CASE REPORT

Pregnancy with a Large Leiomyosarcoma – A Case Report & Literature Review

Nighat Sultana[1], Sadaf Afreen[2], Umbreen Akram[3], Shoaib Nayar Hashmi[4]

[1]Professor Obs & Gynae, Army Medical College/CMH Rawalpindi, [2]Registrar CMH Quetta, [3]Assistant Professor Obs & Gynae, CMH Kharian, [4]Professor of Pathology Deptt CMH Kharian

Correspondence: *Brig Nighat Sultana*
Professor of Gynae & Obs, Army Medical College/CMH Rawalpindi
nighatshafiq88@yahoo.com

Abstract

In the modern era, radiological and pathological investigations have made the diagnosis of gynecological diseases easier and reliable. However, in rare instances, very large masses may mask the actual diagnosis and are mimicked by some other abdomino-gynecological diseases especially in pregnancy, where the diagnosis and treatment options both become very difficult. In case under discussion a very large uterine leiomyosarcoma, presented with second trimester pregnancy. Multidisciplinary team approach and anticipation of such differentials could be helpful in such scenarios.

Key Words: Degeneration (Red) of leiomyoma, Uterine Leiomyosarcoma(ULMS), Mucinous cyst adenoma.

Cite this article as: Sultana N, Afreen S, Akram U, Hashmi SN. Pregnancy with a Large Leiomyosarcoma – A Case Report & Literature Review. J. Soc. Obstet. Gynaecol. Pak. 2018; Vol 8(4):278-280.

Introduction

Uterine cavity pathologies contribute to the majority of gynecological diseases. In this regard fibroid uterus presents as one of the commonest masses with wide array of symptoms including menstrual abnormalities, pain abdomen, compression symptoms of pelvic and leg pain, constipation and urinary problems. Sometimes only as an incidental finding while investigating other gynecological disease, or during caesarian section and laparotomy.[1] It is very rare to find a rapidly growing mass (fibroid) during pregnancy like in this case.

Case Report

A 32 year old woman, primigravida, married for last 1 year, a known case of asymptomatic fibroid uterus for 2 years now presented with gestational ammenorrhoea at 17 weeks with severe pain abdomen and rapid increase in fundal height not corresponding with gestation. Before reaching to CMH Kharian she remained admitted and had conservative management in various tertiary care hospitals of Lahore where she did not get any relief. There was no history of excessive nausea, vomiting, weight loss, anorexia, constipation or urinary symptoms. Menarche was at 11 years of age and no menstrual irregularity was reported. Her past medical, surgical and drug history were not significant. General physical examination was unremarkable except pallor. Abdominal examination revealed a large almost 36 weeks mass arising from pelvis and reaching up to left hypochondrial region. There was tenderness over the mass and it was firm in consistency with dull percussion note with restricted mobility. USG abdomen revealed a large mass occupying the fundal region and upper part of body of uterus, probably fibroid along with 17 weeks intrauterine alive pregnancy. MRI abdomen & pelvis also revealed large mass in left superolateral wall of uterus, most likely fibroid (19.8x16x13 cm) with internal cystic necrotic area, and moderate right sided hydronephrosis with intrauterine fetus. Serum CA-125 was 67.6 (Normal less than 35U/ml), CEA= 3.4 (Normal up to 2.5 ng/ml), serum alpha fetoprotein 64 (Normal less than10 ng/ml). Her hemoglobin was 8.4% which was corrected by 2 Rcc transfusion pre-operatively. Rest of blood investigations were within normal limits. Patient was counseled regarding

termination of pregnancy along with extensive surgery, massive hemorrhage, risk of malignancy and loss of fertility. Exploratory staging laparotomy was planned under general anesthesia. Ureteric stenting was carried out which was possible only on the right side preoperatively. Abdomen was opened through a mid-line incision. Upon laparotomy a gravid uterus with a large fundal mass found to have torsion which was corrected and uterus was taken out of the abdomen cavity the ureteric stent was found on the left side instead of right side. Keeping in view the large fibroid arising from fundal area of uterus, inseparable due to broad base, hysterectomy was performed along with intrauterine fetus and conservation of both ovaries.[2] (Figure 1.1, 1.2).

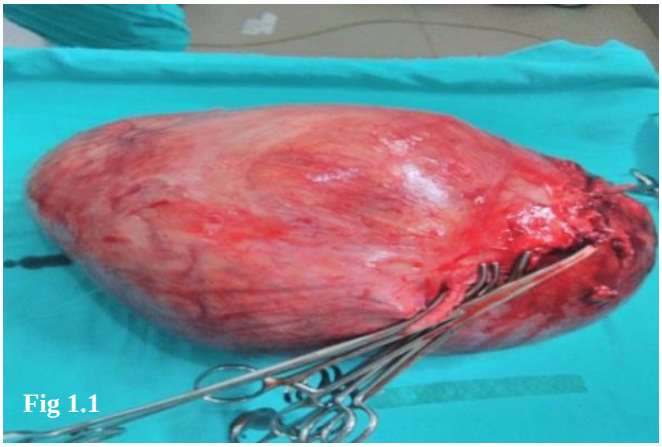

Fig 1.1

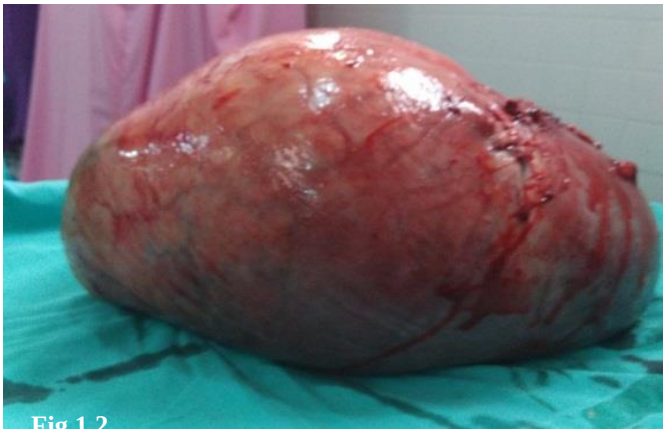

Fig 1.2

Blood loss was moderate. Omental tissue biopsy and peritoneal washings was sent for cytological examination. On gross examination all abdominal visceras were looking normal. Abdomen was closed in layers. Patient was shifted to surgical ITC for further management. Patient had a smooth and uneventful recovery, later discharged and advised follow up with histopathological report. Upon follow up histopathological report revealed that mass as leiomyosarcoma (Stage 1B) with extensive hemorrhage and necrosis on cut sections shown in (Figure 2.1, 2.2), however no malignant cells found in peritoneal washing and omental biopsy. For further advise patient was referred to Shaukat Khanum hospital. She was advised to have follow up visits only without adjuvant therapy was given to this patient.

Fig 2.1

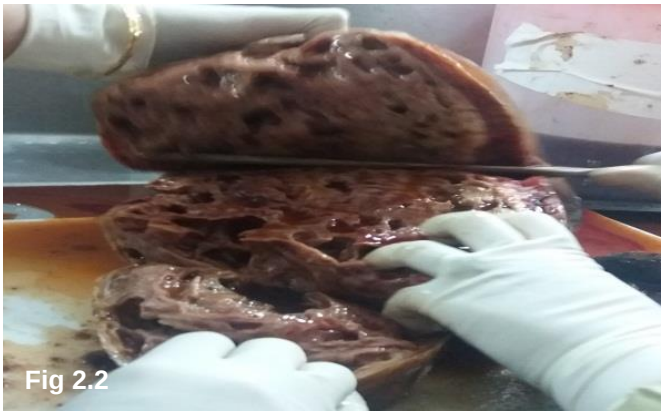

Fig 2.2

Discussion

Uterine fibroids are the most common tumor of female pelvis in the reproductive age group and one of the common cause of hysterectomy as well. During pregnancy hemorrhage, necrosis and degenerative changes in fibroid can complicate the diagnosis.[3] Leiomyosarcoma is a rare entity[4] accounting for approximately 1% of patients with uterine cancer[5] with an estimated annual incidence of 0.64 per 100,000 women.[6] A review by Matsuo et al. covering the period 1955 to 2007 revealed a total of 40 cases of female genital sarcomas diagnosed during pregnancy; 37.5% uterine, 27.5% retroperitoneal, 22.5% vulvar sarcoma, and 12.5% vaginal.[7] Mean age at diagnosis was 27.8 years and the majority of cases were diagnosed in the third trimester. ULMS are

considered neoplasms of high metastatic potential with 5-year overall survival rates varying between 0 and 73%.[8] It mimics leiomyoma but are genetically distinct entities. ULMS are much less common and not hormonally driven and diagnosed only on histopathology. The belief that the risk of ULMS is elevated among women with a "rapidly growing" uterus or leiomyoma was proven false in a study of 1322 women admitted to two community hospitals for hysterectomy or myomectomy. Fibroids rarely, if ever, degenerate into ULMS.[9] Common symptomatology is abdominal distension with pressure symptoms like pain in legs and pelvis, constipation and urinary symptoms along with menstrual irregularities. The typical gross appearance is a large (>10cm), poorly circumscribed mass with a soft, fleshy consistency and a variegated cut surface that is grey-yellow to pink, with foci of hemorrhage and necrosis.[10] Our case presented with short history of rapid abdominal distension and severe pain abdomen. Clinical examination, radiological diagnosis (USG & MRI) pointed towards fibroid uterus with red degeneration but tumor markers raised suspicion of carcinoma like ovarian carcinoma, leiomyosarcoma or choriocarcinoma. Imaging studies and/or clinical findings are not specific for ULMS versus other uterine tumors. Ultrasound examination, magnetic resonance imaging (MRI), or computed tomography (CT) do not reliably distinguish between sarcoma, leiomyoma, endometrial cancer, lymphoma, intravenous leiomyomatosis, or adenomyosis.[11]

Conclusion

Leiomyosarcoma is an aggressive Sarcoma[12] with limited published data that can arise in a number of locations. Although advances have been made in treatment protocols, leiomyosarcoma remains one of the most difficult soft-tissue sarcomas to treat. Hysterectomy is the treatment of choice for large leiomyosarcoma but ovarian conservation is considered in young women.[13,14] Accurate diagnosis, classification, and multi-modality treatment by physicians who are familiar with these tumours are essential for favourable outcome.

References

1. Zimmermann A, Bernuit D, Gerlinger C, Schaefers M,Geppert K. Prevalence, symptoms and management of uterine fibroids: an international internet-based survey of 21,746 women. BMC Womens Health. 2012;12:6.
2. Giuntoli R.L., II, Metzinger D.S., DiMarco C.S., Cha S.S. Retrospective review of 208 patients with leiomyosarcoma of the uterus: prognostic indicators, surgical management, and adjuvant therapy. Gynecol. Oncol. 2003;89:460–469.
3. Muezzinoglu B., Corakci A. Pathological characteristics and clinical outcome of uterine leiomyomas associated with pregnancy. Pathol. Res. Pract. 1996;207(11):691–694.
4. Matsuo K., Eno M.L., Im D.D., Rosenshein N.B. Pregnancy and genital sarcoma: a systemic review of the literature. Am. J. Perinatol. 2009;26(7):507–518.
5. Norris HJ, Zaloudek CJ. Mesenchymal tumors of the uterus. In: Blaustein A, editor. Pathology of the female genital tract. Second ed. New York: Springer; 352; 1982.
6. Harlow BL, Weis NS, Lofton S. The epidemiology of sarcoma of the uterus. J Natl Cancer Inst 76: 399, 1986.
7. Matsuo K., Eno M.L., Im D.D., Rosenshein N.B. Pregnancy and genital sarcoma: a systemic review of the literature. Am. J. Perinatol. 2009;26(7):507–518.
8. Bartsich EG, Bowe ET, Moore GT. Leiomyosarcomas of the uterus: a 50-year review of 42 cases. Obstet Gynecol 71:845, 1988.
9. Jensen ML, Myhre Jensen O, Michalski W, et al. Intradermal and subcutaneous leiomyosarcoma: a clinicopathologic and immunohistochemical study. J Cutan Pathol 23:458, 1996.
10. Kurma RJ. Pathology of the Female Genital Tract, 4th ed. New York, Springer-Verlag; 499.
11. Rha SE, Byun JY, Jung SE, et al. CT and MRI of uterine sarcoma and their mimickers. Am J Roentgenol 181: 1369, 2003.
12. Giuntoli R.L., II, Metzinger D.S., DiMarco C.S., Cha S.S. Retrospective review of 208 patients with leiomyosarcoma of the uterus: prognostic indicators, surgical management, and adjuvant therapy. Gynecol. Oncol. 2003;89:460–469.
13. (Leiomyosarcoma of the uterine cervix associated with pregnancy: A case report and review of literature Dayna D. Whitcombe Philip T. Valente,[c] Ometeotl M. Acosta, and Edward R. Kost , volume 17 Aug 2016 , Gynecologic oncology reports).
14. (Kapp D.S., Shin J.Y., Chan J.K. Prognostic factors and survival in 1396 patients with uterine leiomyosarcoma: emphasis on impact of lymphadenectomy and oophorectomy. Cancer. 2008;112:820–830.