Malignant fibrous histiocytoma is the most common soft tissue sarcoma in adults. The most common sites of involvement include the extremities, retroperitoneum, head and neck. Hand is an unusual location. We report magnetic resonance imaging (MRI) findings of a case with malignant fibrous histiocytoma. It presents as a painless soft tissue mass in the right hand.

Key words: Extremities; Malignant fibrous histiocytoma; MRI; Soft tissue tumors

CASE REPORT

A 51-year-old man presented with a painless, slowly enlarging mass in the right hand for months. No trauma nor surgical history was noted in the right hand. His medical history was not contributory and he denied recent body weight loss. On physical examination there was a 5.0 x 3.0cm firm, mobile mass over the dorsum of his right thumb region. The mass was thought initially to be a ganglion cyst or a hemangioma. Then he received MR imaging for further evaluation.

Radiographs of the hand showed a soft tissue mass over the right thumb area without evidence of bony erosion or periosteal reaction (Fig. 1). MR images revealed a subcutaneous mass with well-defined margin in the dorsal area of the right hand near wrist. The mass displayed low signal intensity on T1-weighted images (TR / TE / excitations: 475 / 22 / 2)(Fig. 2) and high signal intensity on T2-weighted images (TR / TE / excitations: 4470 / 184 / 2). The presence of the internal low signal septations was found within the mass (Fig. 3,4). After gadolinium injection, the mass showed nodular and peripheral enhancement (Fig. 5). The adjacent tendons and bone marrow were not involved. Chest radiograph and sonogram of abdomen
demonstrated no evidence of metastatic disease.

The patient received operation with total resection of the mass. Grossly, the specimen had nodular and rough surface. Extensive necrosis, hemorrhage and myxoid change were also evident. Microscopically, the mass revealed neoplastic mesenchymal cells having markedly pleomorphic and hyperchromatic spindle nuclei and moderate to abundant amount of cytoplasm (Fig. 6). Mitotic figures were frequently seen. The findings were compatible with MFH.

DISCUSSION

Masses in the hand are commonly benign, and aggressive cancers are unusual. In the past, MFH of the soft tissue was a relatively rare diagnosis. In recent years the clinicopathologic findings of this tumor were well defined and stated. MFH is recognized as the most common diagnosis in soft tissue sarcomas. Four other variants of MFH are introduced: myxoid, giant cell, inflammatory and angiomatoid. The storiform-pleomorphic subtype of MFH is the most common subtype and most frequently seen as a deep-seated tumor of extremities in middle-aged or elderly patients [3]. Clinically, it appears commonly as a single, often large mass, frequently with areas of necrosis or hemorrhage. Lower extremities are more frequently affected than upper extremities. MFH rarely arises distally to knee or elbow. The most common anatomic sites are thigh, pelvic girdle, leg, and less commonly knee and upper arm [4].

Plain films of MFH in the hand demonstrate only a nonsepecific soft-tissue mass. Occasionally peripheral calcification or cortical erosion of adjacent bones is found but these findings are usually associated with large tumors and are uncommon in hands [5]. Calcification, which occurs in 5 to 20 % of these tumors, is best seen on CT [4]. MRI has dramatically improved our ability to evaluate soft tissue tumors, and images can be reconstructed in multiple planes so as to define precisely the extent. MFH usually manifests as a lobuted mass. On T1-weighted images, the tumors are typically of intermediate to low signal intensity. On T2-weighted images, the tumors tend to be of high signal intensity, although often quite inhomogeneous centrally. There is a mixture of highly cellular areas of fibrosis and acellular areas composed mostly of collagen within the tumor. These areas are
intermixed with areas of necrosis and hemorrhage. MFH is surrounded typically by a well-defined pseudocapsule of low signal intensity [5]. Gadolinium administration will reveal nodular and peripheral enhancement within the solid non-myxomatous elements of the tumor [2].

In 1994, Miller et al. reviewed MR appearances of 13 malignant fibrous histiocytomas of soft tissue and correlated each with the respective lesion’s histopathology. There was no correlation between MR appearance and histopathology. However, there were three features suggestive of malignant soft tissue neoplasms; they are poor marginal definition, internal low signal septations, and heterogeneous high signal intensity on T2-weighted image [6]. In our case, the MR images showed intermediate to low signal intensity on T1-weighted images and internal low signal septations and heterogeneous high signal intensity on T2-weighted images. Nodular and peripheral enhancement was noted after gadolinium injection. Although the tumor was in the hand, malignant soft tissue tumor was highly suspected. Presence of any trace quantity of fat is an important feature to differentiate liposarcoma from MFH. The atypical appearance of liposarcomas without fat has been reported. Pleomorphic and round cell liposarcomas often do not contain fat and are indistinguishable from MFH [7].

The treatment of MFH typically combines wide local excision and radiotherapy. Hematogeneous dissemination to lung is the dominant form of metastatic spread. However, it is a significant improvement in the 5-year survival rate in patients treated with adjuvant chemotherapy. The phenomenon of clinical enlargement of MFH by intratumoral hemorrhage simulating tumor growth during chemotherapy was reported by Panicek et al in 1991 [8]. In previous reports, a high local recurrence rate was observed, ranging from 21% to 48%. A poor prognosis of MFH with a 5-year survival rate ranging from 25% to 40% was reported [4]. Therefore, early diagnosis and adequate surgical excision combined with radiotherapy and chemotherapy are the important factors to increase the survival rate.

MFH is the most common soft tissue sarcoma but it is rare in hand. MRI provides detailed information of anatomical location and tumor involvement so as to make an early accurate diagnosis.
REFERENCES

恶性纖維組織細胞瘤在手部位之磁振造影影像

呂南翰1 黃國書2 馬景先3 陸新政1 吳志順1

國軍左營醫院 放射線部1
三軍總醫院 放射診斷部2
博正骨科醫院3

恶性纖維組織瘤是成人最常見之軟組織肉瘤。常見發生部位為四肢尤其下肢,頭頸部,及後腹膜腔。我們報告一病例臨床發現一無痛性軟組織腫瘤在右手部位約數月之久。磁振造影 T1 影像呈現中間至低訊號,在 T2 影像呈現高訊號影像及腫瘤內有中隔形成，經注射顯影劑後影像特徵為周邊結節狀加強顯影。手術後證實為恶性纖維組織瘤，由於其發生的部位較罕見，故提出報告。

關鍵詞：肢端；恶性纖維組織瘤；磁振造影影像；軟組織腫瘤