CASE REPORT

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Myxoinflammatory fibroblastic sarcoma: a case report

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Myxoinflammatory fibroblastic sarcoma (MIFS) is a recently described, rare low-grade sarcoma. Generally located in the upper and lower extremities, MIFS clinically mimics a benign cystic mass and is composed of spindle-like or atypical cells and mixed inflammatory infiltrates located in the fibroblastic myxoid stroma. Radiologic images and macroscopic appearance generally resemble a lobulated mass with irregular margins. We present a case of a tumoral mass with neoplastic cells at the center and a smooth surface with a previously undefined appearance. Myxoinflammatory fibroblastic sarcoma is significantly difficult to distinguish clinically from benign lesions and the surgeon should consider the possibility of malignancy in lesions located at the extremities.

Key words: Lesion; myxoid stroma; myxoinflammatory fibroblastic sarcoma; sarcoma.

Myxoinflammatory fibroblastic sarcoma (MIFS), or acral MIFS, is a recently described, rare, low-grade sarcoma. The tumor often affects the hands and feet, followed by the subcutaneous tissues of the wrist and ankle, and is more rarely located on the lower extremities.[1-8] Cases involving more proximal sites, such as the neck, thigh and shoulder, have been described and the term 'acral' was introduced in the WHO 2002 classification.[5] The tumor is commonly seen in middle-aged patients and equally in both genders. Clinically appearance is of a slowly growing, painless mass with irregular margins on the acral sites of the extremity.[7-10] The lesion may often be confused with hygroma or fibroma adjacent to the joint. Further radiological examination is not usually requested, leading to possible misdiagnosis. It may also be confused with inflammatory and infectious diseases, Hodgkin's disease or various sarcomas due to extensive inflammation on histological examination. As this lesion may be mistaken for clinically and histologically benign lesions detected in the hands and feet, the differential diagnosis must be kept in mind.

In this paper, we present a case of MIFS with a previously undefined appearance on MRI.

Case report

A 53-year-old female patient was admitted to the orthopedics clinic with the complaint of a painless mass on her right ankle for approximately 2 months. On physical examination, a mobile, smooth, painless, cystic mass of 2x2 cm was palpated on the anterior-right part of the right ankle at the inferior end of the fibula. A pre-diagnosis of cystic hygroma was made. However, MRI was performed with the suspicion of a solid tumor. A homogenous hyperintense mass surrounded by regular margins on the lateral part of the ankle was seen on T2 sequences (Fig. 1). However, a mass with regular margins, but com-

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prising heterogeneous hypointense areas in the central part was detected on T1 sequences (Fig. 2). Contrast-enhanced imaging showed the mass to take up contrast only in the central part. Although radiological findings did not suggest malignity initially, total excisional biopsy was preferred over closed biopsy due to small size of the lesion. A wide excision was performed under block anesthesia. The tendon sheaths of the extensor longus muscles were also excised, while the distal tibiofibular syndesmosis tendons were preserved.

On pathological examination, the tumor was located subcutaneously and appeared 2 cm in diameter, demarcated, jelly-like, luminous, and hemorrhagic in one area. On histopathological examination, the tumor was seen to contain fusiform cells in addition to Reed-Sternberg-like and virocyte-like cells and inflammatory cell infiltration on a commonly myxoid, partially hyalinized stroma (Fig. 3). Histochemically, the tumor cells were stained positive with actin, vimentin and S-100. The lesion was diagnosed as MIFS under the light of these histopathological and immunohistochemical findings. The lesion had no superficial tumor tissue in surgical examination for demarcation.

No postoperative complications occurred. The patient was followed up for a period of 7 months and had no complaints.

Discussion

Myxoinflammatory fibroblastic sarcoma is a recently described, low-grade sarcoma. Montgomery et al. first reported 51 cases located on the wrist, fingers and ankle.[9] At the same time, Meis-Kindblom and Kindblom reported a similar tumor series composed of 44 cases related with joint and tendon sheath and defined them as 'acral MIFS'.[10] In another study, Michal detailed 5 similar tumors under the heading of 'bizarre giant cell inflammatory myxoid tumor of soft tissue'.[11]

The tumor usually appears as a slowly growing, painless mass located on the upper extremities.[9-16] However, it may also be seen in more proximal soft tissues such as the neck and the shoulder.[6-8] Many tumors are diagnosed as inflammatory or benign lesions on biopsies due to different histological appearances.

Although MIFS is frequently confused with benign masses due to location and clinical features, sufficient

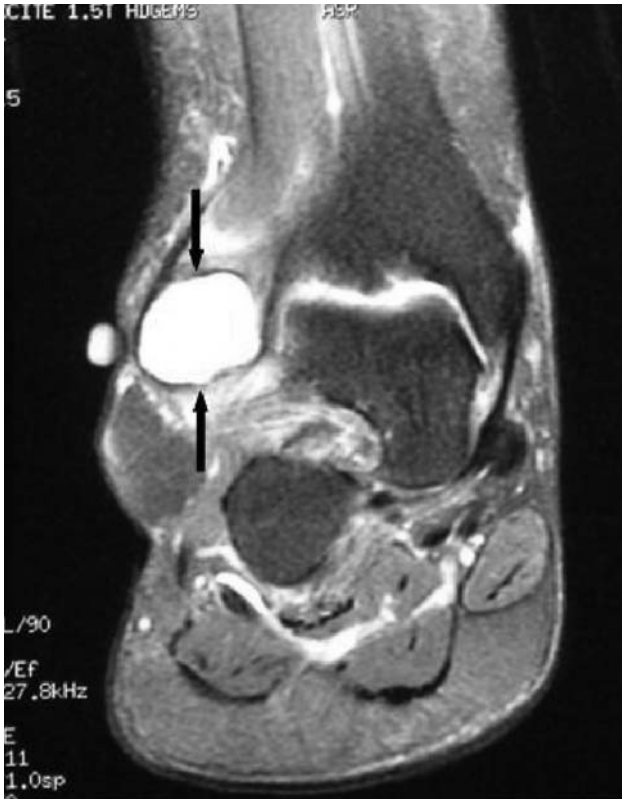

Fig. 1. On MRI of the lesion taken on T2 sequence, a completely cystic appearance full of fluid was obtained. The black arrows show the sharp and regular margins of the lesion. This appearance suggests a benign and cystic tumoral lesion.

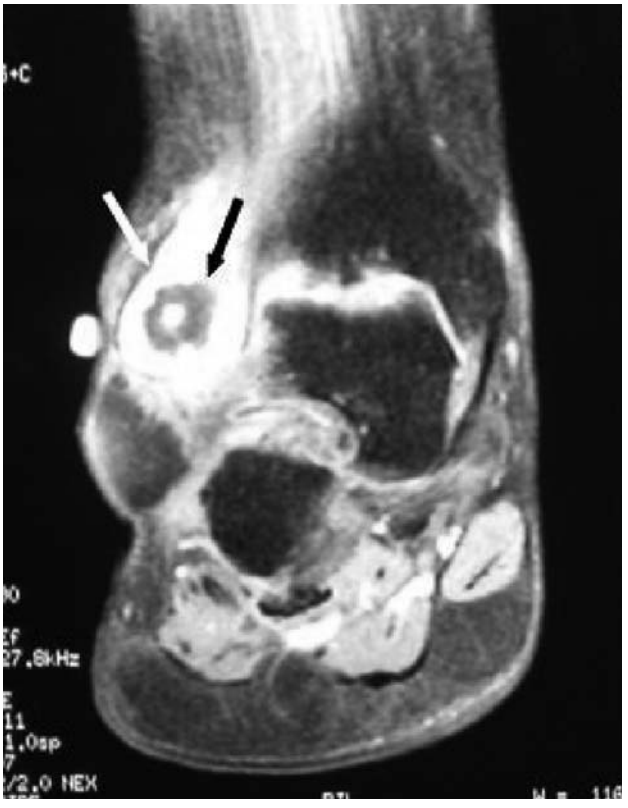

Fig. 2. On MRI obtained on T1 sequence, a hypointense and irregular area (black arrow) is seen in the center of the smooth lesion appearance (white arrow).

data is not available regarding its radiological features. Tateishi et al. compared the MR images of 4 patients with histological findings and reported that myxoid areas in the internal structure of the tumor were seen as heterogeneous contrast-enhanced areas on MRI.[15] Similarly, Lang et al. reported that heterogeneous hyperintense areas may be seen together with focal hypointense areas on T2-weighted sequences and may subsequently be confused with giant cell tumor.[16] The MRI features of our case differ from these previously described findings. In our case, a hyperintense lesion with homogenous internal structure and regular margins was observed suggesting a completely benign and cystic lesion. However, on the sections in the T1 sequence, an irregular hypointense area was present in the center of the lesion. Although initial findings may suggest a benign mass, total excision was performed due to this appearance different than the defined radiological criteria.

Myxoinflammatory fibroblastic sarcoma is a multinodular tumor with irregular margins measuring 1 to 8 cm.[2,4,6,8] It usually appears as a mass most commonly seen on the distal extremities and may be removed by comminution during surgery. Areas that are jelly-like in appearance may be seen in tumors which show large myxoid changes. Histopathologically, MIFS comprises an extensive inflammatory infiltrate and hyalinized areas including neutrophils, plasmocytes, lymphocytes and eosinophils on a myxoid stroma. Dense infiltrates also be seen on low-power microscopy is the most prominent feature. Different areas including solid, myxoid, inflammatory and hyalinized areas are present in MIFS.[1,2,4,5,7,8] Necrosis is rare. Bizarre atypical cells may be seen on more densely cellular areas.[1-3,5,8] These atypical cells may vary from fusiform cells to hystiocytes or epithelioid cells. These fusiform cells usually have moderate nuclear atypia. Large polygonal, ganglion-like cells with basophilic cytoplasm, oval nucleus, vesicular chromatin, prominent nucleoli are observed among the atypical cells. These are Reed-Sternberg or virocyte-like cells seen in the mixed cellular type-Hodgkin's disease.[1-3,5,6,8]

Mononuclear, large bizarre cells immunohistochemically stain strongly positive with vimentin, and variably positive with CD68, CD34 and smooth muscle actin (SMA). They are focally positively stained with cytokeratin. Other determinants (EMA, S-100, HMB-45, melan-A, desmin, F8, CD15, CD30) are negative.[1,3,4,6-8] Similar morphological and immunohistochemical findings were also observed in our case.

While local recurrence is quite high in MIFS, distant metastasis is more rare.[1-3,5] Therefore, close clinical follow-up of patients is recommended. Very rare regional lymph node metastasis has been reported.[2,10] While subcutaneous fat tissue, dermal invasion and focal skeletal muscle invasion may be seen, invasion to the epidermis and bony tissues is not observed. The high local recurrence rate has been suggested to be associated with intralesional removal of the lesion due to diagnostic errors.[11,13,14] In our case, the lesion may be considered to be a benign mass due to its location and physical examination findings. As in many cases, it may be removed intralesionally and malignancy may be found only with pathological examination. In order to avoid local recurrence and repeated surgeries, slowly progressing malignancies should be kept in mind in masses located close to the synovial regions.

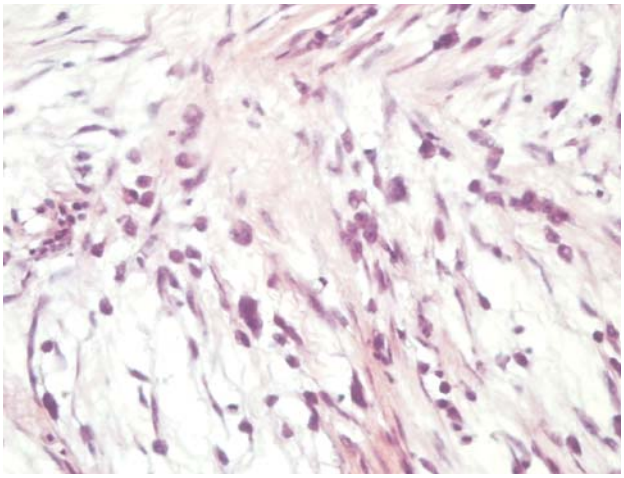

Fig. 3. Inflammatory cells and binuclear Reed-Sternberg-like cells in MIFS (H&E ×400). [Color figure can be viewed in the online issue, which is available at www.aott.org.tr]

Differential diagnosis includes benign myxoid lesions, such as inflammatory lesions, nodular and proliferative fasciitis, tenosynovitis, inflammatory myofibroblastic tumors and myxoma and ganglion cysts.[1,2,4,5,7] In these benign lesions, atypical cells including virocyte-like inclusions are not seen in the inflammatory and myxoid areas. Although preoperative biopsy may be performed, it is difficult to detect the malignancy within the mucinous tissue on needle biopsy. Nevertheless, false-positive results may be obtained. Thus, the mass should be removed through a large excision after the radiological examinations in case there is suspicion in the differential diagnosis.

In conclusion, MIFS is a slowly progressing, low-grade sarcoma with a quite high recurrence rate. Due to the fact that it clinically mimics other benign masses, the local recurrence rate is quite high as a result of insufficient diagnosis and treatment. Previously undefined MRI findings were detected in our case.

Therefore, in the presence of clinical suspicion, distally located cystic-solid masses should be examined with MRI to create the basis for surgical intervention.

Conflicts of Interest: No conflicts declared.

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