CASE REPORT

Ossifying fibromyxoid tumor — A case report

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Ossifying fibromyxoid tumor (OFMT) is a rare soft tissue tumor of uncertain histogenesis, occurring predominantly in deep soft tissues of the extremities. Typically, OFMT presents in adults on the extremities or trunk, as a deep soft tissue mass. Less appreciated is the fact that OFMT may also present as a mass in the superficial subcutis or dermis. We herein report a female who presented with an asymptomatic subcutaneous nodule on the left thigh for 3 years, and who was diagnosed as having typical ossifying fibromyxoid tumor, by unique histopathologic and immunohistochemical studies. Most reported cases have pursued a benign clinical course. However, recent literature emphasized the existence of morphologically atypical and clinically malignant cases of OFMTs. Pathologic criteria for malignancy have been proposed, and reclassification of these tumors as tumors of intermediate malignancy, raise our attention while coping with OFMT clinically.

Introduction

Ossifying fibromyxoid tumor (OFMT) is a rare tumor and was first described by Enzinger et al in 1989, based on 59 cases culled from the archives of the Armed Forces Institute of Pathology over a 25 year period. Most cases arise from subcutaneous or intramuscular parts of the lower extremities, and only about 11% cases involved the superficial skin tissue such as the dermis. Clinically, OFMT usually presents as a solitary, firm, well-defined, slowly growing, painless deep soft tissue or subcutaneous mass. Histologically, the typical tumor is characterized by a fibrous, encapsulated, lobulated neoplasm, with a peripheral rim of bony component. It is composed of a cord- or nest-like arrangement of round, oval, or spindle-shaped uniform cells with vesicular nuclei. Therefore, OFMT should be considered in the differential diagnosis of cutaneous bone-forming tumors.

Case report

A 30-year-old female presented with a well-circumscribed, firm, asymptomatic subcutaneous mass of her left thigh (Figure 1). It was enlarging slowly over the past 3 years. No specific past medical history or family history were obtained according to her statement. An excisional biopsy was performed under the impression of dermatofibroma or other adnexal tumors.

Grossly, a tumor measuring 0.6 × 0.5 × 0.4 cm in size at the superficial subcutis was noted. Microscopically, it revealed a well-defined, encapsulated tumor with a fibrous capsule and a focal existence of lamellar bone component (Figures 2A,2B). Uniform, round to ovoid cells, with eosinophilic cytoplasm, were arranged in a cord- or nest-like pattern within a homogenous fibromyxoid stroma (Figure 3). The nuclei were relatively uniform and vesicular, without any evidence of pleomorphism, hypercellularity, or necrosis. There was no mitosis noted (Figure 4). An immunohistochemical study revealed positive results for S-100 (Figure 5) and negative results for CK and CD34. Scattered tumor cells were positive for SMA. A diagnosis of ossifying fibromyxoid tumor was made. The patient underwent surgical wide excision about 1 month later. The postoperative course was uneventful. We suggested regular follow-up afterwards.

Discussion

OFMT was first described by Enzinger et al in 1989 as a rare deep soft tissue tumor of uncertain differentiation. OFMT usually occurs in adults, with a male predominance (M:F ratio = 1.5:1). Most
cases involved lower extremities, followed by trunk, upper extremities, and the head and neck region. Cutaneous OFMT is characterized, clinically, by a circumscribed, long-standing, asymptomatic, firm, and slowly enlarging mass, and histologically by a well-encapsulated lobular architecture, with round to ovoid uniform cells arranged in cord and nest patterns, within collagenous to myxocollagenous stroma, usually demonstrating peripheral focal spicules of metaplastic bone component. Small satellite tumor nodules outside the main tumor were sometimes noted, and these were also surrounded by a well-defined fibrous capsule, which developed later.

Although Enzinger emphasized its indolent, slowly growing natural course and benign behavior, several subsequent reports have documented, both histologically and clinically, atypical to malignant variants of OFMT. Yoshida et al first reported a tumor that lacked overtly malignant features, but produced local recurrence, distant soft tissue metastasis, and death. Soldano et al reported a case of subcutaneous OFMT which recurred 8 years after initial resection, with aggressive histologic change, such as a high mitotic rate and cell atypia compared to the primary lesion. Suehiro et al reported a case with a huge metastatic OFMT occupying the upper mediastinum and upper half of the right hemithorax, who died 8 months later after surgery, due to multiple brain and opposite lung metastasis. Folpe and Weiss performed a retrospective review of 70 cases of OFMT; they focused on the atypical and malignant cases related with clinical malignant behavior, such as local recurrence and distant metastasis, over the mean follow-up duration of 57 months. The presence of high cellularity or high nuclear grade, with mitotic activity >2 mitotic figures per 50 high power fields (HPFs) revealed a statistically significant potential for malignant behavior and should be subclassified into the group of malignant OFMT, which revealed 60% local recurrence rate and 60% metastasis rate. A malignant OFMT should be regarded as a sarcoma, due to its highly aggressive clinical behavior. Even in typical OFMT cases, overall recurrence and metastatic rates, 17% and 5%, respectively, make us consider them as lesions of intermediate malignancy. Recurrence of typical OFMT usually occurred >10 years after primary excision, highlighting its low growth rate. A conservative approach for conventional typical OFMT, such as complete excision with a margin of normal tissue and long-term follow-up, is considered optimal nowadays. In the Folpe and Weiss classification, those deviating from typical OFMTs, but falling short of proposed criteria for malignant OFMTs, were labeled as “atypical OFMTs”, which possessed a 13% local recurrence rate and a 6% metastasis rate.

The histologic differential diagnosis of cutaneous OFMT includes cutaneous mixed tumor (chondroid syringoma), myoepithelioma of skin and soft tissues, extraskeletal osteosarcoma, and extraskeletal myxoid chondrosarcoma. Cutaneous mixed tumors and myoepitheliomas also contain strands, nests and a reticular pattern of bland tumor cells within a hyaline myxoid stroma. Furthermore, calcification and ossification are not uncommon in both tumors. The presence of a ductal/epithelial component in mixed tumors, which is absent in OFMT, and immunoreactivity for cytokeratin or calponin in mixed tumor and myoepithelioma, can be useful in

Figure 1 A 30-year-old female presented with a well-circumscribed, firm, asymptomatic subcutaneous mass on her left thigh.

Figure 2 (A) A well-defined, encapsulated superficial subcutis tumor with fibrous capsule and focal existence of woven bony component (hematoxylin and eosin stain, ×20); (B) a chip of woven bone was present at the peripheral portion of the tumor (hematoxylin and eosin stain, ×400).

Figure 3 Uniform, round to ovoid cells with eosinophilic cytoplasm were arranged in cord- or nest-type pattern within a homogeneous fibromyxoid stroma (hematoxylin and eosin stain, ×100).
differential diagnosis. Although similar in arrangement of neoplastic cells, the characteristic features of basophilic chondromyxoid stroma, more abundant vacuolated cytoplasm and rarely, ossification in extraskeletal myxoid chondrosarcoma, are different from the OFMT. Cutaneous extraskeletal osteosarcoma is very rare. The unequivocal presence of osteoid, usually accompanying high-grade nuclei of tumor cells, is essential for the diagnosis of osteosarcoma. The OFMT lacks osteoid and mostly contains bland tumor cells.

The lineage of differentiation is uncertain. Nerve sheath and cartilaginous origin were commonly suggested differentiation types according to some immunohistochemical and ultrastructural studies, but are still not conclusively proven. All typical OFMT are immunoreactive for vimentin (100%), and 94% immunoreactivity for S-100 has been reported. Other variable immunoreactivities include CD10 (79%), pancytokeratin (13%), collagen IV (13%), desmin (10%), and smooth muscle actin (2%). However, S-100 protein positivity was found to be lower in some studies, especially those which included atypical and malignant cases. It has been suggested that loss of S100 protein may be related to malignant transformation. However, some experts proposed the possibility that malignant OFMT may be fundamentally different from typical OFMT.

In summary, we present a case of OFMT located in the superficial subcutis. OFMT typically presents as a mass in the deep soft tissue and is less appreciated in the superficial subcutis or dermis. Besides, according to the reported literatures, persistent follow up for local recurrence is needed even without any evidence of pleomorphism, hypercellularity, or necrosis in the primary lesion.

References