Case report

A rare case of primary ovarian leiomyosarcoma during pregnancy in 27 year old patient with 38 weeks gestational age

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Abstract

Ovarian leiomyosarcoma is extremely rare malignant tumour seen in postmenopausal women. Pathogenesis of LMS is still not well understood. A very few cases have been published. We are presenting a case of leiomyosarcoma in pregnant patient. There was a 27 yr pregnant patient at 38 weeks of GA Gestational age. USG revealed a live foetus of 36 wks with normal parameters. There was a huge right adenexal mass with solid-cystic areas extending from pouch of douglous to liver . LSCS with right salpingo-oophorectomy was performed. Histological examination revealed leiomyosarcoma of the ovary. Ovarian leiomyosarcoma may remain assymptomatic in early stage or may manifest with pain in abdomen or loin, weight loss, anorexia etc. LMS is an aggressive tumour and it can occur in reproductive age group also. This extremely rare tumour should be considered in the differential diagnosis of ovarian masses in reproductive age group and pregnant patients also. This case was diagnosed at term stage. If it had diagnosed earlier by sonologist and obstratician ,we would have managed it much better at that stage. So we should be considered ovarian leiomyosarcoma in patients with ovarian masses.

Keywords : Leiomyosarcoma, Pregnancy, Immunohistochemical, LSCS.

Introduction:

Ovarian sarcomas are <3% of all ovarian malignancies[1, 2]. Most common are rhabdo-myosarcoma, fibromyosarcoma, stromal cell sarcoma, and extremely rare sarcomas are Leiomyosarcoma (LMS). Leiomyosarcoma are <0.1% [3] of all ovarian sarcomas. LMS is one of the more aggressive soft tissue sarcoma derived from smooth muscle cells and usually it affects postmenopausal women. We are reporting an unusual case of right ovarian leiomyosarcoma in 27 yr old pregnant patient at 38 weeks gestational age. LMS has poor prognosis because it presents in advanced stage & has an aggressive nature. Survival rates are lowest among all soft tissue sarcomas.

Case report:

Mrs AB a 27 yr old pregnant patient was referred to emergency gynaecology OPD at 38 weeks of GA by last menstrual period (LMP) with acute pain in lower abdomen & right loin since three days. She was G3P2 with previous two caesarean sections (CS). Last CS was three years back. Pain abdomen was continuous, dull aching in lower abdomen and right loin, not associated with any aggravating or relieving factors. There was no significant past medical history. She was booked case in another institute. She had one USG for FWB (done at private hospital)

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at 34 weeks GA, but no comments were made on ovarian tumour. Her pregnancy was uneventful till 38 wks. Her blood group was O positive, HBsAg, VDRL, HIV were non-reactive. Blood sugar was 90 mg%, and haemoglobin was 7.2 g/dl.

On examination patient was conscious and afebrile. Her conjunctiva and tongue were pale. Systemic examination was normal but tachycardia was present. On per abdominal examination uterus was 36 wks, cephalic presentation with floating head, and regular FHS. There were two transverse scars on lower abdomen. Scar tenderness was present on lower abdomen. A bulge, separate from uterine ovoid, was felt in right hypochondrium. It had variable consistency. The limits of mass could not be reached. On PV examination cervical os was closed, uneffaced, and soft in consistency. Intravenous antibiotic was given. Blood was sent for cross match and two units of blood arranged. Urgent USG was done which showed an intrauterine live fetus of 36 weeks in cephalic presentation. A mass of heterogeneous nature, 18×17cm was noted in right adenexa, reaching up to right hypochondrium. Left adenexa appeared normal. Gall bladder, pancreas and bilateral kidneys were also normal.

Patient was posted for surgery by a team of experts which included an obstetrician, general surgeon and urologist. On opening the abdomen peritoneal fluid was aspirated from paracolic gutter and saved for cytology. LSCS was done in usual way and an alive male child of 2.8 kg was delivered. Placenta and membranes were delivered out. Uterus was stitched in two layers. On exploration a mass of 18×16 cm size with variable consistency and intact capsule was seen which was arising from right ovary extending up to right hypochondrium superiorly and up to pouch of Douglas inferiorly. Right fallopian tube was stretched over the mass. Uterus, left ovary and other viscera were grossly normal. Right salpingo-oophorectomy was done after taking informed consent. Haemostasis was achieved. Ureter was identified and found intact. Omental & left ovarian biopsy was taken. Nodes were not palpable. Abdomen closed back in layers. Post operative period was uneventful and she was discharged on tenth postoperative day.

Pathologic examination revealed a tumour composed of interlacing bundles of fusiform cells resembling leiomyosarcoma. There were (>50%) extensive areas of necrosis. Neoplastic cells were round to spindle shaped, with vesicular nucleus, inconspicuous nucleoli & abundant cytoplasm i.e. cells were pleomorphic and hyper chromatic. Mitotic activity was 7-10 per 10 high power fields. Specimen was immunohistochemically positive for smooth muscle actin (SMA) as in (Fig. E), H-Caldesmon, Viamentin, & negative for inhibin, CK-7, CK-20. It was concluded as high grade leiomyosarcoma of ovary with intact capsule. Peritoneal aspirate was negative for malignant cells. Omental and left ovarian biopsy were normal. It was a case of primary ovarian leiomyosarcoma stage 1A with term pregnancy.

Patient was followed weekly for one month. She had no complaint .General physical examination was normal. Per speculum, per vaginal and systemic examination was normal. She was followed monthly for four months. Chest x-ray and CT chest were normal. MRI abdomen and pelvis showed no adenopathy and no ascites. Urinary bladder, bowl loops and uterus were normal. Left ovary showed multiple hyper intense peripherally arranged functional cysts. Right ovary was absent. Patient was posted for total abdominal hysterectomy with left salpingo-oopherectomy.

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Discussion

Primary ovarian leiomyosarcoma is a extremely rare malignancy usually occurs in post menopausal women .To our knowledge such cases of primary leiomyosarcoma with pregnancy at 27yr age has not been reported previously in the literature. "These tumours typically present as a solitary lobular soft fleshy solid mass with haemorrhage and cystic degeneration. Most are unilateral and more than 10 cm in diameter (In this case tumour was 18x16 cm in size). Microscopically these are marked by pleomorphism, abundant (15-30 per 10 HPF) abnormal mitotic figures and coagulative tumour cell necrosis (as in this case shown in Fig. C, C1 & D)"[4].

Pathogenesis of these tumours is not well known. Some theories hypothesize that the tumour originated from hilar blood vessel smooth muscle metaplasia of ovarian stroma or smooth muscle like theca externa cells[2] .Its association with uterine leiomyosarcoma may suggest that they can share same mechanism of development. This theory is explained by rapid growth of such tumours during pregnancy and their positivity for estrogen & or progesterone receptor. Lymph node metastasis occurs very rarely in uterine leiomyosarcoma but most commonly associated with extra uterine disease. So lymph node dissection should be reserved for patients with clinical suspicious nodes. The strongest prognostic variable of LMS is staging of tumour. Patients with early stage disease have a chance of surviving. The treatment of patient with advance or recurrent disease is only palliative. MRI of the lesion and chest CT should be done to evaluate the presence of metastasis in lungs. Treatment of choice is radical surgery f/b adjuvant chemotherapy and radiotherapy. Dixit et al suggest post operative chemotherapy & radiotherapy in large tumour volumes to control local disease & avoid metastasis[1].Rasmussen et al describe the role of repeated cyto-reductive surgery, hormonal treatment and adjuvant ifosfamide in their cases, attributing prolonged survival to surgical intervention.[5] Elizabeth et al describe a case of ovarian leiomyosarcoma and discussed it in supra regional sarcoma meeting. They concluded that intact tumour without metastasis can be resected with close monitoring and imaging .They do not recommended adjuvant therapy.[6]

Conclusion

Primary leiomyosarcoma is rare gynaecological tumour. Very few cases have been published in non pregnant patients. To our knowledge no case has been published of primary ovarian leiomyosarcoma during pregnancy. This prompted us to share this experience. Despite of advances in treatments, leiomyosarcoma are still difficult to treat. During pregnancy symptoms of malignancy as abdominal mass, pain and weight loss, loss of appetite are obscured due to exaggerated pregnancy related symptoms. So with its rarity a gynaecologist should keep it in mind, even in pregnant patient .Extensive sampling along with immune histochemistry studies should be performed to confirm the diagnosis of ovarian tumour. Treatment of leiomyosarcoma should be carried out in specialized centre with expertise in sarcoma care.

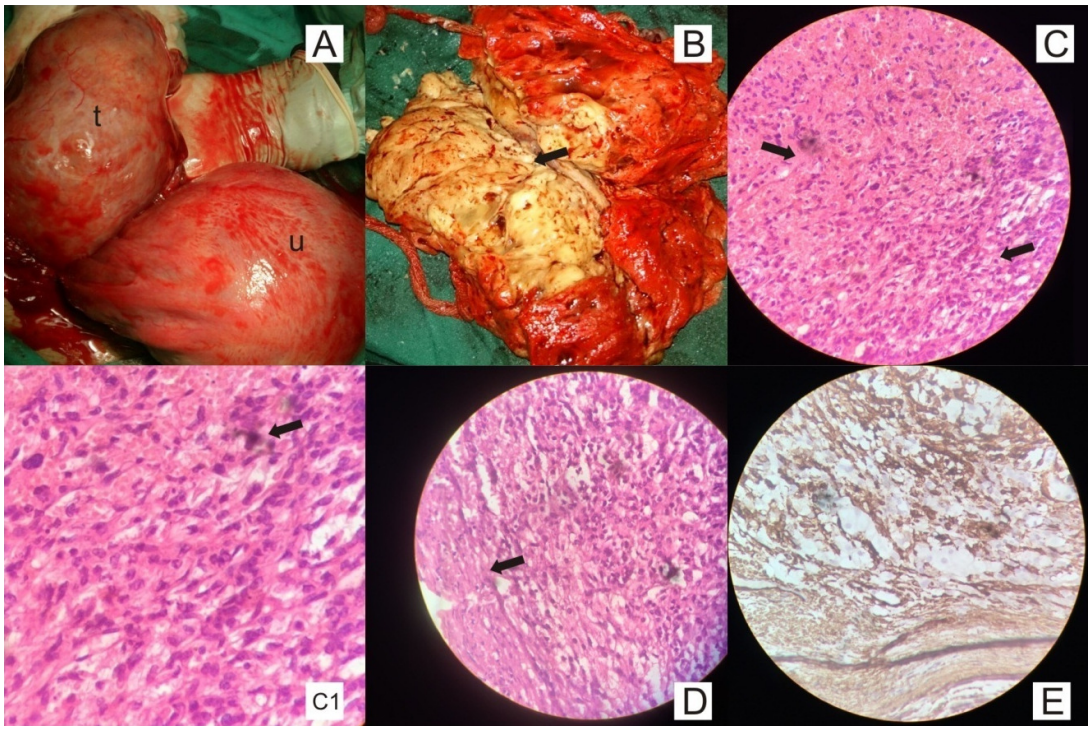

Fig. A Shows intra-operative uterus (u) & right ovarian mass (t); **Fig .B** shows excised tumour with capsule (arrow). Microscopic view **Fig. C & C1** (H &E) low power view (x10) shows extensive necrosis (right arrow) and cells are pleomorphic & hyper chromatic (left arrow). Cells are arranged in fascicles. **Fig. D** shows capsule (arrow). **Fig. E** shows immunohistochemical analysis; tumour cells are positive for smooth muscle actin.

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