Int.j.pathol.2017; 15(1)31-34

Case Report

Wide Spread Low Grade Endometrial Stromal Sarcoma

Anum Usman, Noor Khan Lakhnana and Humaira Zafar
Al Nafees Medical College & Hospital, Islamabad

Abstract: Endometrial stromal sarcoma is an unusual malignant neoplasm of the uterus. It mainly occurs in the age of 40–50 years. It accounts 2 to 6% of uterine malignancies. Usually, the diagnosis is confirmed postoperatively. Common symptoms are abnormal vaginal bleeding, abdominal mass and mild abdominal pain. We report a case of endometrial stromal sarcoma in a 40 years old woman. Biopsy specimens were received at the pathology lab of ANMCH. She presented with abnormal vaginal bleeding since 4 months. Histopathological findings confirmed the diagnosis of endometrial stromal sarcoma which was then confirmed on immunohistochemistry. We aimed to present a case of endometrial stromal sarcoma because of its rare occurrence and problems in establishing histopathological diagnosis

Key Words: Endometrial stromal sarcoma, Uterine malignancy

Introduction

Uterine sarcomas are very rare and may arise from endometrial stroma, smooth muscle and connective tissue. Sarcoma of the uterus is an unusual form of malignancy affecting 2 - 5% patients. It has incidence of 1 - 2 /100000 females in overall population.[1]

Uterine sarcomas as a group have tendency for rapid progression along with bad prognosis. Histopathologically, uterine sarcomas are classified as endometrial stromal sarcomas, leiomyosarcomas and mixed mesodermal tumors.[2] The second most common mesenchymal tumor of the uterus is endometrial stromal tumor.[3] It constitutes to about 0.2% of all uterine malignancies and less than 10% of all uterine sarcomas.[3] Endometrial stromal sarcoma occurs mainly in the age group of peri-menopausal women along with one third being in post-menopausal women.[4] The World Health Organization classifies endometrial stromal tumors in four categories i.e. endometrial stromal nodule, low grade endometrial stromal sarcoma, high grade endometrial stromal sarcoma and undifferentiated uterine sarcoma.[5]

The endometrial stromal nodule lies at the benign end of the spectrum of stromal tumors. It is a solitary sharply circumscribed mass that is confined to uterus. It does not invade veins, lymphatics or the myometrium. The prognosis is excellent. Recurrences do not occur after complete removal of the tumor.[6]

AUTHOR'S CORRESPONDENCE
Dr. Anum Usman
Lecturer
Al Nafees Medical College & Hospital, Islamabad
Email: dranumusman@gmail.com

Low grade endometrial stromal sarcoma has less than ten mitosis per ten high power fields. It has propensity to infiltrate the large vascular spaces. Five year survival rate of low grade endometrial stromal sarcoma is 100%.[7]

High grade endometrial stromal sarcoma is categorized as having more than ten mitosis per ten high power fields. It is very aggressive clinical course with a five year survival rate of 55%.[8]

In this case report, we report a 40 years old woman with abnormal vaginal bleeding for 4 months.

Case Report

A forty year old woman presented with abnormal vaginal bleeding since 4 months. Ultrasound and Doppler imaging revealed a heterogeneous mass in the wall of uterus with internal vascularity. Total abdominal hysterectomy with bilateral Salpingo-Oophorectomy was done with clinical suspicion of fibroid.

On gross examination, specimen consisted of uterus, cervix and both tubes and ovaries measuring 11x10x8cm. The outer surface was lobulated. Cut surface of uterus showed slit like lumen and a necrotic whitish firm growth involving whole thickness of wall in a whorling pattern. The upper cervix also appeared to be involved by the growth. One ovary and tube were enlarged in size, ovary measured 5x5cms and tube had a diameter of 3cm. Cut surface of ovary was whitish. Other ovary and fallopian tube appeared normal. There was also a separate nodule in container, measuring 5x4cm with a white cut surface.

Microscopically, multiple sections from nodule revealed a tumor composed of uniform small cells

having round to oval nuclei and scanty cytoplasm. These cells tend to encircle small vessels in a concentric manner typical of endometrial stromal tumors. There were 5 mitosis per 10 high power fields. Vascular invasion was seen. Despite its low mitotic count the tumor was remarkable in its multifocal spread. These multiple foci of tumor sheets had a pushing expansile pattern of growth (Figure 1). The tumor was present in the myometrium and upper cervix and metastasizing to one ovary and fallopian tube (Figure 2, 3&4). Other ovary and fallopian tube were free of tumor.

Immunohistochemical staining revealed that the neoplastic cells were positive for Vimentin, CD-10 and estrogen and progesterone receptors. The diagnosis of low grade endometrial stromal sarcoma with multifocal spread was established.

Discussion

Endometrial Stromal Sarcoma is a very unusual malignant neoplasm with incidence of about 1-2% .[9]

Endometrial stromal sarcomas are divided into different types on the basis of mitotic activity, vascular invasion, presence of hemorrhage and necrosis[10.]

The tumor has a malignant potential and can spread to fallopian tubes, ovaries, ureters, bladder and vagina. *Jain R*, in his study, had reported metastasis of endometrial stromal sarcoma to distant sites i.e. lung, heart and other sites.[11] Involvement of adenexal structures and cervix is also reported as in a case documented by Geetha-P wherein endometrial stromal sarcoma infiltration was seen in the myometrium and the right fallopian tube.[12] However such extensive involvement with low grade tumor like in our case has not been reported.

The common preoperative differential diagnosis of endometrial stromal sarcoma include leiomyoma of uterus and uterine leiomyosarcoma. One study conducted by *Jin Y* in 2010 revealed that diagnosis of endometrial stromal sarcoma preoperately was difficult and most cases were diagnosed as benign leiomyoma preoperatively in his study. So early accurate diagnosis is very essential and also beneficial for the patient survival.[13]

Immunohistochemistry helps in distinguishing the above mentioned differential diagnosis of endometrial stromal sarcoma. Diffuse CD10 is a useful positive marker for endometrial stromal sarcoma whereas leiomyomas are negative for it. *Jassal C*, in his article, had same results of positive immunohistochemical staining for endometrial stromal sarcoma.[14]

As compared to endometrial adenocarcinoma the uterine sarcomas as a group have a worse prognosis. Mukhopadhyay reported two year survival rate of 50% in uterine sarcomas even at an early stage.[15]However the low grade stromal sarcomas are remarkable in having a better survival rate as compared to other uterine sarcomas as documented by *XueW*, [16]therefore early and accurate diagnosis is important for proper patient management.

Conclusion

Endometrial stromal sarcoma is a rare malignant neoplasm. Usually leiomyoma uterus is the preoperative diagnosis and confirmed only after histopathology of uterus received after hysterectomy.

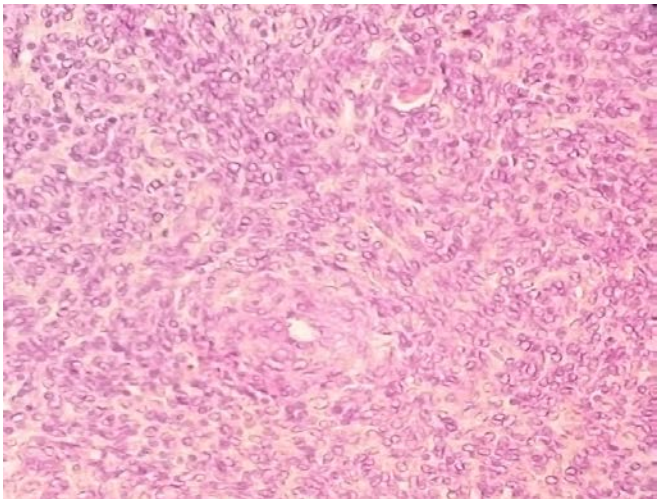

Figure-1: Sheets of bland oval tumor cells arranged concentrically around spiral arterioles (H&E 40x)

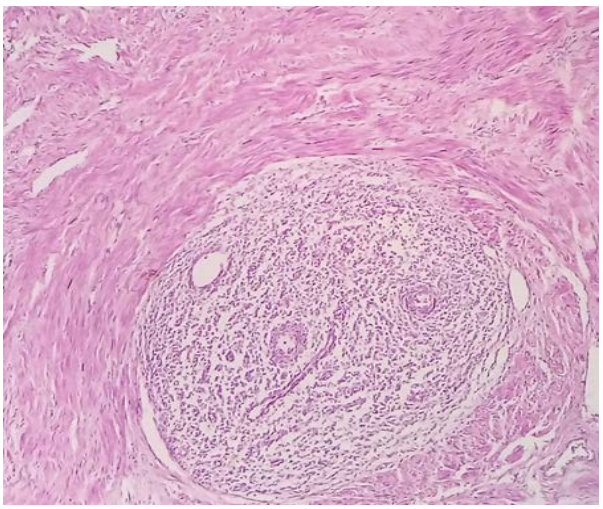

Figure-2: Section from myometrium showing tumor infiltrating the myometrium (H&E 40x)

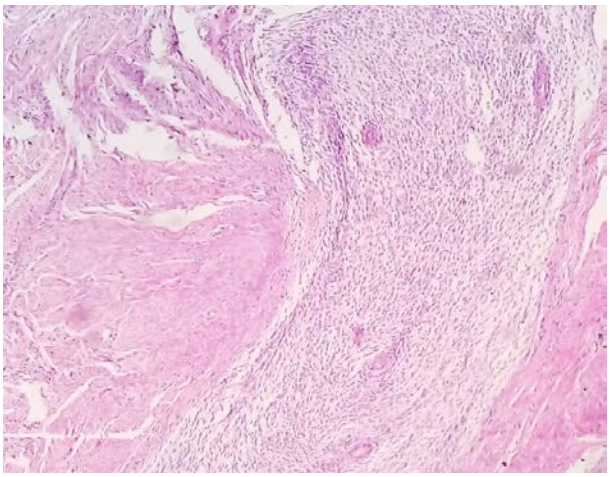

Figure-3: Tumor infiltrates in the cervix stroma. (H&E 10x)

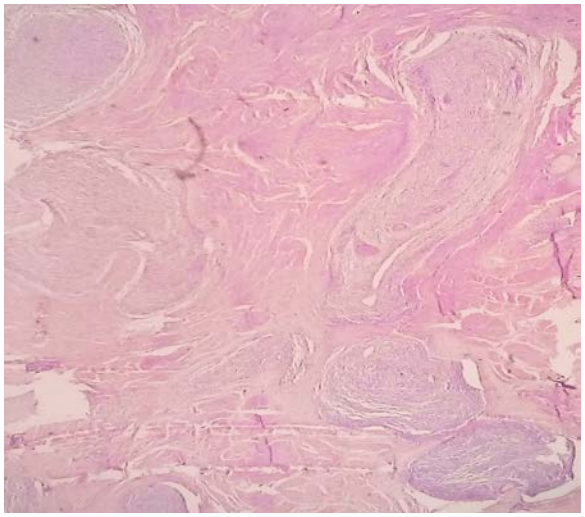

Figure-4: Section from left ovary showing tumor deposits. (H&E 4x)

By reporting our case, we wish to stress the necessity for a high degree of suspicion to diagnose this tumor even in any age group of women. A quick diagnosis and timely intervention are necessary strategies to improve patient's health and survival.

References

1. Jain R, Batra S, Ahmad A, Elahi A, Gupta M andSaith P. Low Grade Endometrial Stromal Sarcoma: A Case Report. Iranian J Med Sci.2015;40(1):81-4.
2. Kim J, Hong S, Sung H, Kyu H and Koh A. A case of multiple metastatic low grade endometrial stromal sarcoma arising from an ovarian endometriotic lesion.JGynecol Oncol.2009;20(2) :122-25.
3. Usha M, Aarathi R, Sujani B and Uravashi T. Low grade endometrial stromal sarcoma presenting as a cervical polyp in a young female: a rare case report. Clin Cancer Invest J. 2014;3(3):257-9.
4. Puliyath G and Nair M . Endometrial stromal sarcoma: A review of the literature. Indian J Med PaediatrOncol 2012;33 (2):1-6
5. Cocklin C and Longacre T. Endometrial Stromal Tumors: The New WHO Classification. AdvAnatPathol. 2014;21(6):383-93.
6. Fdili Z, Chaara H and Bouguern H. Endometrial Stromal Nodule: Report of a Case. Case Reports in Medicine. 2011;2011(1)2:152-59.
7. Kanda M, SonoyamaA,Hirano H, Kizaki T and Ohara N.Transition of low grade to high grade endometrial stromal sarcoma: a case report.Eur J GynaecolOncol. 2014;35(1):4-9
8. Wani R, Bhat N, Nazier J, Bashir H, Jahangeer M, Beigh A et al. Low grade endometrial stromal sarcoma: A case report. J Case Rep OncTher. 2016; 2(1):214-17.
9. Feng W, Hua K, Gudlau E, Yu Y, Zhou X, Baak J. Prognostic indicators in WHO 2003 low-grade endometrial stromal sarcoma. Histopathol. 2013;62(1):675–87.
10. Aseeja V and Taneja B. Endometrial Stromal Sarcoma-A Case Report and Brief Review. ObstetAndGynaecol.2011;2(9):1-8
11. Jain R, Batra S, Ahmad A, Elahi A, Gupta M and Saith P. Low grade endometrial stromal sarcoma: a case report. Iran J Med Sci. 2015;40(1):81-4
12. Geetha P, Rajeshkaran V and Sing S. Endometrial stromal sarcoma. Indian J Med Paediatr Oncol.2010;31(1):21-3.
13. Jin Y, Pan L, Wang X, Dai Z, Huang H, Guo L et al. Clinical characteristics of endometrial stromal sarcomafrom an academic medical hospital in china. Int. J Gynecol Cancer. 2010;20:1535–9.
14. Jassal C, Patnaik B, Divya A and Prasad S. Lowgrade endometrial stromal sarcoma in young age: a clinicopathological report. J ObstetGynaecol India. 2012;62(1):73-5.
15. Mukhopadhyay P, Sharma P, Muraleedharan P and Sarkar S: A case of endometrial stromal sarcoma: J ObstGynecol India. 2008;58(1):73-4.
16. Xue W, Cheung A. Endometrial stromal sarcoma of uterus. Best Pract Res ClinObstetGynaecol. 2011;25(1):719–73

HISTORY	
Date Received:	9-MAY-2017
Date Sent for Reviewer:	11-MAY-2017
Date Received Reviewers' Comments:	25-MAY-2017
Date Received Revised Manuscript:	28-MAY-2017
Date Accepted:	3-JUN-2017

CONTRIBUTION OF AUTHORS	
Author	**CONTRIBUTION**
Anum Usman	A, B, C
Noor Khan Lakhnana	C, E, F
Humaira Zafar	C, D

KEY FOR CONTRIBUTION OF AUTHORS:

A. Conception/Study Designing/Planning
B. Experimentation/Study Conduction
C. Analysis/Interpretation/Discussion
D. Manuscript Writing
E. Critical Review
F. Facilitated for Reagents/Material/Analysis