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Case Report

An Undifferentiated Pleomorphic Sarcoma with Massive Intratumoral Hematoma: A Case Report and Literature Review

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Abstract

We here report a case of undifferentiated pleomorphic sarcoma (formerly malignant fibrous histiocytoma) arising in the femoral triangle where the tumor expanded by intratumoral hemorrhage and mimicked an intramuscular hematoma. T1- and T2-weighted magnetic resonance images showed a cystic mass containing septa and multiple nodules of heterogeneous intensity in the adductor muscle of the right thigh. Excisional biopsy indicated the tumor was an undifferentiated pleomorphic sarcoma. Following *en-bloc* resection, the patient has been free from recurrence or metastasis for 7 years. We discuss the salient features of this case and recent reports on cystic soft tissue tumors.

INTRODUCTION

Undifferentiated pleomorphic sarcoma (UPS) is a soft tissue sarcoma composed of undifferentiated mesenchymal tumor cells and is now regarded as distinct entity following its reclassification from malignant fibrous histiocytoma (MFH). MFH had been considered the most common soft tissue sarcoma of mid and late adulthood. The tumor most frequently arises from the deep fascia or skeletal muscle in an extremity. Occasionally, it exhibits cyst formation, mimicking a hematoma [1], which can lead to a misdiagnosis or delay in diagnosis. Several reports have appeared on large cystic MFH in the abdominal cavity [2, 3] or retroperitoneum that showed rapid hemorrhagic enlargement induced by chemotherapy [4]. We describe here a case with a massive cystic UPS in the femoral triangle where expansion of the tumor, which was 25 cm in longitudinal diameter at initial presentation, was thought to be caused by internal hemorrhage.

CASE REPORT

A 53-year-old man presented with a mass in the right femoral triangle that had grown rapidly over the last 3 months and had not been preceded by trauma. He had been treated for mental illness since the age of 31. Physical examination revealed a strained bulging mass in the right femoral triangle (Figure 1). Despite mild rubor and tenderness over the mass, he did not complain of pain on palpation. Magnetic resonance (MR) imaging revealed an expansive cystic lesion in the adductor muscles of his right thigh, measuring 25.0 × 19.0 × 18.0 cm (Figure 2). T2-weighted images showed a high signal intensity lesion, T1-weighted images showed a relatively high intensity lesion, and both sets of images indicated the lesion contained nodules and septa that had equal signal intensity to muscle. The lesion extended to the surrounding muscles outside of the adductor compartment, contacted the obturator foramen and the lesser trochanter, and was close to the hip joint capsule, femoral artery, and sciatic nerve (Figure 2). Similarly, computed tomography showed the same sized low density area surrounded by tissue

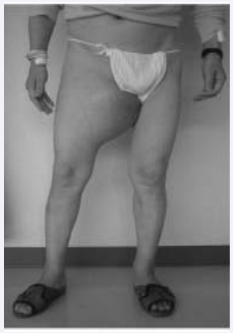

Figure 1 Appearance of patient's legs with an expansive lesion in the right femoral triangle.

with the same density constitution as muscle. Angiography of the femoral artery showed eggshell-shaped staining of the lesion with moderate hypervascularity.

Excisional biopsy from the cyst wall caused the tumor to bleed. Pathological examination showed pleomorphic cells scattered between regions of coagulative necrosis. The patient underwent *en bloc* resection of the tumor without adjuvant therapy. All branches of the profunda femoris artery and great saphenous vein were sacrificed, as were all adductor muscles and the vastus medialis, gracilis, semimembranosus, rotators of the hip joint, and a part of the iliopsoas. The tumor contained hemorrhage and necrotic tissue, with some hemosiderosis. The cyst wall was coated extensively by relatively bland pleomorphic cells with scattered histiocytes, fibroblastic spindle cells, and inflammatory cells and the wall itself contained tumor cells (Figure 3). Proliferation of some capillary vessels was evident. The tumor cells were negative on immunochemical staining for antitrypsin, antichymotrypsin, lysozyme, c-kit, and platelet-derived growth factor. The pathological diagnosis of UPS (formerly known as MFH) was made based on these features. The patient has had no recurrence of the tumor or evidence of metastasis in the 7 years he has been followed and is ambulant with the aid of a cane.

DISCUSSION

MFH had been recognized as the most common soft tissue sarcoma in mid and late adulthood. However, with its reclassification in 2002 by WHO, it is no longer regarded as a distinct diagnostic category; for instance, myxoid and angiomatoid MFH have been re-classified into myxofibrosarcoma and angiomatoid fibrous histiocytoma (AFH), respectively. A diagnosis of UPS is appropriate when no line of differentiation is identified. While it remains controversial, the term MFH is still used by some pathologists and clinicians. Recently, Matushansky reported that MFH is associated with mesenchymal stem cells [5], which may be the cells of origin. Radiologic findings of MFH are

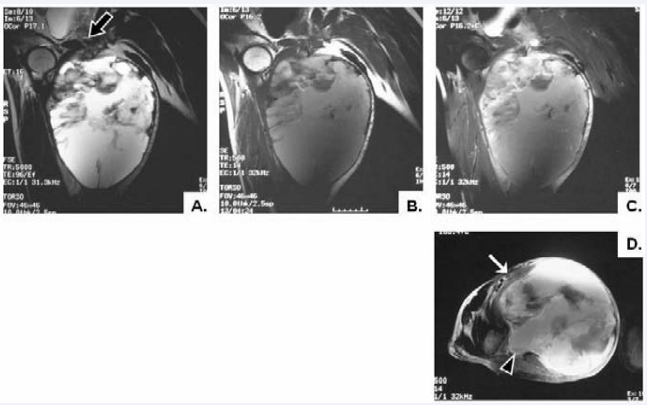

Figure 2 Coronal MR images showed a massive lesion with high signal intensity on T2-weighted imaging (A), faintly high intensity on T1-weighted imaging (B), and partially peripheral enhancement on fat-suppressed T1-weighted imaging after gadolinium injection (C). The mass contained nodules and septa indicating a loculated tumor and it contacted the obturator foramen (thick arrow). D. Axial, fat-suppressed T1-weighted image demonstrated the lesion contacted the lesser trochanter and was close to the femoral artery (thin arrow) and sciatic nerve (arrowhead).

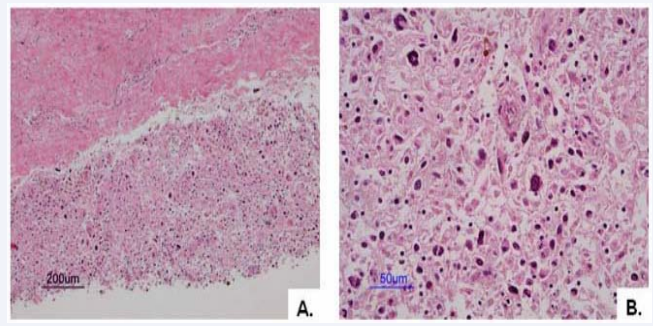

Figure 3 Photomicrographs of the tumor. A. Relatively sparse pleomorphic cells extensively coated the inner surface of the pseudocapsule, which consisted of fibrous elements (hematoxylin-eosin stain; scale bar: 200 μm). B. The pleomorphic area contained inflammatory elements without any characteristic arrangement (hematoxylin-eosin stain; scale bar: 50 μm).

not specific because of its varied morphologic features. Owing to the infrequency of (~5%) extensive cyst formation[1], there is little information about cystic MFH. To our knowledge, there have been no reports of massive cystic MFH in the extremities but three cases in the abdominal cavity or retroperitoneum [2,3]. The incidence of MFH in general arising in the abdominal cavity and retroperitoneum is relatively uncommon (~16%) compared to that originating in the extremities [1]. If MFH occurs in the abdominal cavity or retroperitoneum, it may manifest late and the tumor may be large on initial presentation. It has been reported that the size of the tumor correlates with the incidence of metastasis [1]. However, although both tumor size and anatomic location are significant for the clinical course of MFH in general, such correlations might not apply to cases of cystic MFH.

The differential diagnosis for soft tissue tumor with cyst formation includes synovial sarcoma [6,7], AFH, malignant tumor containing necrosis, schwannoma, and chronic expanding hematoma. Synovial sarcoma occurs near large joints in young adults. Nakanishi et al. reported that cyst formation in synovial sarcoma was related to the histological grade [8]. However, there is no information regarding the relationship between cyst formation and clinicopathologic features in other soft tissue sarcomas. Features of the cyst in the present case included inflammatory infiltrates, a thick fibrous pseudocapsule, and wide diffusion of tumor cells, indicating the tumor might be an AFH, which was first described by Enzinger as angiomatoid MFH [9]. AFH has been regarded as a tumor of childhood and early adult life with intermediate malignant potential, separated from conventional MFH by its relatively benign behavior [10-12]. Fanburg-Smith et al. reported that patients with AFH range up to 71 years, despite a childhood predominance [10]. The current case must differentiaed carefully UPS from AFH.

There are several reports that describe soft tissue sarcoma mimicking hematoma [13,14]. In those cases, the lesions were misdiagnosed as benign, such as traumatic hematoma, which led to poor clinical outcomes. In a report of six cases where sarcoma was misdiagnosed as hematoma, Imaizumi et al. concluded that meticulous evaluation of MR images contrasted with clinical histories including traumas was important for the differentiation [15]. Gomez et al., however, reported three cases of high-grade

sarcoma following trauma and concluded that MR imaging was neither sensitive nor specific enough to rule out malignancy [16]. Reid et al. reported some cases of hematoma that gradually enlarged and were difficult to differentiate between soft tissue tumor and chronic expanding hematoma [17]. In the present case, no traumatic history was identified and the lesion presented as a heterogeneous multi-locular mass with heterogeneous enhancement after gadolinium injection, all of which indicated malignancy.

MR images undoubtedly provide valuable information on soft tissue tumors, and several soft tissue lesions can be diagnosed from MR images alone. However, MR findings of many malignant lesions frequently overlap with those of benign ones. In addition, time-dependent changes such as necrosis and bleeding may complicate the differentiation. Thus far, several attempts at differentiating benign soft tissue lesions from malignant ones by means of ultrasound or 18F-fludeoxyglucose positron emission tomography have proven unsuccessful [18].

The significance of excisional biopsy for malignant soft tissue lesions has been reported [19-22]. In the present case, a percutaneous biopsy under image guidance may have been helpful. Aspiration biopsy is less traumatic for sampling both superficial and deep-seated mass lesions, but in certain cystic sarcomas, it would be difficult to obtain accurate samples. Imaizumi et al. showed that no malignant cells were obtained by aspiration biopsy for 5 patients among 6 with sarcomas mimicking hematoma [15]. Thus, excisional biopsy appears to be necessary for differentiating soft tissue sarcoma from expanding hematoma.

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