CASE REPORT

Aggressive Angiomyxoma Pelvis Presenting As An Ovarian Tumor With Vault Prolapse

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Abstract

Angiomyxoma is a rare tumor arising from the pelvis. Preoperative diagnosis and treatment may be difficult. We report a case of aggressive angiomyxoma pelvis that presented as vault prolapse and was misdiagnosed as ovarian tumor. Conclusion: As surgery is the main treatment, preoperative diagnosis and assessment of extent of the tumor for planning surgery cannot be overemphasized.

Key Words

Angiomyxoma, Pelvic neoplasm, Soft Tissue Neoplasms

Introduction

Aggressive angiomyxoma (AA) is a benign, slow-growing tumor that characteristically occurs in women of reproductive age. Local recurrence is cited in 30% to 40% of case (1). Angiomyxoma is a rare pelvic tumor with varied presentation. Preoperative diagnosis and assessment of extent of the tumor are important for its management. We report a case of aggressive angiomyxoma pelvis that presented as vault prolapse and was misdiagnosed as ovarian tumor.

Case Report

A 35-year-old lady presented with complaints of a mass coming out of introitus and abdominal distension for ten years. She had a vaginal hysterectomy and some abdominal procedures done 10, 5 and 3 years ago. However, there were no records. There was no history of loss of appetite or weight lost. Examination showed a large non-tender mass with restricted mobility filling the whole abdomen. Pelvic examination revealed vault prolapse and a right perineal bulge of 4x4 cm. Ultrasound showed a large solid soft tissue mass with few cystic areas and low echogenecity. Computerized tomography scan (CT) showed that the tumor extended from the pelvis to splenic flexure and along posterior abdominal wall, causing bilateral hydroureteronephrosis (*Fig 1*). Fine needle aspiration cytology (FNA) of the mass showed low cellularity. Monomorphic ovoid cells suggestive of a spindle cell tumor were seen. CA125 was 417 IU. An exploratory laparotomy was done along with the surgeon.

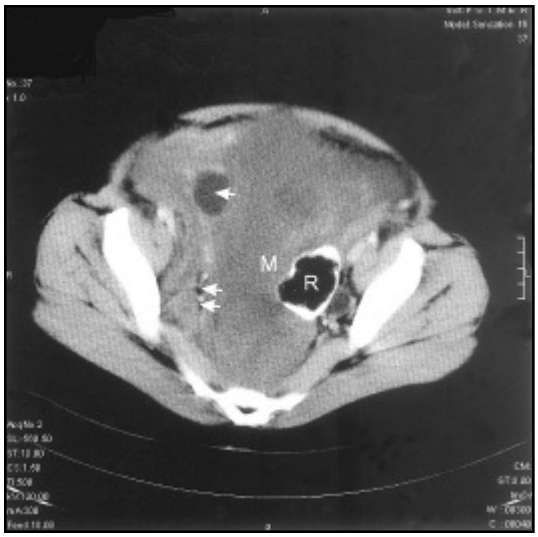

Fig 1. CECT Section Through the Pelvis Shows Heterogeneous Mass (M) with Cystic Component (arrow). Rectum(R) is Displaced to the left. Pelvic Fat (double arrow) is Seen Engulfed in the Mass

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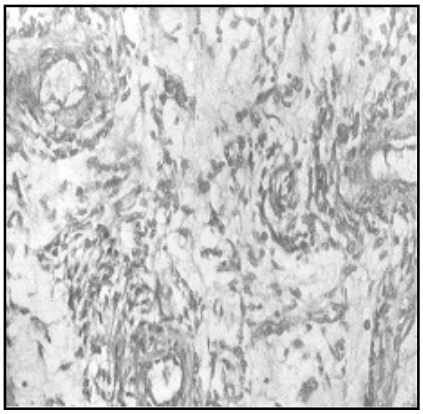

Fig 2. Tissue Section from the Pelvis Mass Shows Varying Sized Blood Vessels Surrounded by Cells with Ovoid to Spindle Nuclei & a Myxoid Stroma (H & E x 280).

A large lobulated, soft, gelatinous mass with solid and cystic areas filling the entire abdominal cavity was found. The left ovary was engulfed by the mass which extended into the left broad ligament, retroperitoneally into the pouch of douglas and into the right obturator fossa. The mass was dissected retroperitoneally while tracing the ureters. Debulking was done, preserving the ureters, bladder and iliac vessels. Few presacral and right iliac vessels were injured. Extensive oozing was controlled by pressure, cautery and gel foam. The patient needed ten units of blood transfusion. Due to excessive blood loss during surgery, diffuse infiltration of the tumor into the bladder and infra levator extension of the tumor, further debulking was defered and abdomen was closed after putting a drain. Postoperatively, she was found to have right ureteric injury at the level of pelvic brim, for which percutaneous nephrostomy was done. The histopathology revealed an edematous and loose myxoid stroma with few proliferative spindle shaped fibroblastic cells devoid of mitotic figures. These findings were suggestive of an aggressive angiomyxoma with involved margins (*Fig 2*).

Postoperatively, C.T. scan done after 3 months showed residual pelvic tumor in the pelvis infiltrating the right ischiorectal fossa, bladder, anal canal, rectum and few small bowel loops. The prognosis and need for extensive abdominoperineal surgery for complete removal were discussed with the patient. She was planned for leuprolide injections that she could not afford. A repeat CT scan done at 5 months postoperatively showed no increase in size. The patient was on close follow up with clinical examination and computerized tomography scan and was planned to undergo abdominoperineal resection in case of progression of disease. However, patient was lost to follow up after 6 months of follow up.

Discussion

Angiomyxoma is a rare mesenchymal tumor arising from the soft tissues of pelvis or perineum that may present as a vulval mass, vaginal polyp, bartholin or a vaginal cyst. It occurs mainly in the reproductive age group (2).The size may vary from 1-60 cm. The diagnosis can be made considering the clinical presentation aided by ultrasound, CT or MRI showing a hypodense mass with translevator extension, displacing rather than invading the pelvic organs (3). FNA reduces the diagnostic possibilities but histopathology alone gives the definite diagnosis. It has low cellularity with spindle shaped cells in a loose matrix containing medium sized arteries having fibrointimal proliferation. Most tumors have been found to have estrogen and progesterone receptor positive status. Tumor cells are atleast focally immunoreactive to desmin, smooth muscle actin, muscle specific actin and vimetin (3).

Surgery is the mainstay of treatment (2). The aim of surgery should be to excise as much as possible without causing urinary or anorectal dysfunction. Recurrence is local and reported in 36-72% (3). Radiotherapy and chemotherapy may not have much role due to the low mitotic activity seen1. GnRH agonist (4) and tamoxifen have been used successfully in few patients. The response can be assessed by clinical assessment, patient's symptomatology and radiographic findings.

References

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