Fig. 1G). The sarcoma was mainly located in the subcutis and deep soft tissue. The tumor cells showed moderate to marked nuclear pleomorphism and frequent mitoses (up to 5/10 HPF). There were focal hyalinization, spotty calcifications,

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and pseudovascular space formation. No angiolymphatic embolus was seen. Immunohistochemically, the ephithelioid cells and spindle cells were positive for cytokeratin and vimentin. Histological findings were compatible with epithelioid sarcoma.

3. Discussion

Epithelioid sarcoma occurs most frequently in young patients; 74% are in the 10- to 49-year age groups [5]. There is a slight male predominance (male to female = 1.8−2.0 to 1)

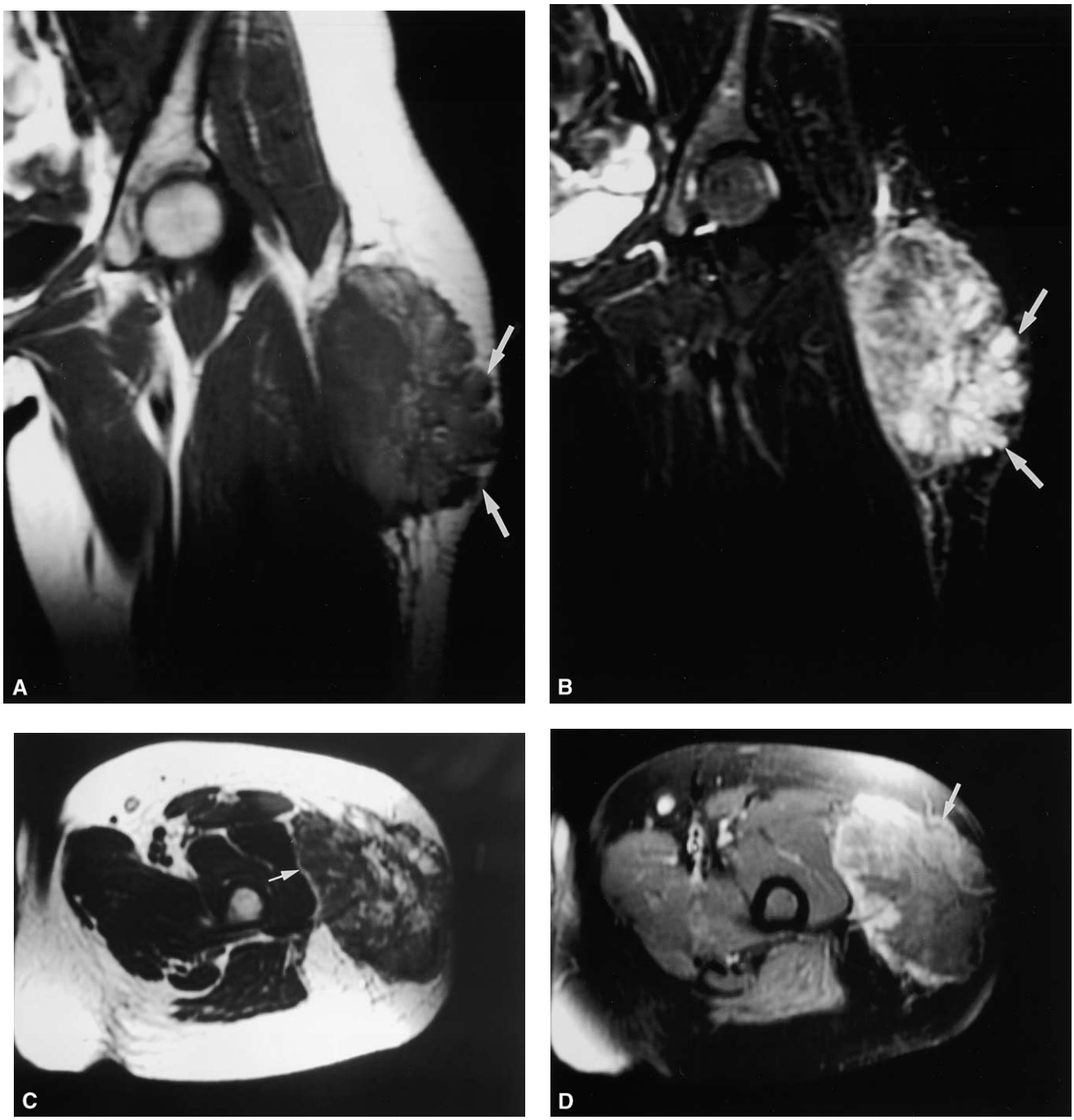

Fig. 1. (A) Coronal T1-weighted (TR/TE = 450/9.5 ms) image shows a subcutaneous soft tissue mass in the upper lateral potion of the left thigh. The tumor has a multinodular pattern (arrows) at its lateral lower margin. The adjacent muscle is slightly compressed by the tumor. (B) On coronal STIR (TR/TE/TI = 5000/46/160 ms) image, the multinodular portion of the tumor has a serpiginous hyperintense signal pattern (arrows), which is consistent with vascular formation on histology. (C) Axial T2-weighted (TR/TE = 4000/106 ms) image shows a fat septum (arrow) separating the tumor from muscle. (D) Gadolinium-enhanced T1-weighted (TR/TE = 666/12.1 ms) image shows peripheral enhancement with a central area of isointensity. The central area of the tumor is consistent with hemorrhage and necrosis on histology. (E) The sarcoma is composed of epithelioid cells intermingled with spindle cells (arrow) (H&E ×200). (F) Necrosis and hemorrhage are noted in the sarcoma (H&E ×100). (G) Increased blood vessels are noted in the peripheral area of the tumor (H&E ×100).

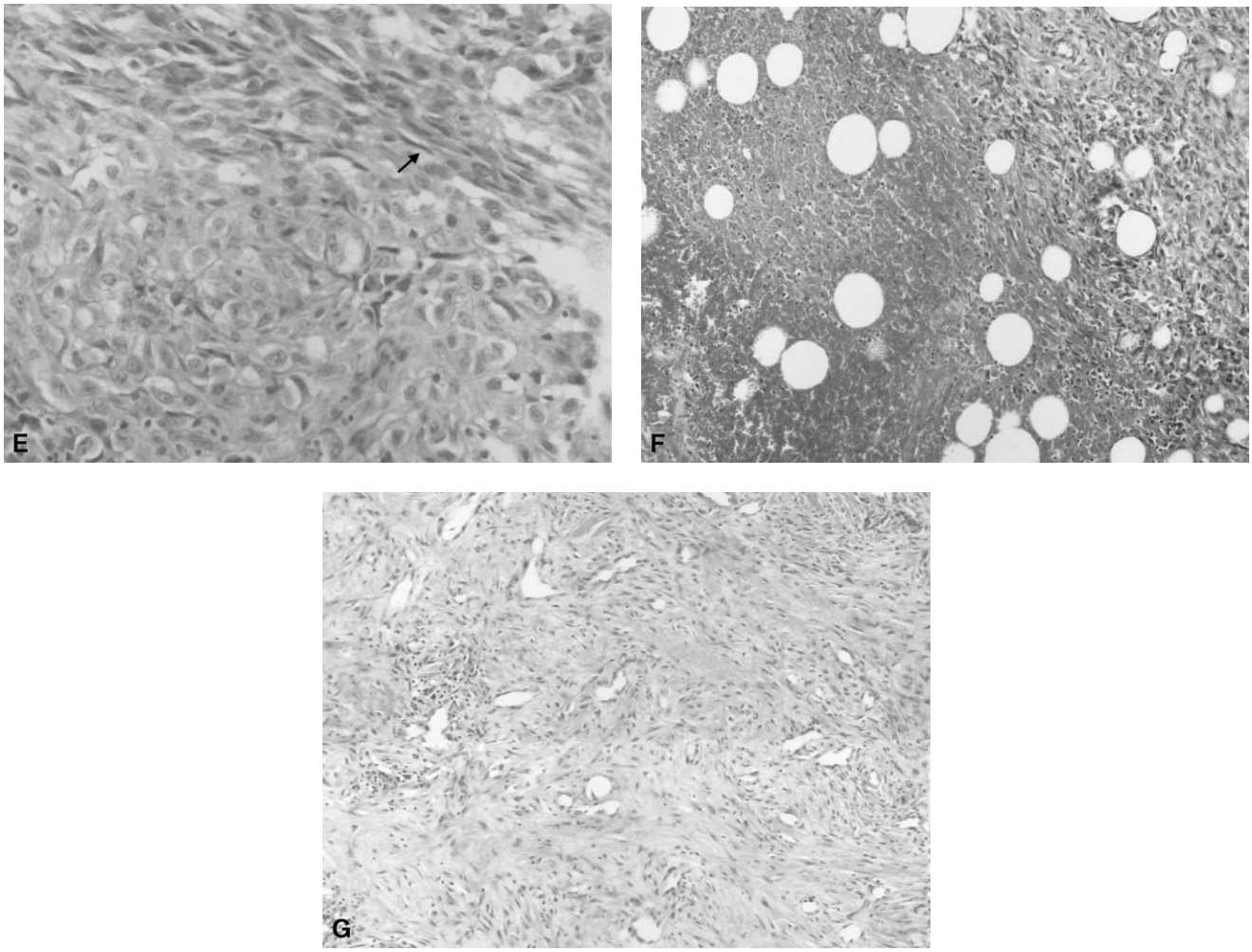

Fig. 1. (*continued*)

[5,6]. The tumor has a predilection for the upper extremities, especially the forearms and hand, accounting for 58% to 60% of patients [6,7]. The second most frequent site is the lower extremities, accounting for 25% to 32% [6,7]. In the proximal and distal lower extremities, the frequency is 12% and 15%, respectively [8].

A distinction between distal and proximal types may be important [5,9], as different locations show various clinical and histological features; the proximal-type tumors can have a more malignant potential and poor prognosis. Of the proximal-type epithelioid sarcoma, 94% occur in the chest wall, inguinal region, thigh and perineum [1]. The tumor size at presentation varies from 0.5 to 19 cm [1,3,6,10,11], and growth duration ranges from 6 months to 5 years [3,4]. Our patient had a 4-month history of a progressively growing mass over the left thigh, which could be considered as a proximal type.

Epithelioid sarcoma has a multifaceted nature, with the tumors at one extreme being small, subcutaneous, discrete and homogeneous and those at the other being large, deep, infiltrative and heterogeneous in appearance [11]. The tumor can be nonhemorrhagic (83%) or hemorrhagic (17%) [7]. Atypical presentations include intra-articular tumors [10], perineural invasion with denervation of muscle atrophy, a subcutaneous honeycomb pattern simulating lymphedema [3], a muscular nodule with no apparent soft tissue mass [2] and an abscess with hemorrhagic fluid [4]. Bone metastasis is rare; only isolated cases have been reported [2,3,5].

Histologically, varying degrees of cellularity, necrosis, fibrosis, hyalinization and surrounding inflammation have been seen [11]. Calcification occurs in 6% to 19% of cases [1,7,12], and is often associated with tumor degeneration. The histological picture of a proximal-type tumor is composed mainly of sheets of large or spindle cells with multinodularity [1].

In our patient, the tumor had a central area of hypointensity with serpiginous peripheral hyperintensity; these findings have not been reported before. The central zone of the tumor consisted of hemorrhage and necrosis, as proved by our histology. However, the spotty calcifications within the tumor noted on histology were not detected on MRI. The peripheral serpiginous appearance may be

attributed to the vascular formation of the sarcoma, as noted on our histology.

In summary, MRI plays an important role in identifying a subcutaneous soft tissue tumor with intratumorous hemorrhage and necrosis; epithelioid sarcoma should be included in the differential diagnosis.

References

[1] Tateishi U, Hasegawa T, Kusumoto M, Yokoyama R, Noriyuki M. Radiologic manifestations of proximal-type epithelioid sarcoma of the soft tissue. AJR Am J Roentgenol 2002;179:973–7.

[2] Nakashima H, Katagiri H, Sugiura H, Yonekawa M, Nishida Y, Yamada Y. Epithelioid sarcoma mimicking a primary osseous multifocal scapula lesion. Skeletal Radiol 2002;31:430–3.

[3] Yamato M, Nishimura G, Yamaguchi T, Tamai K, Saotome K. Epithelioid sarcoma with unusual radiological findings. Skeletal Radiol 1997;26:606–10.

[4] Dion E, Forest M, Brasseur J, Amoura Z, Grenier P. Epithelioid sarcoma mimicking abscess: review of the MRI appearances. Skeletal Radiol 2001;30:173–7.

[5] Chase DR, Enzinger FM. Epithelioid sarcoma. Diagnosis, prognostic indicators, and treatment. Am J Surg Pathol 1985;9:241–63.

[6] Enzinger FM, Weiss SW. Soft tissue tumors. St. Louis (MO): CV Mosby, 1983.

[7] Romero JA, Kim EE, Moral IS. MR characteristics of epithelioid sarcoma. J Comput Assist Tomogr 1994;18:929–31.

[8] Weiss SW, Glodblum JR. Enzinger and Weiss's soft tissue tumors. 4th ed. St. Louis (MO): Mosby, 2001.

[9] Miettinen M, Fanburg-Smith JC, Virolainen M, Shmookler BM, Fetsch JF. Epithelioid sarcoma: an immunohistochemical analysis of 112 classical and variant cases and a discussion of the differential diagnosis. Hum Pathol 1999;30:934–94.

[10] Hurtado RM, McCarthy E, Frassica F, Holt PA. Intraarticular epithelioid sarcoma. Skeletal Radiol 1998;27:453–6.

[11] Hanna SL, Kaste S, Jenkins JJ, Hewan-Lowe K, Spence JV, Gupta M, Monson D, Fletcher BD. Epithelioid sarcoma: clinical, MR imaging and pathologic findings. Skeletal Radiol 2002;31:400–12.

[12] Lo HH, Kalisher L, Faix JD. Epithelioid sarcoma: radiologic and pathologic manifestations. AJR Am J Roentgenol 1977;128:1017–20.