

# Fine Needle Aspiration Biopsy of a Myxoid Leiomyosarcoma with Epithelioid Features and It Metastasized to the Abdominal Wall – A Case Report –

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We present the cytologic findings observed in a fine needle aspiration biopsy specimen of a rare myxoid variant of leiomyosarcoma with epithelioid features and the tumor had metastasized to the abdominal wall. The aspirate showed hypercellularity in a hemorrhagic background. Some large 3-dimensional aggregates of spindle cells were observed. Each cell had a solitary ovoid-to-elongated nucleus with finely granulated chromatin, one or two small distinct nucleoli and an irregular nuclear membrane. There were irregular fascicles of spindle cells with cigar-shaped, blunt-ended nuclei admixed with inflammatory cells. Epithelioid cells with a rather narrow, dense cytoplasmic rim and a well-defined cell border were embedded in a myxoid matrix in a cord-like and cluster arrangement. The matrix appeared as a pale green substance with sharply defined edges. There were very few mitoses. These cytologic features were the same as those of a uterine myxoid leiomyosarcoma that was surgically excised 7 years ago, and immunohistochemical staining revealed the smooth muscle origin of the tumor.

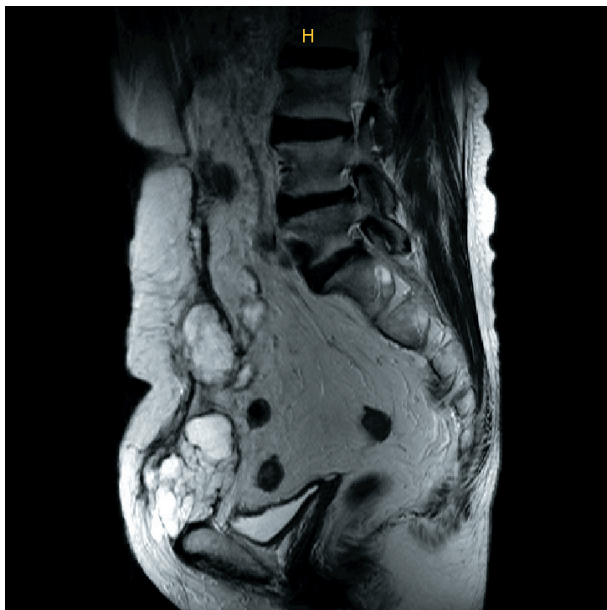
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Myxoid leiomyosarcoma (LMS) of the uterus is an extremely rare neoplasm that was first described by King *et al.* in 1982.<sup>1</sup> This sarcoma is characterized by a jelly-like cut surface with infiltrative borders, a low mitotic rate with minimal cellular atypia and a subsequent malignant course. The cytological findings of a fine needle aspiration biopsy (FNAB) of a uterine myxoid LMS have not been previously described. Here we report on a LMS that metastasized to the abdominal wall in a 73-year-old woman.

## CASE REPORT

The patient was a 73-year-old Korean woman who presented with lower abdominal pain of several months duration. A movable, tender mass was palpated at the lower abdominal wall on

physical examination. The magnetic resonance imaging revealed a 7 × 5 × 12 cm multiloculated and conglomerated mass with mainly solid components that involved the lower abdomen and pelvic wall, and the tumor protruded into the upper pelvic cavity on the T2WI sagittal view (Fig. 1). Other satellite masses were also observed. The patient had undergone a total hysterectomy along with bilateral salphingo-oophorectomy for a large leiomyoma uterus seven years previously, and so a myxoid LMS with focal epithelioid features was diagnosed. A metastatic mass was noted in the abdominal wall 3 years after the hysterectomy, and the mass was removed. Four years after the second operation, another metastatic nodule was found at the previous surgical site; a FNAB was obtained. The laboratory findings showed no specific abnormalities except for anemia.



**Fig. 1.** Magnetic resonance imaging shows a multi-loculated and conglomerated solid mass on the T2WI sagittal image. The mass involves the lower abdomen and pelvic wall and the mass is protruding into the upper pelvic cavity.

### Cytological findings

The aspiration specimen from the metastatic abdominal mass was highly cellular with a hemorrhagic background. Large three-dimensional syncytial tissue fragments of spindle cells were observed at low magnification (Fig. 2A). The background material was obscured by blood. However, there was pale green, thin mucus. We observed well-defined fragments in which the cells floated in myxoid fragment in certain areas (Fig. 2B). At a higher magnification, ovoid nuclei with finely granular chromatin, and one or two small distinct nucleoli, were noted with a markedly irregular and folded nuclear membrane; occasional nuclear cytoplasmic inclusions were also noted (Fig. 2C). These elements formed three-dimensional irregular fascicles admixed with lymphocytes. The spindle cells were observed to be elongated; they had cigar-shaped nuclei with fine chromatin and a barely discernible nucleolus. As the density of the cells decreased, the cell clusters, in which the cells had a wisp of cytoplasm, were observed to be loosely arranged in watery pale-green mucoid material. A small number of sporadic naked ovoid nuclei and spindle cells with bipolar processes were also observed. Within a small number of small-sized, well-defined myxoid tissue fragments, the tumor cells had ill-defined, thin dense eosinophilic cytoplasm and ovoid nuclei. The tumor cells also formed small clusters or cord-like arrangements (Fig. 2D). In rare places, there were a few

fragments in a parallel arrangement along the capillaries, of which only the spindle cells had elongated, blunt-ended nuclei with bipolar processes (Fig. 2E). Any multinucleated tumor giant cells were not found and the mitotic activity was very low.

### Histological findings of the uterus and the abdominal wall mass

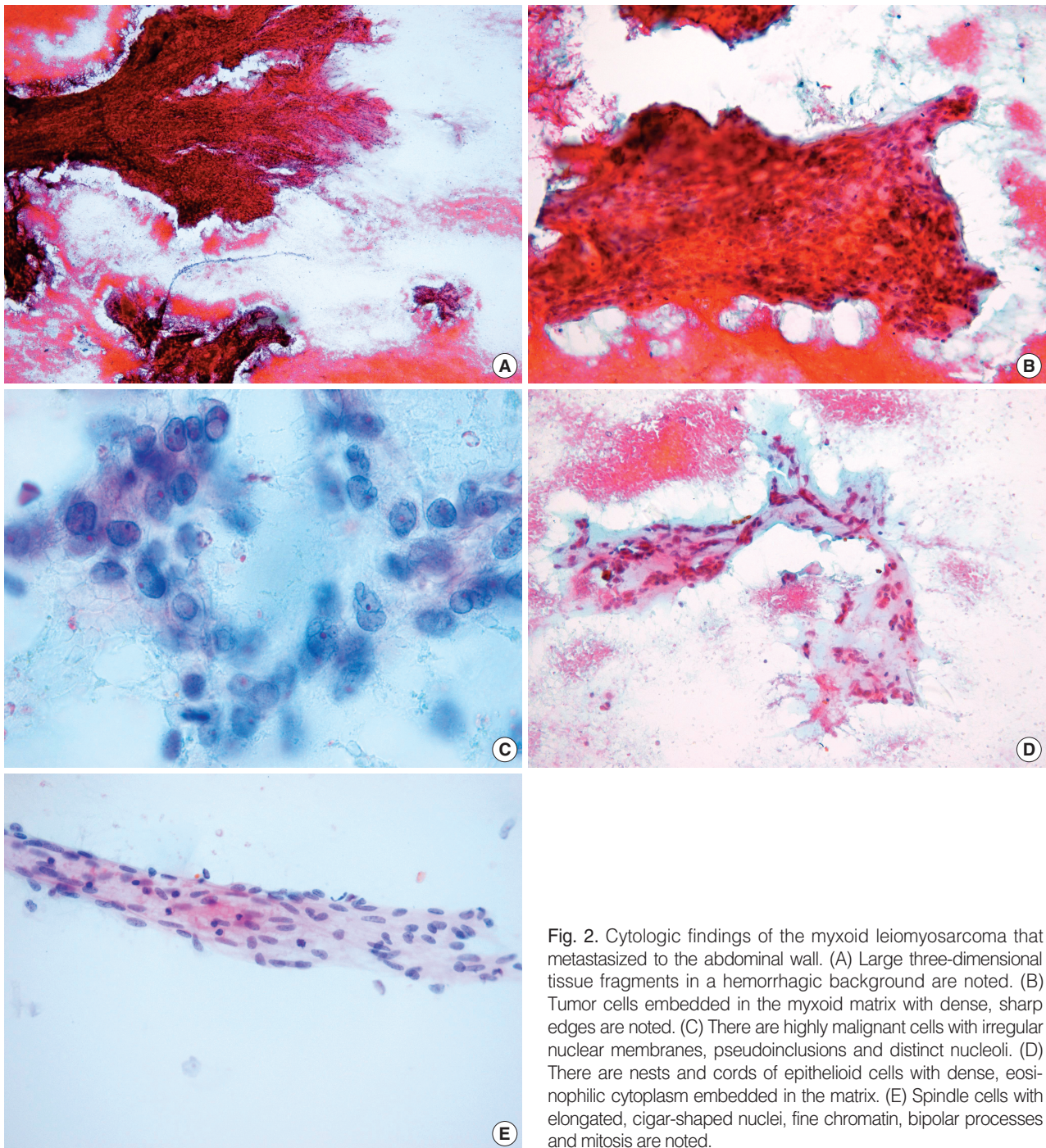
The hysterectomy specimen consisted of the uterine corpus, uterine cervix, bilateral fallopian tubes, and bilateral ovaries. The surgical specimen was  $10 \times 15 \times 8$  cm and it weighed 1,340 g. A relatively well-circumscribed subserosal mass measuring  $10 \times 8 \times 7$  cm was attached to the anterior uterine wall. On sectioning, the tumor had an amber green gelatinous cut surface with focal hemorrhage. The histological examination revealed a myxoid LMS with partial epithelioid features. The tumor cells had an elongated-to-ovoid shape and moderate-to-marked atypia, and small but clear nucleoli were identified. The spindle cells floated in the mucoid matrix and formed short irregular fascicles, and the epithelioid cells with moderate eosinophilic cytoplasm and vaguely defined cell borders formed small irregular cords and nests. Many narrow and delicate capillary structures were observed and blood-filled cysts were also noted. Some of the polygonal cells showed microvesicular and foamy cytoplasm, and xanthomatous changes were also observed. Yet the mitotic activity was very low (1/10 high power field [HPF]). The myxoid tumor nests had infiltrated into the neighboring myometrium. Immunohistochemical staining was positive for smooth muscle actin and desmin, and it was negative for CD10 and cytokeratin.

The abdominal mass was located inside of muscle and it measured  $7 \times 5 \times 12$  cm. Abundant mucoid material was found on the sections. On the histological examination, the irregular cords and aggregates of malignant cells were settled in the mucoid background, and the malignant cells had infiltrated into the skeletal muscle and fascia (Fig. 3A). The features were the same as those of the tumor found in the uterus on histological examination and immunohistochemical staining (desmin positivity) (Fig. 3B); therefore, the diagnosis was metastasis from the uterine myxoid LMS.

## DISCUSSION

FNAB is not a widely accepted technique for evaluating soft tissue tumors. This is because of its poor histological cytological concordance, the morphological heterogeneity within the same

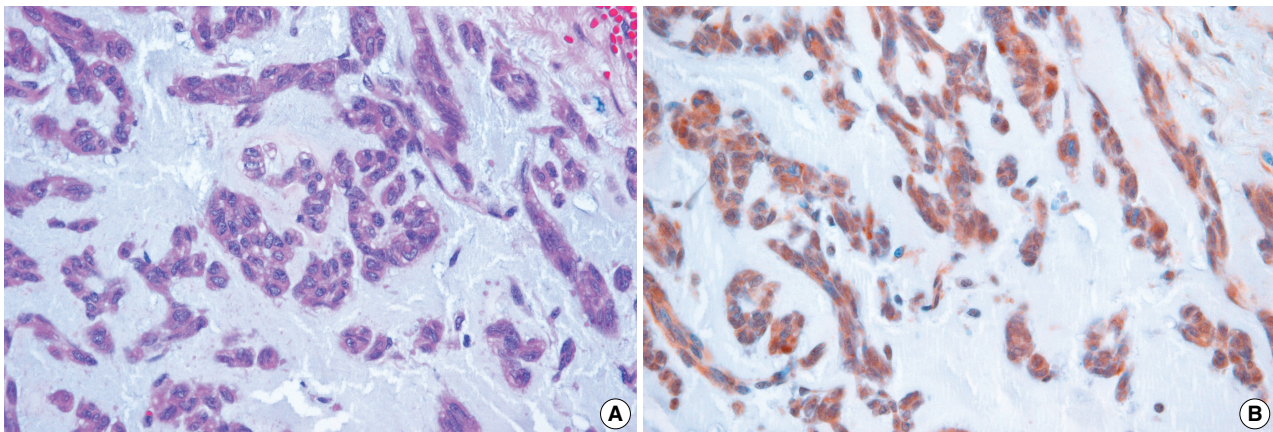




**Fig. 2.** Cytologic findings of the myxoid leiomyosarcoma that metastasized to the abdominal wall. (A) Large three-dimensional tissue fragments in a hemorrhagic background are noted. (B) Tumor cells embedded in the myxoid matrix with dense, sharp edges are noted. (C) There are highly malignant cells with irregular nuclear membranes, pseudoinclusions and distinct nucleoli. (D) There are nests and cords of epithelioid cells with dense, eosinophilic cytoplasm embedded in the matrix. (E) Spindle cells with elongated, cigar-shaped nuclei, fine chromatin, bipolar processes and mitosis are noted.

tumor and the development and clinical recognition of borderline tumors with intermediate malignancy among the soft tissue tumors. However, compared to other biopsy techniques, FNAB is relatively easy to perform and it is cost-effective. Myxoid changes are not rare findings in soft tissue tumors. Such findings allow making a histopathological and cytological diagnosis. The tumors that are rich in a myxoid matrix include lipo-

blastoma, myxoma, neurofibroma, nodular fasciitis, proliferative myositis, chondroid lipoma, myxoid liposarcoma, myxofibrosarcoma, extraskeletal myxoid chondrosarcoma and unusual forms of a LMS and dermatofibrosarcoma protuberance.<sup>2</sup> In the current case, the finding of epithelioid cells embedded in the myxoid matrix suggested that we consider metastatic mucinous carcinoma in the differential diagnosis. Yet for the cases with mucinous



**Fig. 3.** Histologic findings of the myxoid leiomyosarcoma that metastasized to the abdominal wall. (A) The cancer cells float in the mucoid matrix and form small irregular cords and nests and the cancer cells have moderate eosinophilic cytoplasm and well-defined margins. (B) Based on immunohistochemical studies, the tumor cells are positive for desmin.

carcinoma, the tumor cells show strong cohesiveness and a relatively clear cellular membrane or boundary. These features are not consistent with the findings of the case reported herein. The most characteristic features of our case were the three-dimensional tissue fragments of ovoid to spindle-shaped cells, which were assumed to be a sarcoma rather than a carcinoma. In addition, there was an absence of the large prominent nucleoli and melanin pigments that can be observed in cases of melanoma with myxoid change. Most of the cases of the above-mentioned benign myxoid soft tissue tumors have been hypocellular with a bland chromatin pattern, and these tumors are characterized by 1) the presence of stellate cells with three or more cytoplasm processes in a myxoma, 2) fatty tissue, multivacuolated cells and chondroid-like fragments with cells in the lacunae and irregular nuclear membrane in a chondroid lipoma, 3) ganglion-type cells with abundant, dense cytoplasm and one or two oval, peripherally displaced nuclei containing a prominent nucleolus in cases with nodular fasciitis and proliferative myositis, and 4) spindle cells with wavy and comma-shaped nuclei in cases of myxoid neurofibroma.<sup>2</sup>

For the cases of myxoid liposarcoma,<sup>2-4</sup> the findings include optically empty vacuoles, a myxoid matrix with delicate branching capillary segments and most importantly lipoblasts. For cases of extraskelatal myxoid chondrosarcoma,<sup>2-5</sup> the characteristic findings are cells embedded within lacunae, chondromyxoid fragments with sharp edges, conspicuous cytoplasm vacuoles and round chondroblasts that form short cordons. These findings differ from those found in our present case. Myxofibrosarcomas<sup>2-4</sup> are characterized by prominent plexiform collagen-thickened capillary vessels, a granular myxoid background with cell detri-

tus and pleomorphic large or giant cells. In addition, the dermatofibrosarcoma protuberance is composed of a storiform arrangement of spindle cells with evenly distributed chromatin and small, inconspicuous nucleoli.<sup>2</sup>

For the patient reported on here, the tissue fragments consisting of a parallel arrangement of spindle cells with cigar shaped nuclei might have been a diagnostic clue for smooth muscle differentiation. In addition, although rare, the typical histopathological findings of a LMS were also observed, such as bipolar processes, dense, acidophilic fibrillar cytoplasm and the centrally located nucleus containing fine chromatin and a barely discernable nucleolus. There were no paranuclear vacuoles. However, nuclear cytoplasmic inclusions are one of the findings observed in cases with a LMS. Furthermore, ovoid pleomorphic nuclei with a dense, eosinophilic cytoplasm rim help with determining the epithelioid differentiation of LMS. Histologically, in the current case, there were some xanthomatous cells; these were thought to be due to degenerative changes. On electron micrograph, ballooning of mitochondria and dilated vesicular structures have been reported in LMS.<sup>6</sup>

In general, diagnosing a soft tissue tumor according to the aspiration cytology should include differentiating it from a pseudosarcomatous lesion. The most common lesion is nodular fasciitis; the hypercellularity, bland chromatin pattern and clinical features are helpful in differentiating these lesions from a sarcoma. Miralles *et al.*<sup>7</sup> reported a 4% false-positive diagnosis rate and they concluded that in most cases fat necrosis was incorrectly diagnosed as a liposarcoma. Wakely *et al.*<sup>5</sup> reported that sarcomas showed a moderate-to-high level of cellularity more often than that of benign lesions (sarcoma, 95%; benign, 18%) and more



frequent moderate-to-marked nuclear atypia (59% vs 9%, respectively). Yet it should be noted that, marked nuclear atypia can also be observed in cases of pleomorphic lipoma. A majority of benign and malignant myxoid tumors have a matrix that is relatively watery with a translucent and relatively poorly defined outline. But for most cases of extraskelatal myxoid chondrosarcomas, much denser mucin with sharply-defined smooth edges, as was seen in the tissue fragments of our case, has been observed as the characteristic findings. In addition, the myxoid benign specimens have an amorphous, semi-transparent appearance that is not fragmented but rather, the appearance is that of a thin, diffuse film.<sup>5</sup> In the case reported here, tissue fragments of the thicker myxoid matrix with sharp edges were observed. Such findings should suggest the possibility of a malignant myxoid tumor, although more cases are needed to confirm this. In addition, confirmation of whether the cells in the matrix have nuclear atypia or lacunae is helpful. This case illustrates the findings of metastatic myxoid LMS with epithelioid features in an abdominal wall aspirate from a 73-year-old woman. The characteristic cytologic findings include hypercellularity, ovoid to spindle-shaped vesicular nuclei with an irregular membrane, some cigar-shaped nuclei and myxoid tissue fragments with floating epithelioid cells without lacunae.

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