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A Giant Cystic Leiyomyoma with Pseudomeig Syndrome Mimicking Ovarian Malignancy: A Case Report and Review of Literature

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Abstract

Leiyomyomas are an important cause of morbidity and the commonest cause of hysterectomy in the women of reproductive age group. Inspite of modern diagnostic techniques, few leiyomyoma cases may pose a diagnostic dilemma due to varied clinical and radiological presentation. Giant leiyomyoma with degeneration or parasitic myomas are such entities which closely mimic ovarian masses radiologically. Judicious diagnostic approach, conscientious perioperative management and multidisciplinary teambased care are the key for good patient outcome in such cases.

Keywords: giant, leiyomyoma, pseudomeig, ovarian, malignancy.

Introduction

Leiomyomas or fibroids of the uterus is the most common type of benign tumor arising from the uterine smooth muscle. It is believed that both estrogen and progestrone stimulation is responsible for smooth muscle proliferation which leads to leiyomyoma formation. The size of the fibroids can vary from subcentrimetric to giant large myomas occupying the whole of peritoneal cavity. Giant myoma although rare can be potentially life threatening due to the pressure effects on the vital organs. At times, such large fibroids pose a diagnostic challenge to the clinicians due to its varied clinical presentation. This case posed diagnostic dilemma due to clinical and radiological presentation of ovarian mass but per-operatively and on histopathology, it turned out to be a giant leiomyoma attached to uterus with a band.

Case summary

Mrs X, 39 years, P3L3 presented to gynaecology outpatient department of LokNavak hospital, New Delhi with history of heaviness of abdomen which gradually progressed to a large abdominal lump over a period of one year. Patient had become bedridden for last one month due to cachexia and heavy mass which was associated with loss of appetite and shortness of breath. Her menstrual cycles were normal and there was no significant past medical or surgical history. She had three children, all normal vaginal deliveries and last child was 10 years old. There was no family history of malignancies. On examination, her BP was 130/70 mm Hg and pulse rate was 80 beats/min. She had respiratory rate of 20 breaths per minute. On general physical examination, there was mild pallor, no icterus, cyanosis, pedal edema or significant lymphadenopathy. On examination of chest, air entry bilaterally was reduced in the lower zones with coarse crepts. The cardiovascular and neurological examination was normal. The per abdomen examination revealed a grossly over distended abdomen corresponding to term size uterus which was non tender, with smooth surface and firm to cystic in consistency. The lower limit of the mass could not be reached. The patient also had a third degree enterocoele on local examination due to increased intraabdominal pressure. On per vaginal examination cervix was normal in consistency but pulled up. There was fullness in all the fornices and the same mass could be felt through all the fornices. Uterus could not be felt separately. Pervaginorectal examination was normal with free parametrium and rectal mucosa. All routine blood investigations were normal except for haemoglobin report of 9gm%. PAP smear was negative for intraepithelial lesion or malignancy.

The abdominal ultrasound revealed a well defined hypoechoic lesion in the posterior wall of the

uterine fundus measuring 2.7 cm, likely peduncle of the mass. The right adnexa showed a large lobulated heterogenous echogenic mass measuring (22x17 cm) with multiple cystic areas and evidence of central vascularity. Mild free fluid was present in the peritoneal cavity. Uterus was normal but ovaries were not visualised separately. CECT abdomen and pelvis was done which showed a normal uterus & large solid cystic mass of 25.3x18.9x17.8 cm with heterogenous enhancement in the pelvic cavity extending to the peritoneal cavity, displacing the bowel loops posteriorly and laterally and anteriorly abutting the abdominal muscle wall. Multiple enlarged vessels were seen in the mass. There was no calcification or fat attenuation. There was massive ascites with nodular peritoneal enhancement. Both the ovaries were not seen separately.

Based on these clinical and radiological findings provisional diagnosis of ovarian malignancy was made. However, the ascitic fluid analysis was negative for malignant cytology. The tumour markers for ovarian malignancy were normal. CECT chest showed moderate pleural effusion on the right side with underlying basal atelectasis.

To break the dilemma, FNAC and trucut biopsy performed. were А provisional plan for neoadjuvant chemotherapy was made as the patient was unfit for surgery in view of moderate pleural effusion after confirmatory diagnosis on biopsy. However, both FNAC and trucut biopsy showed non malignant features on histopathology. In view of the gigantic size of the mass and its dubious origin, surgical intervention was planned with adequate pre-operative work-up, a high risk consent and building up hemoglobin with one unit of blood.

Exploratory laparotomy followed by total abdominal hysterectomy with bilateral salpingo oophorectomy and pelvic floor repair was done along with excision of the mass in collaboration with the gastrointestinal surgeon. Peroperative blood loss was around 800 ml. Patient was two units of blood; transfused with one preoperative and one postoperatively. Patient

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withstood the surgery well. Per operative findings revealed a large abdominal heterogenous mass solid and cystic areas measuring with approximately 50x50x50 centimetres in size weighing around ten kilograms (figure 1) seen attached to the uterine fundus with a small band (figure 2) but buried into and taking its blood supply from the mesentery, requiring incision from xiphisternum to pubic symphysis. Bilateral ovaries were normal. The histopathology confirmed the diagnosis of giant leivomyoma. Patient was discharged on day fourteen after stitch removal. Post operative period was uneventful.



Figure 1: Giant leiyomyoma with solid and cystic areas measuring approximately 50x50x50 centimetres weighing around ten kilograms



Figure 2: Band attaching the mass to the fundus of uterus

Discussion

Leiomyomas are an important cause of morbidity and the commonest cause of hysterectomy in the women of reproductive age group ¹. Due to the hormonal factor the incidence increases with reproductive age and gradually decreases after menopause. The most common presenting symptom of fibroids is abnormal uterine bleeding which is heavy and prolonged 2 . In addition, women with uterine fibroids may suffer from dysmenorrhoea and non-cyclic pelvic pain³. The giant myomas present more often with pressure symptoms and vague complaints. Grey scale ultrasound is the preferred modality of diagnosis which may further be complemented with MRI if required. On ultrasound, fibroids typically appear as well-defined, solid masses with a whorled appearance; of similar echogenicity to the myometrium or may be hypoechoic. Although fibroids typically have a characteristic appearance on ultrasound, degenerating fibroids can have variable patterns. Only few cases of giant uterine tumors have been reported in the recent literature. The largest uterine fibroid ever reported weighed 63.3 kg and was removed postmortem in 1888⁴. The potential for uterine leiomyomas to grow to an extreme size before causing symptoms is quite remarkable. This is likely due to the relatively large volume of the abdominal cavity, the distensibility of the abdominal wall and the slow growth rate of these tumors. Leiomyomas have been misdiagnosed as adenomyosis, hematometra, uterine sarcoma and ovarian masses.⁵

In 1937, Joe Vincent Meigs (1892–1963), an American professor of the Harvard Medical School of Gynaecology drew widespread attention of the medical fraternity to the Meigs syndrome⁶. The following four characteristics were described by Meigs in 1945 to define the syndrome: The tumour is a benign fibroma or a fibroma like tumour of the ovary (such as the coma and cell tumours), granulosa ascites, pleural effusion(s) and removal of tumour must cure the patient⁷. Other benign cysts of the ovary (such as struma ovarii, mucinous cystadenoma and teratomas), leiomyoma of the uterus, and secondary metastatic tumours to ovarv if associated with hydro thorax are referred to as 'Pseudo- Meigs' syndrome⁸. The concurrent presence of ascites and pleural effusion in the

patient raised a strong suspicion of Meigs/ pseudomeigs syndrome.

Fortunately, intense research has benefitted the treatment of leivomyomas with large a armamentarium of therapeutic modalities in terms of conservative, medical and surgical options. Management options are usually individualized based on the severity of the symptoms, the size and location of the fibroid, thepatient's age and the patient's desire for future fertility ⁹. The current established management of uterine fibroids may involve one of the following approaches or a combination of expectant management, surgical management, medical management or uterine artery embolization¹⁰. For myomas of such large dimensions the mainstay management is surgical. Only the most experienced gynecologic surgeons should attempt such an operation. Even then, intraoperative consultations from gynecologic colorectal, and urology oncology, general, surgeons may be helpful. So, it is always prudent to tie up preoperatively with teams likely to be involved. Intraoperatively, the patient should be positioned to allow adequate ventilation and reduce vena cava compression¹¹. The skin incision should allow easy manipulation of the mass and exploration of the upper abdomen. Preoperative mechanical bowel preparation may decrease the risk of bowel injury and aid per-operative visualisation.

Conclusion

In the era of advanced diagnostic modern medicine, few cases may still pose a challenge to the most experienced surgeons. Although fibroids typically have a characteristic USG appearance, degenerating fibroids can have variable patterns. Clinical modalities, including USG, CT, and MRI can also fail to diagnose the fibroid preoperatively if they are giant as in this case. Surgery remains the mainstay of management. Surgical tie up should preoperatively be done with gastrointestinal surgeon, general surgeon, urosurgeon and vascular surgeon as per case requirement. Experienced anaesthesiologist and

intensive care facility are part and parcel of management. Such cases should be operated with adequate blood and blood products in hand. Keeping all due arrangements prior to surgery results in good outcome as happened in this case. Conscientious perioperative management and multidisciplinary patient care are essential to prevent morbidity and mortality.

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