Focal myxoid change in synovial sarcoma is not uncommon, although the presence of predominantly myxoid stroma is very rare. Recognition of synovial sarcomas with massive myxoid feature is important because these can easily be mistaken for other myxoid soft tissue neoplasms. We report a case of a synovial sarcoma with massive myxoid feature in the left thigh of a 54-year-old woman. Wide excision of an 8.5 × 7.0 × 5.0 cm, well-circumscribed and lobulated tumor was performed. The cut surface was gray, soft, and myxoid. Histological examination showed proliferation of spindle cells in the predominantly myxoid stroma. There were small areas with features more typical of synovial sarcoma, including uniform, spindled cells with fascicular growth patterns, collagenous stroma, mast cell infiltration, and hemangiopericytoma-like vascular patterns. Immunohistochemical examination showed focal positivity of the tumor cells for epithelial membrane antigen (EMA). Tumor cells were all negative for cytokeratin (AE1/AE3), cytokeratin 7, S-100 protein, smooth muscle actin, and desmin. Ultrastructurally, tumor cells showed desmosomes and microvilli. Our case underscores that, in order to make a correct diagnosis, immunohistochemical and ultrastructural examination is essential.

Key Words: Synovial sarcoma; Soft tissue; Myxoid; Sarcoma

CASE REPORT

A 54-year-old woman presented with a mass in her left upper thigh which had been present for six months. The egg-sized mass was first noted six months before and the mass began to grow more rapidly from two months before admission. Her medical history was noncontributory. Physical examination revealed a hard, movable mass in the left upper thigh. No pain and tenderness were present. Magnetic resonance (MR) images showed a soft tissue mass in the left upper thigh. There was low signal on the T1-weighted image and high signal on the T2-weighted image (Fig. 1). A wide excision was performed.

Synovial sarcoma is a mesenchymal spindle cell tumor which displays variable epithelial differentiation. It accounts for 5 to 10% of soft tissue sarcomas. Synovial sarcoma mainly occurs in young adults and occurs more commonly in males than in females. Histologically, synovial sarcoma is either biphasic or monophasic. Focal myxoid change in synovial sarcoma is not uncommon. The presence of predominantly myxoid stroma had rarely been reported. Krane et al. described synovial sarcoma with predominantly myxoid change as myxoid synovial sarcoma. Myxoid synovial sarcoma can be easily confused with other myxoid spindle cell neoplasms. Therefore, the recognition of myxoid synovial sarcoma is important. Here, we describe a case of a synovial sarcoma with a massive myxoid feature of the left thigh of a 54-year-old woman, and review the relevant literature.
positively with alcian blue and was inhibited by prior hyaluronidase treatment. On the immunohistochemical stain, tumor cells showed focal positivity for EMA, particularly in the fascicular spindle cell area (Fig. 3D). Tumor cells were all negative for cytokeratin (AE1/AE3), cytokeratin 7, S-100 protein, smooth muscle actin, and desmin. Ultrastructurally, tumor cells showed desmosomes and microvilli (Fig. 4). Our diagnosis was myxoid synovial sarcoma. Adjuvant chemotherapy was administered. Twelve months after the surgery, the patient remains in good health.

**DISCUSSION**

Synovial sarcoma is a soft tissue sarcoma which usually arises adjacent to joints or tendon sheaths, particularly in the lower extremities. Its histogenesis is unknown. Most of the patients present with a palpable, deep-seated mass that is often painful. Some tumors have radiologically irregular calcification that is occasionally massive.

Synovial sarcoma has a wide histologic spectrum that includes three variants: monophasic, biphasic, and poorly differentiated. Common histologic features of synovial sarcoma include a variably collagenous stroma with a hemangiopericytoma-like vascular pattern, intratumoral calcification, and mast cells. Myxoid change is not uncommon, but it is generally so focal as to be diagnostically inconsequential.

The term myxoid synovial sarcoma was first described in 1999 by Krane et al. who defined myxoid synovial sarcoma as a condition in which myxoid stroma constitutes greater than 50% of the studied surface area. Krane et al. documented a series of myxoid synovial sarcoma in which marked myxoid change initially obscured the diagnosis and led to confusion. Moffatt et al. reported the cytologic features of synovial sarcoma with predominantly myxoid stroma.

We reviewed our case, as well as seven previously reported cases of myxoid synovial sarcoma. The age of the patients ranged from 12 to 54 years (median age 20 years). The male-to-female ratio was 1:1.25. Six tumors arose in the lower limbs (four in the thigh and one in the knee), two arose in the upper limbs (one in the elbow and one in the forearm), and one arose in the head and neck (arytenoid region).

According to a report by Krane et al., myxoid synovial sarcoma constituted only 2% of all synovial sarcoma encountered in a consulting practice. The extent of myxoid change in synovial sarcoma was variable. The present case had myxoid stroma which constituted more than 90% of the tumor volume. In our case, the presence of uniform, spindle cells with a fascicular growth

**Fig. 1.** MR images of the left thigh tumor. (A) Axial T1-weighted image shows low signal intensity. (B) Axial T2-weighted image shows high signal intensity.

**Fig. 2.** Gross findings of the left thigh tumor. The left thigh tumor shows an 8.5 × 7.0 × 5.0 cm, well circumscribed, lobulated, and myxoid appearance.
pattern, with foci of collagenous stroma, mast cells, and focal hemangiopericytoma-like vascular pattern suggested features of synovial sarcoma. In Krane’s series, two of seven cases showed small foci of glandular (biphasic) differentiation. In the present

Fig. 3. Histologic findings of the left thigh tumor. (A) The tumor shows a spindle cell proliferation in the predominantly myxoid stroma. (B) The tumor shows transition from cellular area to myxoid area. (C) The tumor cells show uniform, spindle shaped nuclei. (D) The immunohistochemical stain shows focal positivity for EMA.

Fig. 4. Electron microscopy of the tumor cells. (A) Tumor cells show desmosomes (original magnification, ×15,000). (B) Tumor cells show projecting microvilli (original magnification, ×10,000).
case, no glandular differentiation was observed.

The origin of myxoid material in synovial sarcoma is unknown. It has been suggested that myxoid material may be associated with a degenerative process or a product of stromal fibroblasts. Myxoid material has never been found in the cytoplasm of tumor cells, thus, it is unlikely to be produced by tumor cells.

The predominantly myxoid change is not uncommon in many soft tissue neoplasms. Because of the relative chemosensitivity of synovial sarcoma, it is important that they be distinguished from other myxoid spindle cell sarcomas. The histologic differential diagnosis of synovial sarcoma with massive myxoid feature includes aggressive angiomyxoma, low grade fibromyxoid sarcoma, myxoid liposarcoma, myxoid leiomyosarcoma, myxofibrosarcoma, and myxoid malignant peripheral nerve sheath tumor (MPNST). Aggressive angiomyxoma has prominent blood vessels and desmin positive tumor cells. Low-grade fibromyxoid sarcoma is distinguished by its whorled growth pattern and its more prominent fibrous stroma. Myxoid liposarcoma typically contains multivacuolated lipoblasts and anastomosing vessels embedded in myxoid background material. Myxofibrosarcoma may resemble synovial sarcoma, but the nuclear pleomorphism and hyperchromatism in myxofibrosarcoma are greater than those present in myxoid synovial sarcoma. Myxoid leiomyosarcoma contains intersecting fascicles with more abundant eosinophilic cytoplasm and cigar-shaped nuclei. Using immunohistochemical methods, myxoid leiomyosarcoma is immunoreactive with smooth muscle actin and desmin. In our case, tumor cells were negative for smooth muscle actin and desmin. The most problematic differential diagnostic of the present case is myxoid MPNST. A perivascular accentuation (or whorling) of the tumor cells and the presence of heterologous elements favor a diagnosis of MPNST. Immunohistochemical analysis can be helpful in distinguishing synovial sarcoma from other sarcomas.

In conclusion, we have described a case of synovial sarcoma with a massive myxoid feature arising in the left upper thigh. Synovial sarcoma with massive myxoid feature should be considered in the differential diagnosis of other myxoid soft tissue neoplasms.

REFERENCES

11. Lopes JM, Bjerkeløv B, Holm R, Bruland O, Sobrinho-Simões M, Nesland JM. Immunohistochemical profile of synovial sarcoma with emphasis on the epithelial-type differentiation. A study of 49 primary


