A RARE CASE OF MALIGNANT MESENCHYMOMA OF THE UTERUS

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Abstract:
Malignant mesenchymoma is a rare tumor and location in the uterus is even rarer. We describe a case of malignant mesenchymoma of the uterus in a postmenopausal female.

Keyword: Malignant mesenchymoma, leiomyosarcoma.

INTRODUCTION:
Malignant mesenchymoma is a rare malignant tumor, which by definition shows differentiation into at least two different malignant mesenchymal components\(^1\). The most common location is in the retroperitoneum\(^2\), but occurrences in several other locations have been reported. Malignant mesenchymoma arising from the uterus is extremely rare and information about its management is lacking. Despite the fact that uterine leiomyomas are very common neoplasms, malignant transformation of these tumours is exceedingly rare and it is assumed that most uterine leiomyosarcomas arise de novo. Here we report a rare case of malignant mesenchymoma with leiomyosarcomatous and chondrosarcomatous areas (which can also be called as leiomyosarcoma with heterologous differentiation).

CASE REPORT
55 years old postmenopausal female presented with lower abdominal pain and retention of urine. She had excessive bleeding with passage of clots per vaginum since 3 weeks. Clinicians found a polyp protruding from the cervix. Polypectomy was attempted with the presumptive diagnosis of cervical fibroid polyp. Since there was a heavy bleeding the procedure was converted into a total abdominal hysterectomy.

GROSS FINDINGS
We received total abdominal hysterectomy specimen showing normal endometrium and myometrium with a defect at the level of the cervix. Also multiple grey white masses were received separately with the largest mass measuring 10x8x4 cm which had a white & firm cut surface. Other masses were friable, greyish white and fleshy with focal firmer areas. Representative samples have been taken.
from different areas and processed.

**Figure 1**: Uterus with a large defect at the level of the cervix

**Figure 2**: Grey white soft tissue masses with some having fleshy and some having fibrous appearance

**HISTOPATHOLOGICAL FINDINGS**

Sections from the greyish white masses revealed a neoplasm composed of fascicles and sheets of spindle cells with moderate eosinophilic cytoplasm and elongated nuclei. Most of the areas showed diffuse nuclear atypia with hyperchromasia, bizarre nuclei, atypical mitotic figures and tumor giant cells. Mitotic count was 20 per 10 high power fields. Extensive areas of coagulative necrosis were found. In some areas the tumour had features of benign leiomyoma. Histologically benign smooth muscle merging with malignant smooth muscle cells formed the bulk of the tumour. There was also evidence for divergent mesenchymal differentiation in the form of chondrosarcomatous differentiation which is seen admixed with the spindled malignant cells. The tumour was seen to be arising from the cervical stroma and the uterus was normal. Ex-

**Figure 3**: Spindle cells arranged as fascicles – 4X

**Figure 4**: Neoplastic cells exhibiting bizarre hyperchromatic nuclei and mitoses – 10X
Figure 5: Spindle cells with nuclear atypia and mitoses – 40X

Figure 6: Cells having bizarre nuclei and tumour giant cells – 10X

Figure 7: Coagulative type of necrosis which is sharply demarcated from the neoplasm – 4X

Figure 8: Malignant cartilage merging with adjacent spindle cells – 40X

Figure 9: Malignant cartilage – 40X

IMMUNOHISTOCHEMISTRY:
We have done vimentin, desmin and smooth muscle actin which showed diffuse strong positivity in the malignant spindle cells of the tumour. Chondrosarcomatous areas showed focal positivity for S-100 and vimentin. Cytokeratin and myoglobin were negative.
Uterine leiomyomas are extremely common neoplasms that may be asymptomatic or cause abnormal uterine bleeding or abdominal discomfort. Malignant transformation of a benign leiomyoma into a leiomyosarcoma is an extremely uncommon event and it is assumed that most uterine leiomyosarcomas arise de novo. However, documented cases of malignant transformation of leiomyomas do exist. The term ‘malignant mesenchymoma’ was introduced by Stout [1] to define tumors of the soft tissues of mesenchymal origin, which are composed of two or more cell types any of which, if taken by itself might be considered as a primary malignant neoplasm.
Since fibrosarcomatous areas were noticed in most of the mesenchymal tumors, Stout [1] noted that this form should not be counted as one of the two elements required. More recently, Enzinger and Weiss [3] advised the use of Stout's criteria and insisted on the fact that each of the two or more tissue elements has to be sufficiently differentiated to permit clear recognition of its histogenetic type with the light microscopy, immunohistochemistry or ultrastructural examination. In this case we found leiomyosarcomatous and chondrosarcomatous areas in the same tumour thus fulfilling the criteria for malignant mesenchymoma. Post operative ultrasound abdomen revealed no other masses. Distinction between metaplasia in a malignant mesenchymal tumour and divergent mesenchymal stem cell differentiation is interpretative and not necessarily exclusive. Divergent differentiation of stem cells, for instance resulting from mutations in genes controlling differentiation, may thus result in different phenotypes. Malignant mesenchymomas with osteosarcomatous, chondrosarcomatous and liposarcomatous areas have been reported in the literature [4]. Malignant mesenchymomas have a propensity to occur in the retroperitoneum [3]. Other reported locations of malignant mesenchymoma include thigh, lower leg, lung, orbit, larynx, heart, liver, pleura, spermatic cord, urinary bladder, prostate, kidney, and even an incisional scar of the abdominal wall [5]. Although the location and presentation of this tumor are diverse and no common causative factor had been established, there have been reports of malignant mesenchymoma being associated with radiation therapy [6] and prenatal phenytoin exposure [7]. Despite limited experience in most centers, the general consensus is that malignant mesenchymoma is a high-grade neoplasm associated with poor prognosis [8]. Due to its extreme rarity, there is insufficient data to suggest the best modality or combination of treatment for this condition. However, the common treatment so far for malignant mesenchymoma regardless of the location has been complete surgical resection of the tumor, with or without adjuvant therapy [9]. In our case, the major malignant component was leiomyosarcoma. The mainstay of treatment for uterine sarcoma is total hysterectomy and bilateral salpingo-oophorectomy with or without lymphadenectomy [10]. Adjuvant radiation decreases local recurrence rates and may have a palliative role but has not been clearly shown to improve overall survival [10]. Adjuvant chemotherapy has not been shown to improve survival but chemotherapy may be used for recurrent or widespread disease [10,11]. Retrospective studies have shown that the stage of the disease, mitotic count, and age at diagnosis are independent prognostic variables for disease-free and overall survival [12,13]. In summary, malignant mesenchymoma is a rare tumor and the case presented here suggests that mesenchymal stem cells in the myometrium normally differentiating into smooth muscle cells may also differentiate along other lines.

REFERENCES:


3 Enzinger RM, Weiss SW. Malignant soft tissue tumors of uncertain type. In, Soft Tissue Tumors


