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A diagnostic dilemma for the gynaecologist - an ovarian mass with an unexpected twist- a rare case of leiomyosarcoma arising from retroperitoneum

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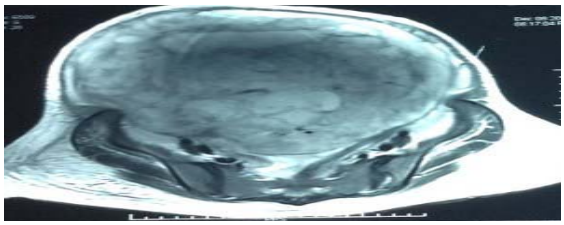
Abstract : Ovarian mass is a commonly encountered clinical condition for the gynecologist. Several tumours in the pelvis and retroperitoneum can mimic an ovarian mass or malignancy and can cause a diagnostic dilemma for the gynecologist. Such tumours need to be dealt by a multi-disciplinary team. This case report highlights about a 30 year old female who was diagnosed to have a huge sub serous fibroid complicating her second pregnancy which increased in size to about 40x30 cm after delivery. Upon evaluation by MRI and CT scan the diagnosis was revised as an ovarian mass occupying entire abdominal cavity from pelvis to the sub hepatic plane. Intraoperatively the tumour was found arising from the retroperitoneum with transverse colon and IVC adhesions with the uterus and ovaries being normal. The tumour was removed and histopathology revealed a leiomyosarcoma. Postoperatively patient recovered and is in continuous followup. This case is reported for the challenge it posed to the gynecologists in the diagnosis and management.

Keyword : Ovarian mass, retroperitoneum, leiomyosarcoma.

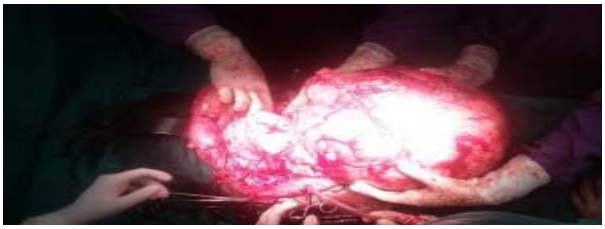
A 30 year female was evaluated at an elsewhere institution for her second pregnancy. Serial antenatal ultrasounds revealed a large subserous fibroid on the right side which was increasing in size. Her antenatal period was uneventful and she had undergone an emergency caesarean section at term for prelabour rupture of membranes and fibroid complicating pregnancy. She delivered an alive female child on 29/10/15. Her post-operative period was uneventful. She noticed a rapid increase in size of her abdomen and pedal edema one and half months after child birth. History of dyspepsia, loss of weight and loss of appetite were present. No history of dyspnea. No bladder and bowel disturbances. Her family history revealed breast carcinoma in her mother. She was breast feeding the baby. Baby expired on 35th day of life due to acute gastro enteritis. She resumed her periods on 03/12/2015. She was evaluated again at elsewhere institution. Her ultrasonogram revealed a large heterogeneous mass occupying the entire abdomen extending from the epigastrium to the pelvis and both flanks with no evidence of free fluid, uterus and ovaries not visualized separately. She had undergone magnetic resonance imaging which showed a large abdomino pelvic mass of mixed signal intensity measuring 32x19x15 cm extending from pelvis to sub hepatic plane on right side displacing the bowel loops. The lesion appears lobulated with cystic changes. The lesion could not be delineated from the mid fundal aspect of the uterus. The impression was given as ? Large subserosal fibroid with degeneration.



MRI picture

With the above investigations in hand, she was referred to our institution for further management. On admission she was cachectic and with severe anemia. No jaundice. Her BMI was 17.2. She was thoroughly investigated in our institution. Her blood parameters revealed Hemoglobin of 6 gm%, leukocytosis, thrombocytosis and hypersegmented neutrophils. Her renal function and liver function tests were normal. Her contrast enhanced computed tomographic images showed a large abdomino pelvic heterodense mass with heterogenous enhancement occupying whole abdominal cavity indenting the undersurface of liver of size 26x14x24 cm, uterus and both ovaries not visualized separately from the mass, the possibility of ovarian malignancy was suggested by the radiologist. Her CA 125 value was 82.26 U/ml (normal less than 35.0) , serum beta HCG was 0.10 mIU/ml, serum AFP was 1.25 IU/ml, serum LDH was 445 U/l (reference range 225 – 450). Oncologist opinion was obtained. Taking into account her CT report and her raised CA 125 values with gross abdominal distension with mass, she was planned for staging laparotomy. Intraoperatively the mass was seen over the Inferior Venecava and just above the right

common iliac vein closely adherent to transverse colon. The uterus and both ovaries were normal. The tumour was removed en bloc and Hemicolectomy and end to end anastomosis were done. Intra operative blood loss was 2000 ml which was adequately replaced. Post operatively, the patient was shifted to high dependency unit and closely monitored. The patient developed infra renal IVC and right common iliac vein thrombosis which was treated with anti coagulants. Post operatively the patient recovered well.

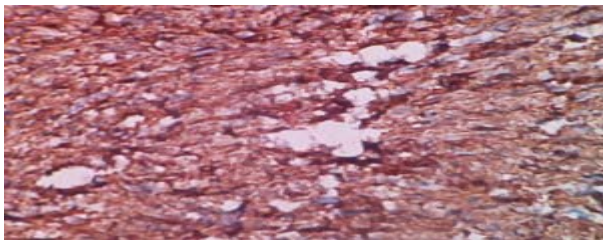


INTRA OPERATIVE PICTURE

CUT SECTION

HISTOPATHOLOGY:

The histopathology was reported as low grade spindle cell sarcoma probably leiomyosarcoma or gastro intestinal stromal tumour. Immune histo chemistry was done with SMA and CD 117. SMA was positive confirming the diagnosis of leiomyosarcoma. The patient was started on adjuvant chemotherapy of ifosfamide, doxorubicin and paclitaxel and is in regular follow up.



IHC - SMA postive

Discussion

Retroperitoneal sarcomas are rare tumours accounting for 1 – 2% of all solid malignancies. Incidence of these sarcomas is 0.3- 0.4 % per 100000 population (1). The most common are liposarcoma and leiomyosarcomas (2). These retroperitoneal sarcomas can pose a diagnostic and therapeutic challenge to the gynecologist because of the nature of these tumours to mimic that of an ovarian tumour which may be difficult to be differentiated by CT or MRI imaging, rarity, late presentation and complex anatomical location(11). Retroperitoneal tumours are commonly misdiagnosed as ovarian pathologies as it is extremely difficult to differentiate from an ovarian tumour as the ultrasound findings and clinical symptoms are very similar (3). Symptoms due to retroperitoneal leiomyosarcomas are vague and appear due to the sudden increase in size or due to compression of retroperitoneal structures. Most patients present with abdominal distension and pain along with a palpable mass, urinary and gastrointestinal symptoms (4). Pedal edema can occur due to compression of the IVC (5). The diagnosis of retroperitoneal leiomyosarcoma is mainly by imaging methods and confirmed by histopathology and immunohistochemistry. Ultrasonogram is the first line of investigation to be done. The investigation of choice for retroperitoneal sarcoma is contrast enhanced computed tomography. It plays an important role in localization, extent, local invasion and assessment of metastases. But imaging features of most sarcomas are nonspecific(6). Nowadays magnetic resonance imaging is playing an increasing role in the evaluation of retroperitoneal sarcomas because of the excellent tissue and spatial resolutions, vascularity, internal characterization (7), (8). Histopathology gives the final diagnosis along with immunohistochemistry with SMA (smooth muscle actin) (9) The management of the retroperitoneal leiomyosarcoma is complete surgical excision with wide clear resection margins. The adherent structures can be removed en bloc to achieve a good oncological clearance. Leiomyosarcoma adherent to the IVC can be treated by resection of the involved IVC with synthetic caval replacement(10). Adjuvant treatment includes chemotherapy with ifosfamide, doxorubicin and paclitaxel. Combined modality treatment of surgery with radiotherapy has been shown to improve the local recurrence rate of the retroperitoneal sarcomas Retroperitoneal sarcomas have a poor prognosis with overall 5 year survival between 39 and 68%. Local recurrence is common in these tumours and is responsible for as high as 75% of sarcoma related deaths Retroperitoneal sarcomas pose a real problem to gynecologists as they mimic ovarian tumour and particularly in antenatal period where they are usually diagnosed as fibroid complicating pregnancy because of the similar ultrasonogram features suggesting degeneration. Although literatures are not aplenty for leiomyosarcomas with hormonal receptors we do have literatures suggesting liposarcomas associated with ER expression associated with pregnancy which directs us for further research for hormonal influence in sarcomas and the consequent trials for hormonal therapy which can be a revolution in the treatment of sarcomas(12),(13)

Conclusion

The gynecologist must be well aware of the fact that a huge retroperitoneal sarcoma must always be kept in the back of the mind for all huge ovarian tumours especially for the one which presents during or immediately after pregnancy. The role of hormonal influence in the sudden increase in size after pregnancy needs further detailed clinical trials which can revolutionize the treatment of these aggressive sarcomas

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