

Case Report

Endometrial Stromal Sarcoma Mimicking Adenomyosis with Ovarian Carcinoma - A Case Report

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ABSTRACT

Endometrial stromal sarcoma is a very rare malignant tumour that constitutes 0.2 % of all uterine malignancies. It affects slightly younger woman as compared to otherwise uterine malignancies with mean age of 42-52 years. Usual presentation is in perimenopausal age with abnormal uterine bleeding, uterine enlargement. It mimics endometrial carcinoma, leiomyoma, leiomyosarcomas, adenomyosis and ovarian tumour. A high index of suspicion is needed for its diagnosis. Hysterectomy is the treatment of choice. We report a case of 40 year old female patient presenting with lower abdominal pain, menorrhagia and abdominal distension. Ultrasound and CT abdomen and pelvis revealed enlarged uterus with heterogeneous myometrium, subserous fibroid and multilocular cystic mass with solid mural nodules. A diagnosis of adenomyosis with malignant ovarian neoplastic mass was made. Hysterectomy was performed. Histopathological diagnosis was endometrial stromal sarcoma with bilateral ovarian infiltration.

Key words - Endometrial Stromal Sarcoma, Adenomyosis, ovarian carcinoma, leiomyosarcoma.

CASE REPORT

A 40 year old female patient presented with pain in lower abdomen since 5 months, increased PV bleeding during menses and abdominal distension since 3 months. There was no history of fever, weight loss, bowel bladder disturbance. On per abdominal examination, there was uniform distension of abdomen corresponding to size of 36 weeks gravid uterus. The mass was both firm and cystic in consistency, side to side mobility of mass was restricted and lower pole of the mass could not be reached. On PV and per speculum examination, vagina was healthy and cervix was pulled up. Mobility of the mass was restricted. Cervical movements

were not transmitted to mass. Bilateral adnexa could not assessed due to extent of the mass. Rectal mucosa was free on per rectal examination. Mass corresponded to size of 36 weeks gravid uterus. A differential diagnosis of ovarian neoplastic mass, uterine fibroid with cystic degeneration or retroperitoneal mass was made. Laboratory investigations were within normal limits. RFT's, LFT's were normal. CA 125 -52.7 U/ml.

Ultrasound (USG) abdomen pelvis showed an enlarged and bulky uterus with marked heterogeneous echotexture of myometrium with a subserous fibroid. Endometrium was not distinctly visualised. A large multilocular cystic mass was noted

superior to uterine fundus with multiple septations and solid components, measuring approximately 30 (Transverse) X25 (Cranio-caudal C) x18 (Antero-posterior) cm with multiple internal septations and solid components. It was seen occupying almost entire abdomen and pelvis in midline

showing minimal vascularity. Both ovaries were not visualised separately. Both kidneys showed mild fullness of pelvi-calyceal system due to compression of both ureters by the mass. Diagnosis of adenomyosis of uterus with CA ovary was made.

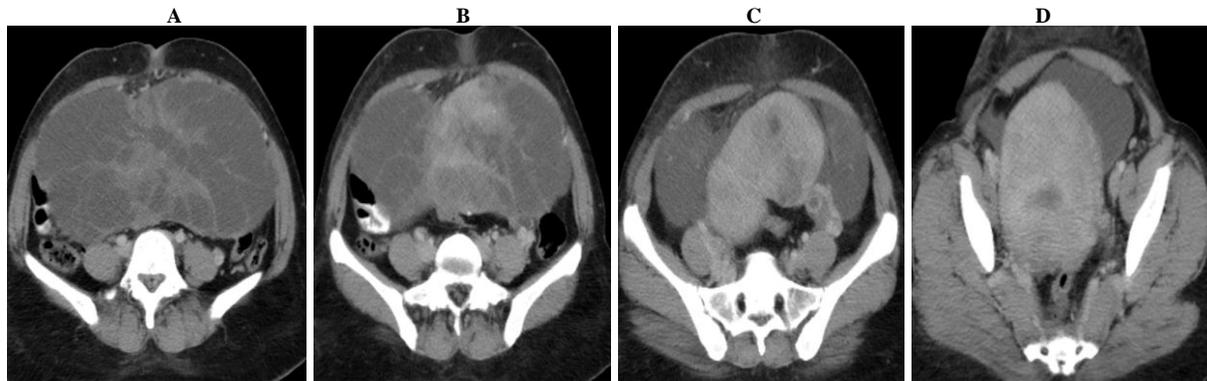


Figure 1 (A-D)- Axial CECT abdomen showing multilocular cystic mass in pelvis superior to uterine fundus with solid mural nodules (A,B); enlarged and bulky uterus with heterogeneous myometrium with subserosal fibroid on right side(C,D).

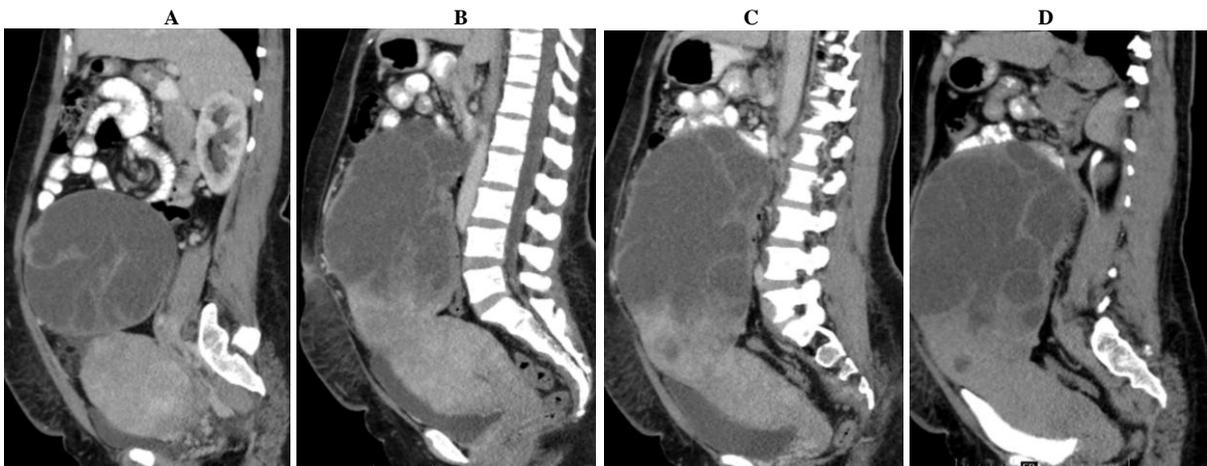


Figure 2 (A-D)- Sagittal CECT abdomen showing multilocular cystic mass in pelvis superior to uterine fundus with solid mural nodules , enlarged and bulky uterus with heterogeneous myometrium with obscuration of intervening fat planes.

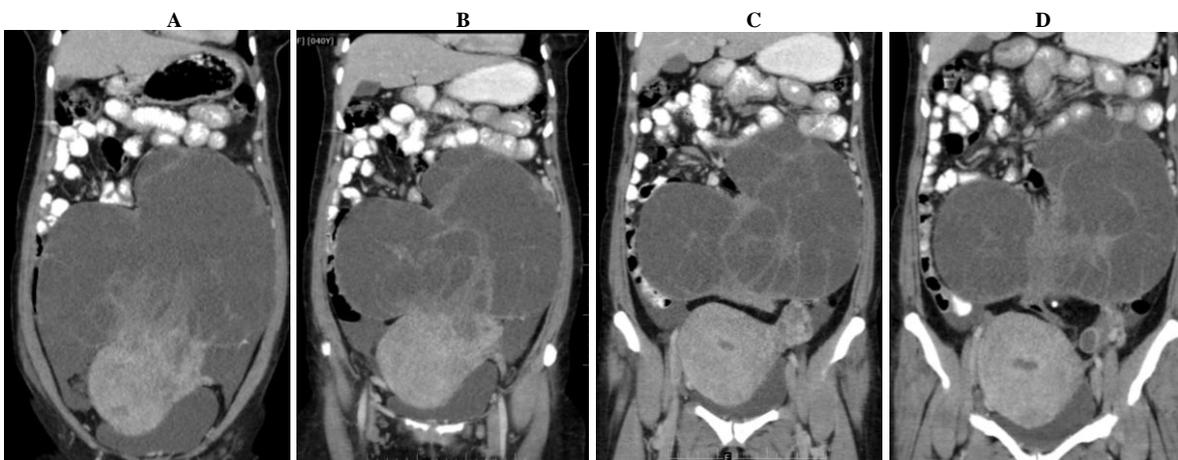


Figure 3(A-D)- Coronal CECT abdomen showing multilocular cystic mass in pelvis superior to uterine fundus with solid mural nodules , enlarged and bulky uterus with heterogeneous myometrium with obscuration of intervening fat planes and mild ascites in bilateral paracolic gutters.

CT abdomen and pelvis (Figures-1,2,3) showed enlarged and bulky uterus with mild heterogeneous myometrium with fundic subserosal fibroid. A large multilocular cystic mass measuring approximately 30 (T)X25 (CC)X18(AP) cm was noted in pelvis just superior to uterine fundus with multiple thin and thick septations and solid mural nodules showing heterogeneous enhancement on contrast study. No calcification was seen. Fat plane between the mass and uterine fundus was obscured. Mild ascites was noted in bilateral lower paracolic gutters. Endometrial cavity was not distinctly visualised. There was no abdominal or pelvic lymphadenopathy. No hepatic metastasis. A diagnosis of adenomyosis with fundic subserous fibroid and malignant neoplastic ovarian mass was made. Intraoperative a large cystic mass was noted arising from uterine fundus which was densely adherent to omentum and bowel. Total abdominal hysterectomy with bilateral salpingo-oophorectomy with omentectomy with lymphnodes resection was done (Figure 4).



Figure 4- Postoperative specimen showing uterus along with cystic mass superior to uterine fundus.

Histopathological examination (Figure-5 A,B) showed small oval to spindle cell resembling endometrial stroma, tongue like infiltration between muscle bundles of myometrium, multiple tumours emboli, infiltration of serosal surface of the uterus by tumour cells. Mitotic figures of 4 to 5/ 10 hpf were seen. Sections from both ovaries showed infiltration by tumour cells and

multiple tumour emboli (Figure-5D). A diagnosis of endometrial stromal sarcoma of uterus with multiple tumour emboli, bilateral ovarian infiltration, with no perineural invasion was made. Parametrium, omentum and surrounding tissues were free from tumour cells Figo staging III A, TNM staging 3A NO MO / stage IIIA.

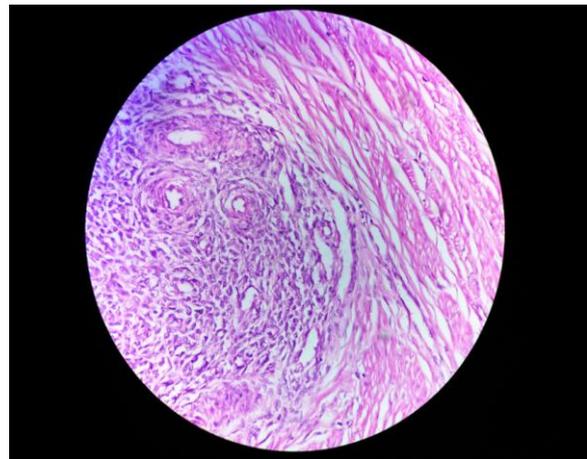
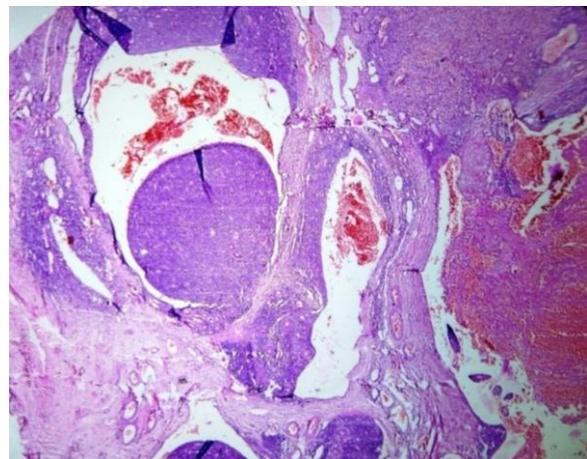


Figure 5-Histopathological examination-

A) High power view-show small oval to spindle cells resembling endometrial stroma, tongue like infiltration between muscle bundles of myometrium, multiple tumours emboli.



B) Ovary-Scanner view- Sections from both ovaries showed infiltration by tumour cells and multiple tumour emboli

INTRODUCTION

Uterine neoplasm can be endometrial or mesenchymal type. Endometrial carcinoma is the most common endometroid tumour of the uterus. Mesenchymal tumours are subclassified as smooth muscle, endometrial stromal and mixed epithelial mesenchymal tumours. [1]

Uterine sarcomas represent 2 to 5% of all uterine malignancies. They are of 3

types- carcinosarcoma (mixed mullerian tumours) which is the most common (40-70%) followed by leiomyosarcoma (40-50%) and endometrial stromal sarcoma which is the least common (<10%) of uterine sarcomas. [2,3]

Endometrial stromal sarcoma is a very rare malignant tumour that constitutes 0.2 % of all uterine malignancies. Its annual incidence is 1 to 2 per million woman. It affects slightly younger woman as compared to otherwise uterine malignancies with mean age of 42-52 years. 10 to 25 % of affected woman are premenopausal. It is an indolent tumour with local recurrence and distant metastasis even after 20 years of initial diagnosis. Mean age at presentation is 42 to 53 years. Patient usually present with pain and abnormal vaginal bleeding. They are typically soft and polypoid and may fill the endometrial cavity. Necrosis and haemorrhage are common. Histologic grades are – endometrial stromal nodal (ESN, benign), low grade, high grade and undifferentiated. The staging of uterine sarcoma is like endometrial carcinoma. Stage I limited to uterine body, Stage II- spread to the cervix, Stage III- spread outside the uterus but within the pelvis, Stage IV- distant metastasis. [4]

DISCUSSION

ESN is a benign tumour composed of cells which resemble normal proliferative phased endometrial stromal cells. ESN is considered as a long as finger like protrusion into the adjacent myometrium are <3mm in depth and <3 in the number with no vascular invasion. More extensive myometrial invasion and endolymphatic and invasion vascular is suggested of LGESS. Low grade ESS is distinguished from high grade ESS by a mitotic rate of <10 mitotic figures (MF) /10 ppf and clinically by a more protracted course. Recurrence occurs late with local recurrence more common than distant metastasis. [6,7]

The pathogenesis of ESS is unknown. Exposure to unopposed estrogen, tamoxifen and polycystic disease of ovaries

are complicated. Latest WHO classification is not made on mitotic count but on the basis of unclear pleomorphism and necrosis. UES (Undifferentiated endometrial sarcoma) represent a high grade sarcoma which bears no histological resemblance to endometrial stroma. Abnormal uterine bleeding is present in 90% of woman while 70 % cases show uterine enlargement. About 30 -50% of ESS has intrauterine spread at the time of diagnosis. The main tumour mass is intramyometrial, most ESS involve the endometrium and hence uterine curettage is useful in preoperative diagnosis. Sometimes the lesion is completely within the myometrium when uterine curettage is not diagnostic. Since ESS has great similarity with normal endometrium, it may not be possible to diagnose with certainty and curettage fragments. Hence definitive diagnosis can be made only on hysterectomy specimen. Rarely it can present at extrauterine site -commonly ovary. It can be primary or metastatic lesion from an occult tumour of the endometrium or from a previous undiagnosed case when hysterectomy was done for uterine leiomyoma. [4] It can extend into adjacent structures like fallopian tubes, ligaments, ovaries and vessels.

Differential diagnosis is uterine leiomyoma, endometrial polyp, adenomyosis, mullerian adenocarcinoma. Extrauterine ESS mimic sex cord stromal tumours of ovary, gastrointestinal stromal tumours, hemangiopericytoma, lymphangiomyomatosis and mesenchymal cystic hamartoma of the lungs. [4]

On microscopy, ESS arises from endometrial stroma (connective tissue) rather than the glands. On macroscopy, it shows polypoid mass extending into broad ligament, ovaries and fallopian tubes. Lymphatic tumour plaques are seen as yellow, rosy and ball like masses. On microscopy, it shows monotonous ovoid cells to spindle cells with minimal cytoplasm, prominent arterioles with angiolymphatic invasion, upto 10-15 mitotic figures per 10 hpf in active areas, tongue

like infiltration in myometrium between muscle bundles, may have foam cells or hyalinization in the stroma and may exhibit epithelioid, myxoid and fibrous change. [8]

Immunoreactivity with antibodies to CD-10 and smooth muscle actin and desmin are used to differentiate between ESS and leiomyoma. Usual treatment is total abdominal hysterectomy with bilateral salpingo-oophorectomy and pelvic and para-aortic selective lymphadenectomy. In low grade ESS and recurrent disease hormonal therapy tamoxifen, gonadotrophin releasing hormone (GnRH) analogues medroxyprogesteron and aromatase inhibitors is suggested. As gynaecologist and radiologist are not much familiar with this tumour, it may be mistaken for endometrial polyp, leiomyoma, adenomyosis, subserous fibroid, leiomyosarcoma of uterus. High index of suspicion is needed in a rapidly enlarging uterus with heterogeneous myometrium and extruterine mass. [4]

On MRI, ESS has typically high signal intensity on T1WI and heterogeneously highly signal intensity on T2WI. The tumour margin are typically irregular with marginal nodular lesion. Intramyometrial worm like nodular extension are often seen with areas of hemorrhage and necrosis. Most of them show marked enhancement than normal myometrium. They have to be differentiated from endometrial carcinoma, leiomyomas with cystic degeneration, leiomyomas. Endometrial carcinomas do not show irregular marginal lesions, intramyometrial nodules or marginal nodules and do not show extensive enhancement as ESS. Leiomyomas usually have regular margins and appear hypointense on T2WI. [9] Leiomyosarcomas may show irregular margins. Adenomyosis affects the myometrium diffusely. It appears hypointense with respect to myometrium on both T1WI and T2WI. [9]

On USG, it can be seen as intramural mass protruding into the endometrial cavity or as intramural mass. The margins can be

well defined or ill- defined. They can be hyperechoic with respect to myometrium or heterogeneous with multiple anechoic cystic areas. Low grade ESS has four patterns on USG – predominantly solid mass with cystic areas, predominantly unilocular cystic mass, a well defined solid mass and an ill defined infiltrative solid mass mimicking adenomyosis. A more nodular or coarse appearance of the myometrium is suggestive of low grade ESS rather than adenomyosis. [5]

CONCLUSION

As it a rare entity and mimic endometrial carcinoma, leiomyoma, leiomyosarcomas, adenomyosis and ovarian tumour (with intrauterine) involvement, a high index of suspicion is needed. Usual presentation is in perimenopausal age with abnormal uterine bleeding, uterine enlargement. Hysterectomy is the treatment of choice diagnosis is done on histological examination. Grades of ESS can be detected by studying characteristics varying of the tumours. Prognosis is better in low grade ESS.

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