ORIGINAL RESEARCH PAPER

GYNAECOLOGY

Volume : 6 | Issue : 11 | November 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

A case report of low grade Myxoid Leiomyosarcoma

KEYWORDS: leiomyosarcomas,uterine tumours,low grade myxoid variety

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ABSTRACT

INTRODUCTION: Uterine sarcomas account approximately 3% of all uterine cancers and are considered rare. Leiomyosarcomas constitute for 30 % of all uterine sarcomas. Treatment modalities are not standardized and there are few controlled studies evaluating different treatment approaches. CASE SUMMARY: A 45year old female came with complaints of mass per abdomen.On examination tense abdominal distension was present .MRI of abdomen and pelvis reveals a large lobulated predominantly cystic lesion measuring 25x18cms in size with enhancing septations and solid components in the pelvis extending superiorly into the abdomen. CA-125 was 25.9U/ml. Laparotomy revealed a large mass occupying the pouch of Douglas and lateral pelvic wall in the retroperitoneal space. TAH with BSO was done along with removal of the mass.Histopathology report revealed retroperitoneal mass showing features of myxoid leiomyosarcoma-low grade. CONCLUSION: Surgical management remains the mainstay for treatment of leiomyosarcomas.

Introduction: Uterine sarcomas are rare mesodermal tumours that account for approximately 3% of uterine cancers(1).The main lesions are carcinosarcomas, leiomyosarcomas and endometrial stromal sarcomas, the first two have highest incidence in black women in the United States(2).Standard treatment protocols are not available and there are few controlled studies evaluating different therapeutic approaches. Uterine sarcomas can be divided basically into two types-1)Pure-only malignant mesodermal elements are present(eg. leiomyosarcomas and endometrial stromal sarcomas).2) Mixed-malignant mesodermal and malignant epithelial elements are present(eg.carcinosarcomas).

Leiomyosarcomas usually arise denovo from uterine smooth muscle, although rarely they may arise in a pre-existing leiomyosarcomas. Usually occur most commonly in the 45-55 year age group and account for approximately 30% of uterine sarcomas(3). Myxoid leiomyosarcoma is a rare variant of uterine leiomyosarcoma characterised by bland cellularity, myxoid matrix and aggressive behaviour.10-37% of leiomyosarcomas of all tissue sarcomas arise from the retroperitoneum.

The only treatment of is total abdominal hysterectomy and bilateral salphingo-oopherectomy, although in young patients it is reasonable to preserve the ovaries, particularly if the tumour has arisen in a fibroid. Because of the propensity for early haematogenous spread, adjuvant chemotherapy after hysterectomy to eliminate micrometastases is an attractive concept.

Case report: A 45year old came with complaints of mass per abdomen which she noticed one month back, rapid growth in size to attain the present size. She also gives history of associated pain abdomen and distension. She gives history of primary infertility with her married life being 7 years, took treatment for the same for 2 years and then discontinued. She doesn't give any history of weight loss nor decreased appetite. Her menstrual cycles were irregular from past 5years with interval of 30-45 days, changes 3-4 pads per day with passage big clots and spasmodic dysmenorrhea. She didnt give any significant family history of carcinomas.no previous history of any chronic medical illness.

On examination she had pallor confined to lower palpebral conjunctiva, no lymphadenopathy noted. Per abdomen examination revealed abdominal distension, tense with a abdominal girth of 71cm.Fluid thrill was present. On local examination the cervix was flushed with vagina. Uterus size couldn't be assessed. Fornices were free with no tenderness or forniceal mass. Per rectal examination couldn't reveal any pathology.CA-125 was 25.9U/ml.

MRI of abdomen and pelvis reveals a large lobulated predominantly cystic lesion measuring 25x18cms in size with enhancing septations and solid components in the pelvis extending superiorly into the abdomen. The left ovary was not visualised, the uterus and right ovary was pushed to right side. Radiological diagnosis of ovarian cystadenocarcinoma was made.

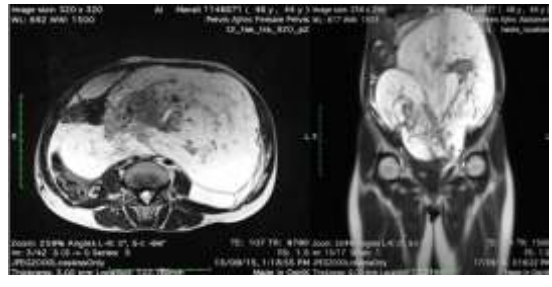

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Figure 1:MRI picture of the large lobulated mass in pelvis with predominantly cystic lesion and solid areas in the pelvis extending superiorly into the abdomen

She was taken up for staging laparotomy. Intraoperative findings were a large mass occupying the pouch of douglas and lateral pelvic wall in the retroperitoneal space. The mass was lying directly over the left ureter and extend into the broad ligament on either side. Retroperitoneal dissection was done to release the mass. Total abdominal hysterectomy with bilateral salphingo-oopherectomy was done along with removal of the mass.

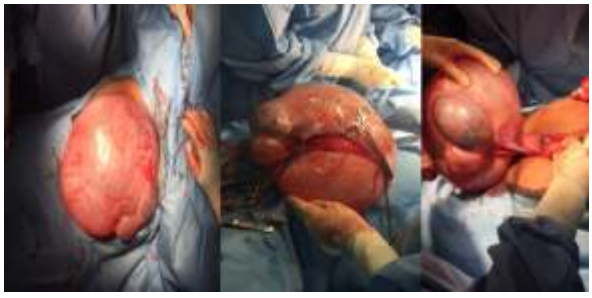

Figure 2: Laparotomy picture showing the large retroperitoneal mass

Histopathology report revealed retroperitoneal mass showing features of myxoid leiomyosarcoma-low grade, cervix showed chronic papillary cervicitis, uterus-subserosal leiomyomata, ovaries and tubes showed normal histology. Peritoneal fluid cytology showed no evidence of malignancy. Patient recovered well in post-operative period and was discharged after 7 days.

Discussion: Based on studies performed so far, Myxoid Leiomyosarcoma are very rare. Treatment is surgical resection. Low grade sarcomas irrespective of histological features overall exhibited good prognosis with median survival of 44 months.

Parra-herran et al(4) in their article titled,"Myxoid leiomyosarcoma of the uterus: A clinicopathologic analysis of 30 cases and review of the literature with reappraisal of its distinction from other uterine myxoid mesenchymal neoplasms" reviewed 30 cases initially diagnosed as uterine myxoid leiomyosarcomas to characterize its clinicopathologic features. They found 11.1% overall survival among cases with five year follow-up, significantly worse than reported survival rates for conventional Leiomyosarcomas. They concluded that myxoid leiomyosarcomas are aggressive neoplasm characterized by infiltrative tumour borders and variability of other features(mitotic count,atypia and necrosis).

Surgical resection i.e Total abdominal hysterectomy with bilateral salphingo-oopherectomy is the mainstay of treatment for frankly malignant leiomyosarcomas although in young patients it is reasonable to save ovaries, particularly if the tumour has arisen in a fibroid (5).A large study of uterine sarcomas recorded in cancer registry of Norway from 1956 to 1992 reported no change in 5 year survival rate after the introduction of chemotherapy into the treatment protocols(3,6).

Conclusion: Accurate diagnosis is critical because therapies may differ widely for conditions which are included in the differential diagnosis of myxoid leiomyosarcoma. The only proven benefit for patients with leiomyosarcomas is total abdominal hysterectomy with bilateral salphing-oopherectomy. Pelvic radiation and chemotherapy are proven to be of no benefit for these patients.

References:

1. Zaloudek CJ, Norris HJ. Mesenchymal tumours of the uterus.In:Fengolio C,Wolff M,eds.Progress in surgical pathology,vol.3,New York:Masson,1981:1-35
2. Brooks SE et al.Survillence,epidemiology ad End results analysis of 2677 cases of uterine sarcoma 1989-1999.Gynaecol Onco 2004;93:204-208
3. Berek and hacker textbook of Gynaecologic Oncology 5th edition. Chapter 10-Uterine cancer.
4. Parra-herran C et al,"Myxoid leiomyosarcoma of the uterus:A clinicopathologic analysis of 30 cases and review of the literature with reappraisal of its distinction from other uterine myxoid mesenchymal neoplasms"Am J Surg Pathol.2016 Mar;4 0(3) :285-301.doi:10.1097/PAS
5. Giuntoli RI et al. Retrospective review of 208 patients with leiomyosarcoma of the uterus: prognostic indicators, surgical management and adjuvant therapy.Gynaecol Oncol 2003;89:460-469.
6. Nordal RR et al.Uterine sarcomas in Norway 1956-1992:incidence.survival and mortality.Eur J Cancer 1997;33:907-911.