A Case of Malignant Fibrous Histiocytoma

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A 57-year-old Korean man with malignant fibrous histiocytoma (MFH) on the leg is presented. Histopathologic examination was consistent with a pleomorphic-storiform subtype of MFH with highly pleomorphic cellular features and frequent mitosis. According to recent evidences, MFH might be a heterogenous group of poorly differentiated tumors rather than a distinct entity. Our case was only positive to CD68 and vimentin immunohistochemically. (Ann Dermatol 12(1) 64–67, 2000).

Key Words: Malignant fibrous histiocytoma

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma of middle and late adulthood1. A case of a 57-year-old Korean man with two, tender, firm subcutaneous nodules on the right lateral shin necessitating wide resection is presented together with a review of literatures.

CASE REPORT

A 57-year-old man had noted a growing tender nodule on his right lateral shin. Because the size of the lesion gradually had increased and another similar nodule on the right shin had developed during the following 1 year, the patient had visited a local clinic. One of the two nodules had been excised at the local clinic. According to the referring letter, one 1×1.5 cm, tender nodule had been excised on the right lateral shin and the pathologic diagnosis had been made as MFH. On presentation to us, he was a healthy man without any systemic symptoms. A past medical and family history were not significant. He had no fever, chill, facial swelling, dyspnea, weight loss, or bone pain. A skin examination revealed a solitary 1×1.1 cm, tender, skin colored, relatively ill-defined, round, firm, subcutaneous nodule on the shin with adjacent linear scar of the previous excision site (Fig. 1). There was neither lymphadenopathy nor other palpable masses. Results of the following laboratory studies were within normal limits or negative; a blood cell count, urinalysis, liver function tests, and chemical battery. On chest X ray, there were no abnormal findings. We carried out an excisional biopsy with a 1-cm margin over the entire nodule. Histopathological examination revealed that a cellular tumor was located deeply under the fascia (Fig. 2) and was composed of cells with large, pleomorphic cells of admixed fibroblast-like cells, histiocyte-like cells and multinucleated cells (Fig. 3). Many mitotic figures were detected. The tumor cells were arranged in irregular intertwining bands composed of ill-defined bundles of atypical spindle cells, forming a vague storiform pattern (Fig. 4). In some parts, infiltrate of many polymorphonuclear cells and atypical mitotic figures of tumor cells were also detected (Fig. 5). There were some myxoid areas and some bizarre multinucleated cells at the periphery of the tumor. A review of the outside-slide which was prepared previously was similar to the new biopsy specimen but there were also foamy cells. In immunohistochemical staining, the tumor cells were diffusely stained with vimentin (Fig. 6A). CD68-positive cells were scattered comprising less than 30% of tumor cells (Fig. 6B). Other im-
Fig. 1. A 1 × 1.1 cm, tender, skin colored, relatively ill-defined, round, firm, deep nodule on the shin with adjacent linear scar of the previous excision site.

Fig. 2. The cellular tumor is deeply situated under fascia (H & E, ×40).

Fig. 3. The densely cellular tumor was composed of large, pleomorphic cells of admixed fibroblast-like cells, histioyte-like cells and multinucleated cells (H & E, ×400).

Fig. 4. The tumor cells were arranged in irregular intertwining bands forming a vague storiform pattern (H & E, ×100).

munohistochemical studies using a battery of monoclonal antibodies against CD34, smooth muscle actin (SMA), desmin, and S-100 protein were all completely negative. Based on the histological studies, the two nodules could be categorized into storiform-pleomorphic subtype of MFH, however there were also combined features of inflammatory or myxoid type of MFH. The patient is well now without recurrence, 4 months after the wide excision.

DISCUSSION

Fibrous tumors of the soft tissue are usually benign, but some fibrous neoplasms such as dermatofibrosarcoma protuberans (DFSP), atypical fibroxanthoma (AFX), and MFH can be very destructive locally with a high recurrence rate after local ex-
MFH is one of the most common soft tissue sarcoma of adult life in Korea. In a statistical analysis of soft tissue tumor of 33 malignant soft tissue sarcomas, 7 cases were MFH the second most common tumor next to liposarcoma. However, the 7 cases visited general surgery (head & neck, trunk) and orthopedic surgery (lower extremities). The location of deep soft tissue may render patients with MFH visiting other surgery departments rather than dermatologic departments. This may be the cause of rare reports of MFH in dermatologic journals despite the common occurrence.

The most common sites of origin are the proximal extremities, particularly the thigh and buttock. The tumors are multilobular fleshy masses, often apparently circumscribed on gross examination, although the microscopic growth pattern is frequently infiltrative among fascial planes and between muscle fibers accounting for the high rate of local recurrence. Involvement of the dermis, occasionally with ulceration is rare. About two-thirds of the tumors occur in striated muscle and less than 10% are confined to the subcutis. A rare case report of dermal origin was reported recently. Histopathologically, MFH is a highly cellular, pleomorphic tumor composed of fibroblast-like spindle cells, histiocyte-like cells, foam cells and giant cells. Mitotic figures including atypical one are plentiful. Four subtypes are recognized and the most common variant is storiform-pleomorphic type composed of spindle cells in storiform pattern, plump histiocyte-like cells, and pleomorphic multinucleated giant cells.

The other three types are as follows; myxoid (myxofibrosarcoma), giant cell type with osteoclast-like giant cells (malignant giant cell tumor of soft parts), inflammatory type with dense infiltrate of neutrophils and xanthoma cells (malignant xanthogranuloma). However, there is much overlapping in these subtypes and even variability of subtypes in individual tumors of same patient. This overlap and histopathological variability were also seen in our case.

MFHs behave biologically as a neoplastic rather than reactive mesenchymal tumor with definite invasive and metastatic potential. Metastasis is related to tumor size, depth, and grade. With the help of more effective surgical therapy such as Mohs' surgery, the rates of local recurrence and metastasis have decreased. In a study published in 1992, a local recurrence rate of 25% and a metastatic rate of 34% and an overall survival rate of 50% were reported.

In differential diagnosis, DFSP is a CD34 positive fibrohistiocytic tumor of intermediate malignancy characterized by aggressive local growth and propensity to recur but metastasis is extremely rare. Histologically, storiform-pleomorphic type of MFHs composed of spindle cells in storiform pattern may resemble DFSP but the tumor cells are far more pleomorphic with plump histiocyte-like cells and multinucleated giant cells. AFX is a superficial malignant tumor that arises on sun-exposed surfaces and persues almost an invariably benign course. MFH is the most aggressive of the fibrohistiocytic tumors with a high local recurrence.
rate and significant metastatic rate usually associated with a poor prognosis. The histogenesis of these three tumors are still uncertain. In MFH, no pathognomonic immunophenotype exists although vimentin, alpha-1-antitrypsin, CD68, and factor XIIIa were frequently positive in tumor cells. Some postulate that MFH arises from histiocytes which is supported by the CD68 positivity in the tumor cells. However, CD68 has been shown to stain various lineage cells in addition to histiocyte and macrophage derivation. CD68 has been shown to stain non-hemopoietic cells such as hepatocytes, renal glomeruli and renal tubules and a sensitive and specific marker for neoplasms of myeloid and myelomonocytic in the form of intense granular cytoplasmic staining, and that some lymphomas and leukemias of B-lineage can exhibit dot reactivity. Moreover, CD68 has recently been shown to stain non-hemopoietic tumors such as granular cell neoplasms and some malignant melanomas. Considering the great variability of the staining, care must be exercised in utilizing CD68 for determining the histogenesis or cellular differentiation of tumors. Although most authors favor either a histiocytic, a fibroblastic or a dual fibroblastic-histiocytic origin, the tumor cells of MFH do not display a characteristic histiocytic immunophenotype. Sometimes, a wide range of intermediated filaments such as muscle specific actin, alpha-smooth muscle actin or even keratin have been identified in MFH. Fletcher et al. have suggested that MFH is a morphologic pattern of mesenchymal and non-mesenchymal tumors in which a large proportion could be reclassified specifically with help of adequate tissue preparation and further studies. After all, MFH might be considered as a heterogenous group of poorly differentiated tumors rather than a distinct entity.

REFERENCES