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Epethelioid LMS-A case report

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Abstract

Fibroid most commonly presents in the reproductive age group. Leiomyosarcoma of uterus occurs in 1-2 %of uterine sarcoma. Epithelioid Leiomyosarcoma of myometrium is rare malignant tumor that arises from the smooth muscle lining the walls of the uterus. A 45 year old female presented with abnormal uterine bleeding, Ultrasonography showed a fibroid measuring 5x5 cm in body of uterus. Hysterectomy revealed a polypoidal growth in endometrial canal with haemorrhage and necrosis. H & E sections revealed histological features of epithelioid Leiomyosarcoma of uterine wall. IHC confirmed the diagnosis with SMA, ER, PR, Ki-67 markers being positive. Prognosis of this tumor is poor with 5 year survival rate depending on the size and stage of the tumor at presentation. Hence, accurate histopathological diagnosis confirmed on IHC is essential.

Keywords: Epithelioid, Leiomyosarocma, uterus.

Introduction

The most common tumor of the uterus is leiomyoma. Leiomyosarcoma of uterus is rare, it is defined as a malignant tumor composed of cells showing distinct smooth muscle features [1,2]. Leiomyosarcoma is a relatively rare form of cancer, comprising of 5-10 % of soft tissue sarcomas. Leiomyosarcoma of uterus accounts for only 1-2 % of uterine malignancies, with epithelioid Leiomyosarcoma is a variant of LMS. LMS with prominent epithelioid features is a well recognized entity in soft tissues, uterus and other sites. It is distinguished on cytoarchitectural grounds from the majority of Leiomyosarcoma that arise in the uterus. Most occur in women over 40 years of age who usually present with abnormal vaginal bleeding (56 %), palpable pelvic mass (54 %) and pelvic pain (22 %) [3]. Preoperative distinction between Leiomyoma and Leiomyosarcoma may be difficult. Occasionally, the presenting features are related to tumor rupture (hemoperitoneum), extra uterine extension (one third to one half cases), or metastases. Leiomyosarcoma originating from Leiomyoma is rare.

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In the present case, 3 possibilities were considered, epithelioid LMS, sarcomatoid carcinoma and epithelioid carcinoma.On basis of histopathological diagnosis and IHC markers, diagnosis of epithelioid LMS was established.

Case report

A 45 year old female presented with abnormal uterine bleeding. Ultrasonography revealed a fibroid measuring 5x5 cm in the body of uterus. Hysterectomy was done. Uterus measured 10x8x8 cm. Cut section showed dilated endometrial canal occupied by a polypoidal growth measuring 5x5 cm, greyish white cut surface and growth seen infiltrating the myometrium(Fig 1). Cervix, both ovaries and both fallopian tubes were grossly unremarkable. H & E stained slides from the polypoidal growth showed round to polygonal cells showing pleomorphism, inconspicuous nucleoli, separated by septas. Numerous multinucleated cells and large polygonal cells are seen. Mitotic figures are seen (8/ 10 HPF). The tumor showed increased vascularity with areas of necrosis and haemorrhage. The tumor cells were seen invading the myometrium along with presence of vascular tumor emboli(Fig 2).Histological diagnosis of epithelioid variant of Leiomyosarcoma was made.

Immuno histochemistry was done for confirmation which showed Positivity for SMA(Fig 3),Vimentin, ER (Fig 4), PR (Fig 5), Ki-67 (Fig 6).Negative markers were S-100, NSE, Synaptophysin.

Fig 1:Gross , Cut section shows dilated endometrial canal occupied by growth.

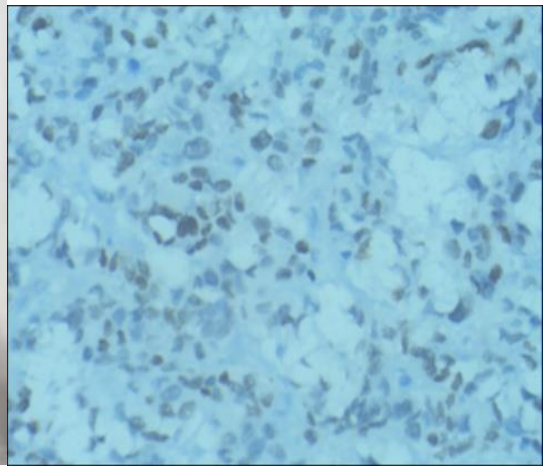

Fig 2:H&E , 400X, section shows pleomorphic Round to polygonal cells, bizarre cells, mitoic activity.

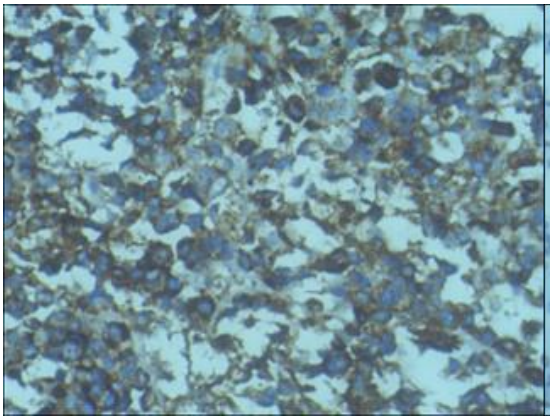

Fig 3: SMA, 400x

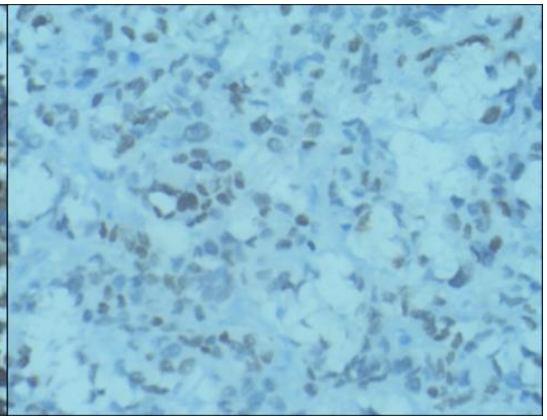

Fig 4:ER,400X

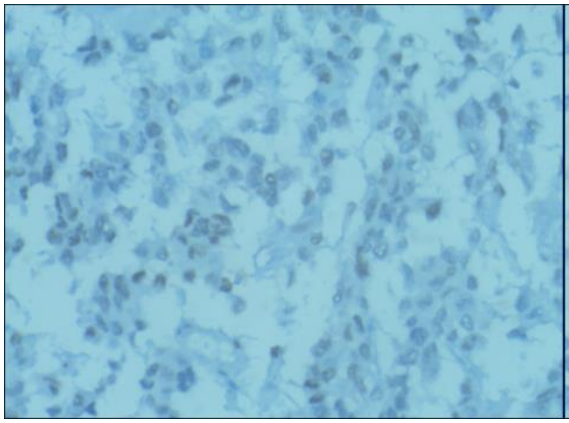

Fig 5: PR 400X

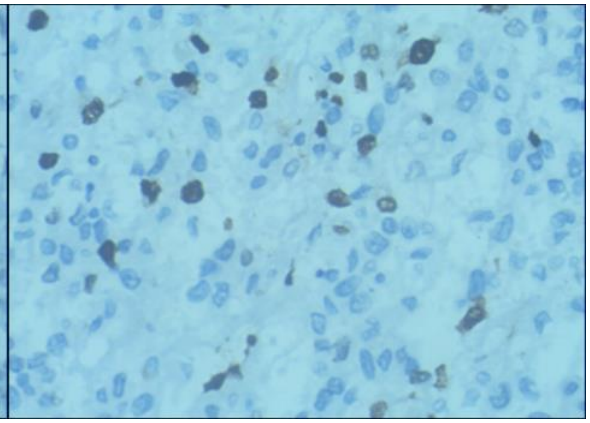

Fig 6:Ki-67

Discussion

The histological diagnosis of the present tumor is difficult. The histology showed spindle cell areas and epithelioid areas. After excluding carcinosarcoma, Leiomyosarcoma is the most common pure uterine sarcoma comprising of approximately 1 % of all uterine malignancies. Most occur in women > 40 years of age who present with abnormal vaginal bleeding (56 %), palpable pelvic mass (54 %) and pelvic pain (22 %). Signs and symptoms resemble leiomyoma [3]. Less common symptoms are weight loss, weakness, lethargy, fever. Occasionally, the presenting features are related to tumor rupture (hemoperitoneum), extrauterine extension (1/2 to 1/3rd) of cases or metastases. Very rarely leiomyomasarcoma originate from a leiomyoma.

Uterine LMS are a rare and aggressive form of uterine cancer as compared to more common endometrial carcinoma and have poorer prognosis. Uterine LMS comprise 1 % of patients with uterine cancers, annual incidence is 0.64 per 100000 women [4,5].

They have high metastatic potential with 5 year survival rates of 0-73 % [6,7].

5year survival rate is 50-65% in stage I, 0-20 % in more advance stage at the time of diagnosis.

Poor prognosis is due to two factors- high incidence of recurrence and ease with which the disease can spread to other organs through blood and lymphatic systems [8]. Diagnosis is usually not made before surgery, so many patients present with advanced disease.

The histopathological diagnosis of uterine LMS is based on hypercellularity, severe nuclear atypia, high mitotic rate (> 15 mitotic figures / 10 HPF) [9,10]. One or more supportive clinicopathologic features such as peri- or post menopausal age, extra uterine extension, large size (>10 cm), infiltrative border, necrosis, atypical mitotic figures are frequently present. Epithelioid and myxoid LMS are two rare variants which are difficult to recognize microscopically as their pathologic features differ from spindle cell LMS. Nuclear atypia is usually mild in both tumor types and mitotic rate is commonly 3 mitotic figures/ 10 HPF.

In Epithelioid LMS , necrosis may be absent and myxoid LMS are often hypocellular . In absence of severe cytologicatypia and high mitotic activity, both tumors are diagnosed as sarcomas based on their infiltrative borders [11]. Differential Diagnosis of Epithelioid LMS areEpithelioid sarcoma, sarcomatoid carcinoma and epithelioid carcinoma. Epithelioid sarcoma, occurs mostly in the extremity and shows characteristic granulomatous appearances with mild atypia, with absence of smooth muscle antigens.

Sarcomatoid carcinoma rarely occurs in the uterus. Almost all cases show squamous cell carcinoma with sarcomatous changes with absence of smooth muscle antigens. So possibility of epithelioid LMS is most likely as there is presence of smooth muscle actin, smooth muscle actin (HHF-35), h-caldesmon which is highly suggestive of smooth muscle differentiation.

Very few cases of epithelioid LMS are reported in the literature. Wang et al reported clinical pathological parameters such as tumor cell necrosis and lymphovascular invasion as the presenting symptom of epithelioid LMS and reviewed 27 cases (17 spindle, 5 epithelioid, 2 myxoid and 3 mixed) of LMS [12]. Toyoshima et al reported a case of epithelioid LMS of uterine cervix [13].

5 year survival rate for small isolated LMS tumor surgically excised with wide, clear margin is 80-90 %. Larger, high grade tumor is likely to recur or metastasize in 80 % cases [8].

Conclusion

Epithelioid LMS is a rare entity with poor prognosis. Hence accurate diagnosis at an early stage is essential for better 5 year survival rate.

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