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Case report

AGGRESSIVE ANGIOMYXOMA PRESENTING AS HUGE BROAD LIGAMENT TUMOR- A RARE CASE REPORT

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ABSTRACT

Aggressive angiomyxoma was first described in 1983 by Steeper and Rosai, and fewer than 150 cases have been reported in the world medical literature. Aggressive angiomyxoma is a rare mesenchymal tumor occurring predominantly in the pelvic-perineal region of females of age group 30 to 50 yrs. These tumors are benign and often reach too large dimensions before becoming clinically symptomatic. We report such a case presented as broad ligament tumor, a very unusual presentation. Patient underwent laprotomy and tumor was successfully excised.

Keywords: Aggressive angiomyxoma, Pouch of Douglas.

INRTODUCTION

Aggressive angiomyxomas (AA) are rare, slow growing benign tumors which are, predominantly, located on the perineum of reproductive age women.[1,2] These lesions are characterized as soft, non-encapsulated tumors with finger-like projections infiltrating the surrounding soft tissues. The tumour presents as a large multilobular or polypoid mass or swelling. It is important to diagnose this condition because a tumor is locally infiltrative and surgery is usually the first line of treatment, radical surgery with wide margins and long-term follow-up is advised.[3]

CASE

40 yrs female came to OPD with complain of pain in abdomen, difficulty in passing stools and urine and feeling of abdominal mass since 2 months in OPD of Pravara Rural Hospital, Loni. Dist. Ahmednagar in Maharashtra in India.

Mass associated with pain and bowel complain was initially small and gradually progressed to present size. There was no history of fever, cough, cold, nausea, vomiting, and bleeding per-vaginal, apparent weight loss. Last menstrual period was 10 days back.

Past menstrual history was regular with cycle and flow and there were no clots and no dysmenorrhea.

Patient was Para 2 Live 2. Both were full term normal deliveries & history of tubal ligation done 15 yrs back.

Past, personal and family history was insignificant.

On examination, general condition of the patient was good and vitals were stable. No peripheral lymphadenopathy and other signs of malignancy found.

Per abdomen: Non tender abdominal mass felt corresponding to 24-26 weeks; oval, on Rt. side of lumbar spine, arising from pelvis, slightly mobile, firm in consistency, around 20*15cm. Rt. iliac fossa completely obliterated.

Per vaginal examination: Uterus normal size, firm, mobile, separately felt from the mass which was felt in Rt. Adnexa.

Rt. Adnexa was fixed; bogginess felt in Rt. fornix, Lt. Fornix was relatively free.

Per Rectal examination: large firm mass felt anteriorly free from rectal mucosa, POD was obliterated & mass compressing rectal lumen.

All hematological investigations were done and the investigation reports were within normal limits.

USG suggestive of a large tumor of size 21*20*8cm arising from Rt. Adnexa. Ovaries seen separately from tumor on Transvaginal sonography examination. Uterus of normal size with normal ecotexture of ovaries.

Tumor with some cystic and some hyper lucent areas in between. On Color Doppler vascularity set in.

Fine needle aspiration cytology **(FNAC):** suggestive of Tumor cells cytologically bland and have a spindled, ovoid or stellate appearance with ovoid nuclei and evenly dispersed chromatin. CT scan and MRI evaluations could not be done due to no affordability. CA125 levels was 9IU.

On the basis of the clinical picture and Ultrasound review patient diagnosed as large Rt. Sided Broad ligament tumor.

Exploratory laprotomy under General anesthesia performed. Large whitish shiny tumor nearly of 20*20 cm drawn out of peritoneal cavity which was arising from Rt. Adnexa of the uterus and its extension as peduncle in POD and also in the ischiorectal fossa noted.

With combined surgical approach Tumor excised completely by securing bowel, bladder and ureters.

Along with the tumor uterus with bilateral ovaries which were observed to be normal removed by the conventional method of Hysterectomy. Haemostasis achieved completely with moderate intraoperative loss. No remnants of tumor left.

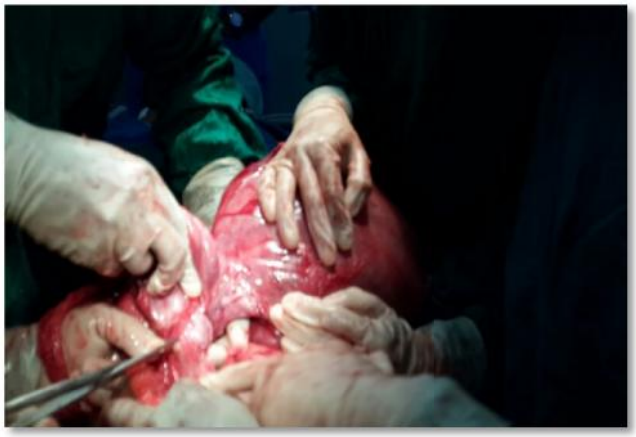

Fig 1: Tumor attached to broad ligament seen.

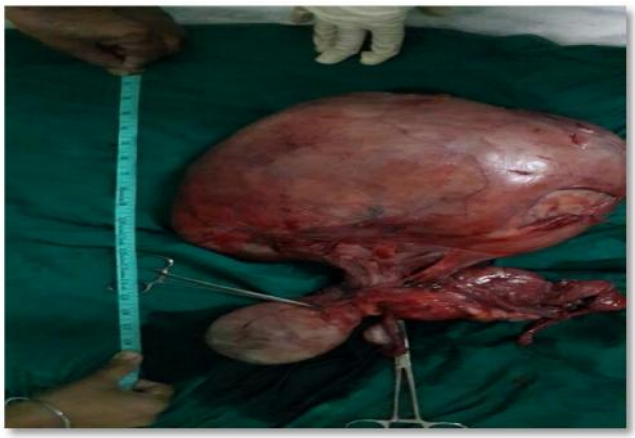

Fig 2: External appearance with pedunculated extension of tumor.

Post-op was uneventful. Repeat haemogram was 11gm%. Patient discharged on 10th Post operative day.

Histopathology gross: Shiny white brown Tumor mass measuring 22*20*7cm arising from lateral Wall of body of uterus. Cut surface: Myxomatous & hemorrhagic areas. Tumor is extended as pedunculated globular 8.5*7*3cm attached to the lower side of uterus near Cervix. Another extended mass 9 *7*2. 5cm noted on the posterior aspect of the Uterus.

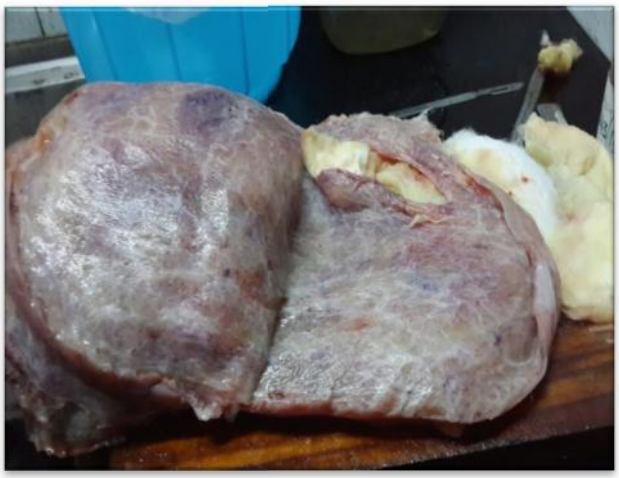

Fig 3: Cut Surface of tumor.

Section from tumor show sparsely cellular tumor composed of numerous haphazardly arranged small to large blood vessels set in myxoid stroma.

Stroma shows collagen fibrils, scattered smooth muscle bundles.

The Tumor cells cytologically bland and have a spindled, ovoid or stellate appearance with ovoid nuclei and evenly dispersed chromatin.

Stroma is distinctly myxoid.

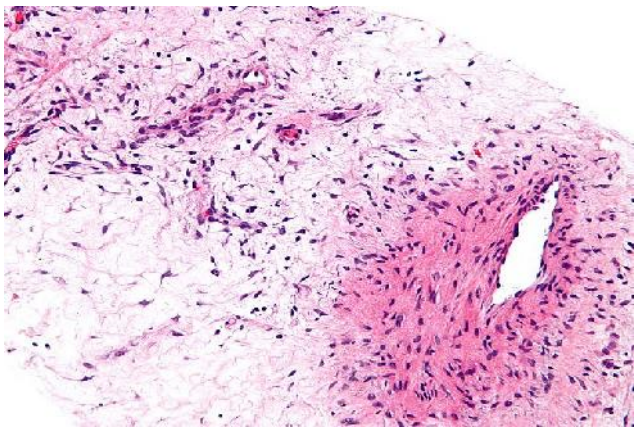

Fig 4: Section from pelvic mass showing varying sized blood vessels surrounded by cells with ovoid to spindle nuclei & a myxoid stroma (H & E x 280).

DISCUSSION

Aggressive angiomyxoma is a rare tumor of mesenchymal origin first described in 1983 by Steeper and Rosai.

The age distribution is wide, with the peak incidence at 31 to 35.[4] Female to male ratio of slightly more than 6:1 . The size may vary from 1-60 cm.[5]

On gross examination it is rubbery and white or soft and gelatinous. The tumor presents as a large multilobular or polypoid mass or swelling.[5]

Locally infiltrative but non-metastasising; that may present as a vulval mass, vaginal polyp, bartholin or a vaginal cyst, ovarian cyst, etc. These lesions are characterized as soft, non-encapsulated tumours with finger-like projections infiltrating the surrounding soft tissues.[6] The tumor grows slowly, and its benign nature is suggested by the histology and by the fact that it shows no tendency to metastasize. However it usually tends locally to recure.[6] The rarity of this condition makes the preoperative diagnosis fairly difficult, It has been also related to hormonal activity which explains female dominance.[7]

The diagnosis can be made considering the clinical presentation aided by ultrasound, CT or MRI shows a hypodense mass with translevator extension, displacing rather than invading the pelvic organs.

FNA reduces the diagnostic possibilities but histopathology alone gives the definite diagnosis.[8]

Differential diagnosis:

Benign: myxolipoma, myxoid neurofibroma and myxoid leiomyoma to myxofibrosarcoma, myxoid variant of liposarcoma, leiomyosarcoma.

Malignant: fibrous histiocytoma and botryoid rhabdomyosarcoma.

The distinctively striking vascular component in aggressive angiomyxoma helps in ruling out most of the above mentioned neoplasms as differentials.

The optimal treatment for AA is wide local excision with tumor free margin, as this tumor is locally invasive and tends to infiltrate deep into pelvic soft tissues.[9]

Pre-operative knowledge of tumor extent is important in determining surgical approach and MRI features of AA are characteristic.[9]

Recurrence is local and reported in 36-72%.

This surgery is challenging because of the infiltration and the difficult dissection and value of extensive surgical resection to obtain clear margins has been questioned.[10] In the past, most authors advocated wide excision even if genitourinary and digestive tract resections were necessary.

Radiotherapy and chemotherapy may not have much role due to the low mitotic activity seen.

GnRH agonist and tamoxifen have been used successfully in few patients.[11]

The response can be assessed by clinical assessment, patient's symptomatology and radiographic findings.

Because of the abdominal and pelvic organ manipulation, hospital stays are generally longer, and patients are at higher risk of developing problems such as deep venous thrombosis and pulmonary embolism while they await the return of bowel function after the procedure.

There are many unanswered questions about treatment and follow-up strategies for this rare disease; because this tumor is slow growing and is often symptomatic only when the tumor is large, radiographic follow-up is best.

CONCLUSION

Although a rare diagnosis, aggressive angiomyxoma can present with unusual features. Detailed radiological examination is helpful in suspecting the problem, but histology is the gold standard for diagnosis. It should be distinguished from benign myxoid tumors with low risk of local recurrence. Therefore, recurrence of tumor may be avoided by wide local excision which is curative and prognosis of such tumor is good.

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