

A Rare Case of Uterine Myxoid Leiomyosarcoma: a Case Report

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ABSTRACT

We present the rare case of a 61-year-old female patient who was submitted in the hospital with metrorrhagia and pelvi-abdominal pain. Echographic examination revealed an heterogeneous uterine mass measuring 190/130/110 mm. Therefore, total hysterectomy with bilateral ooforectomy was performed. Grossly, the uterus presented a tumoral mass with areas of hemorrhage, necrosis and abundant mucoid degeneration. On light microscopic examination a malignant neoplastic proliferation with features of myxoid leiomyosarcoma was noted. In this paper, we presented this rare form of leiomyosarcoma with an emphasis on its particularities that have not been mentioned in the literature.

INTRODUCTION

Myxoid leiomyosarcoma is a rare variant of uterine leiomyosarcoma characterized by bland cellularity, myxoid matrix and aggressive behavior (1).

Leiomyosarcoma is the most common uterine sarcoma and accounts for 1% of uterine malignancies (2). Few cases of myxoid leiomyosarcoma were reported in the literature and the data was insufficient for conclusive epidemiological studies regarding the incidence and prevalence of this pathology. The preexisting leiomyomas does not represent a risk factor for this malignancy (3).

The reported a five-year survival rate of 15-35% with a more favorable prognosis for patients showing stage I tumors and for premenopausal females (4).

In this paper is presented a rare case of uterine myxoid leiomyosarcoma with its histological and immunohistochemical particularities.



CASE REPORT

A 61-year-old female patient accusing metrorrhage and pelviabdominal pain presented at the emergency room of the University Emergency Hospital in Bucharest. The patient was submitted in the Obstetrics-Gynecology department for further investigations. In anamnesis, the patient mentioned a cervix conization procedure which was performed at 45-year-old. Speculum examination reveals yellow fluid discharge. On vaginal palpation a long cervix is described and on bimanual examination the annexes are not palpable, but the uterus appears enlarged (200/150/100 mm)

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with tumoral transformation and a hard consistency. Serology blood tests revealed hypochromic microcytic anemia. A heterogeneous myometrium mass measuring 190/130/110 mm was seen on transvaginal echography.

After the surgical evaluation, total hysterectomy with bilateral oophorectomy was performed in order to treat the uterine tumor. The surgical specimen is send to the Pathology Department of the hospital and was fixed in 10% buffered formalin for 24 hours. Following macroscopic examination, the tissue fragments were processed and embedded in paraffin. Afterwards, three-micron thick sections were stained with hematoxylin and eosin.

Immunohistochemical study was made on unstained paraffin sections using an indirect trisadial Avidin-Biotin complex method. The

sections were deparaffinated in toluene, dehydrated in alcohol series, rehydrated and washed in phosphate buffered saline. Then, they were incubated with primary antibody overnight, washed with carbonate buffer and developed in 3,3'-diaminobenzidine hydrochloride/hydrogen peroxide nuclear counterstaining with Mayer's Hematoxylin. The following markers were used: desmin, myogenin, SMA (smooth muscle actin), CK-19 (cytokeratin-19), Ki-67, CD10, protein S-100 and CD34. The immunoreactivity was evaluated as follow: diffuse positive (>75% positive cells), positive (25-75% positive cells), focal positive (<25% positive cells) and negative.

To calculate the mitotic index, the area with the largest number of mitotic figures was used for the count of mitoses per 10 high-power fields. □

RESULTS

Grossly, a uterine mass measuring 170/70/60 mm is described. The tumor is yellow-grey-tan in color with areas of hemorrhage and foci of necrosis representing 20% of the tumor mass as well as areas with abundant myxoid degeneration.

On microscopic examination, the presence of a malignant smooth muscle cell proliferation is recorded. The neoplastic cells are spindle with pale-eosinophilic cytoplasm and indistinct cell membrane. The cells are arranged in fascicles, cords and are surrounded by a myxoid matrix. Moderate cytological atypia is present: minimal/absent cell pleomorphism, diffuse hyperchromatic and pleomorphic nuclei with atypical mitosis. The mitotic index accounts 20 mitotic figures per 10 HPF. The presence of tumor cell necrosis is noted.

Considering the diagnosis criteria (abundant myxoid matrix, moderate atypia, cellularity ranging from low to high, tumour cell necrosis evidence and high mitotic index), the final diagnosis is moderate-differentiated myxoid leiomyosarcoma (G2 - differentiation grade).

Immunohistochemistry examination reveals the positivity for smooth muscle cell differentiation markers with diffuse positivity for desmin and focal positivity for SMA. Also, positivity for myogenin is noted. CK-19 is focal positive. Ki-67 is 40% positive which indicates the aggressiveness of the tumor. CD10, S-100 and CD34 are negative. □

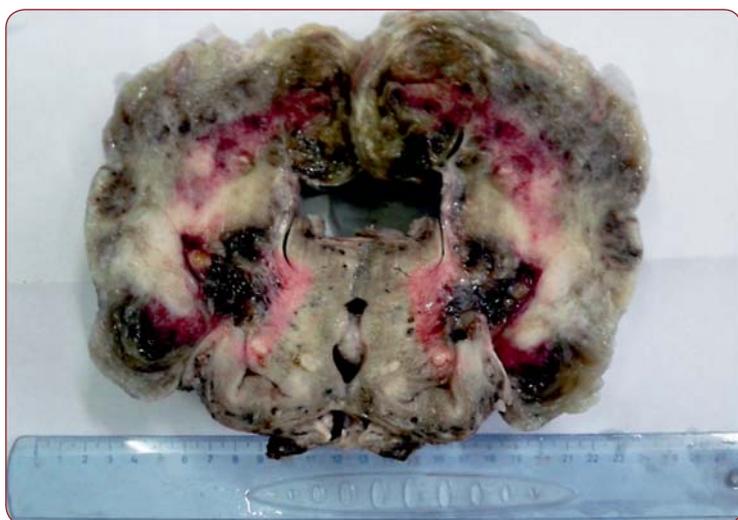


FIGURE 1. Uterine tumoral mass with areas of hemorrhage and necrosis.

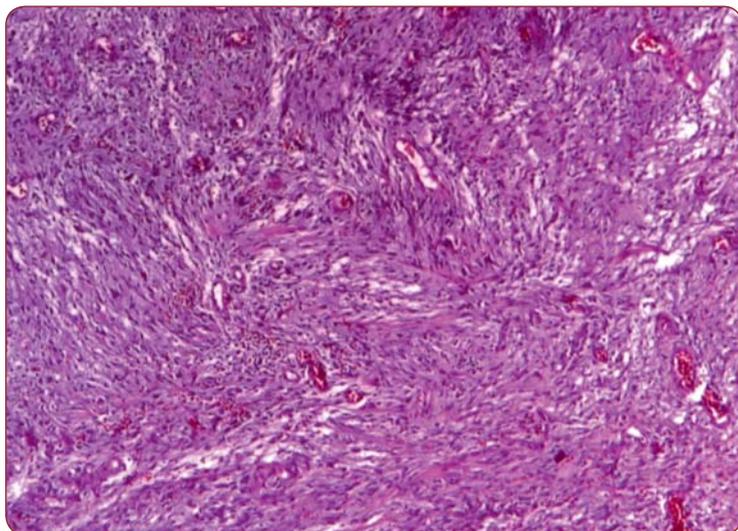


FIGURE 2. Smooth muscle cell proliferation enclosed in a myxoid matrix.

DISCUSSION

Myxoid leiomyosarcoma is a malignant smooth muscle cell proliferation with an abundant myxoid matrix. They are characterized by low cellularity and low mitotic index (4). Because of the hypocellularity and the bland cytological aspect, the myxoid leiomyosarcoma represents a diagnosis challenge. For an accurate result, multivariate diagnosis criteria are taken into consideration, such as: mitotic index value, the grade of nuclear atypia and the presence of tumor cell necrosis (5,6). In our case, the tumor has areas that shows increased cellularity which determines a higher mitotic index.

This malignancy affects females with a mean age of 54 (a decade later than the mean age for leiomyomas).

Important prognosis factors for this tumors are the extrauterine extension of the tumor, vascular invasion, maximum diameter of the tumor and the mitotic index value (2,7,8). Even though the mitotic index is low, the prognosis is poor. Also, the 5-year survival rate is higher than in conventional leiomyosarcoma (6,9). This implies a worse diagnosis for our patient given the histological features and the high mitotic index.

If the tumor is less differentiated, the smooth muscle cell markers immunopositivity is weaker (4). In our case, the histologic differentiation of the tumor explains the focal positivity for SMA.

Because of the treatment and prognosis implications, a differential diagnosis with myxoid leiomyoma and inflammatory myofibroblastic tumor must be taken into consideration. Myxoid leiomyoma is a benign smooth muscle cell proliferation. Cytological atypia are absent and, rarely, minimal. Often, mitosis are absent. In-

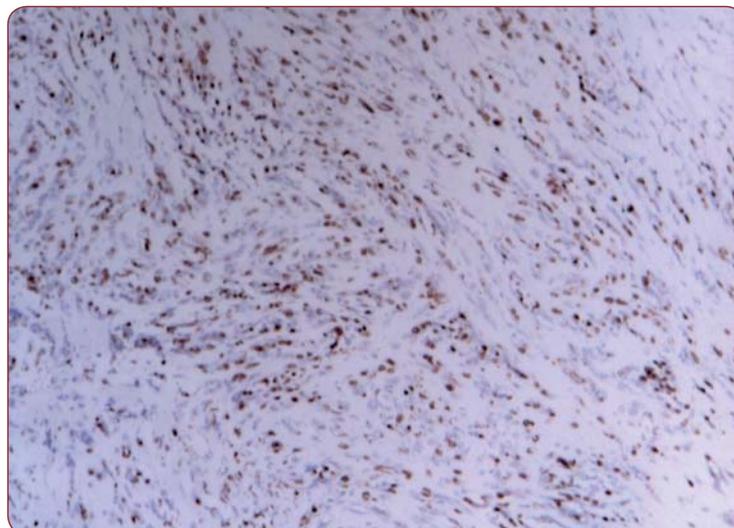


FIGURE 3. Positive tumoral cells staining for Ki-67.

flammatory myofibroblastic tumor has a maximum of 10 mitosis/10 HPF with no atypical mitotic figures. Usually, a lymphoplasmacytic inflammatory infiltrate is present and tumor cell necrosis is absent. □

CONCLUSION

Based on the studies performed so far, myxoid leiomyosarcoma is very rare. Because of its characteristics, this tumor has a poor prognosis. The particularity of the presented case consists in the high mitotic index and high cellularity areas, features that do not characterizes the myxoid leiomyosarcomas mentioned up to this point.

Conflict of interests: none declared.

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