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Case Report

A Case Report on Angiomyxoma

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Abstract :

Aggressive Angiomyxoma (AA) is a rare variety of soft tissue tumour of pelvis and perineum occurring almost exclusively in adult females. AA is most often found in or in proximity to the lower pelvis, more specifically perineum, vulva, vagina or inguinal regions. Here we report a case of angiomyxoma. She is a 40 years old house wife, presented with a brownish, soft, multilocular, pedunculated nontender solid mass in the left side of the vaginal wall. Though initially it was provisionally diagnosed as a case of cervical polyp, ultimately histopathology proved it to be a case of Aggressive Angiomyxoma.

Key words : Aggressive, Angiomyxome.

Introduction :

Aggressive Angiomyxoma is a rare variety of non metastasining soft tissue tumour of pelvis and perineum occurring almost exclusively in adult females (female to male ratio is 6.6: 1)[1]. This was first described in 1983 by STEEPER et al[2]. No etiologic factors are known. It is most often found in women in reproductive age with a peak incidence in the fourth decade of life[2]. AA is most often found in or in proximity to the lower pelvis, more specifically perineum, vulva, vagina or inguinal regions. Tumour size is often underestimated by physical examination. Most AA are big, size may vary from 1-60 cm^3. These tumours are macroscopically lobulated and may adhere to surrounding soft tissue. Microscopically, cells with a spindled or stellate morphology are seen, embedded in a loose matrix consisting of wavy collagen and oedema (Figure 1). Cellularity is generally low to moderate. Infiltration into fat, muscle, and nerves are seen. The hallmark of AA is vessels of varying caliber haphazardly scattered throughout the tumour parenchyma, whereas mitotic figures are scarce[4].

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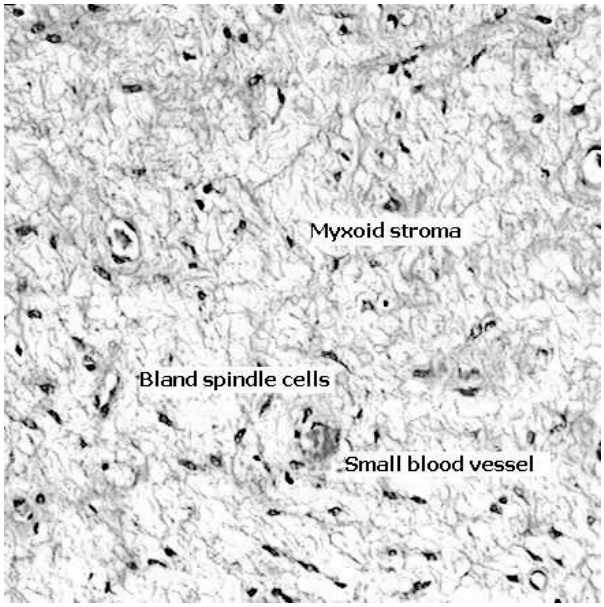

Figure 1 : Histopathology of Angiomyxoma

Immunohistochemically, most AA are positive for desmin, smooth muscle actin, muscle-specific actin, vimentin, oestrogen receptor, and progesterone receptor. Some tumours are positive for CD34[5].

Complete surgical excision is the gold standard because of its tendency to recur locally. Most surgeons aim at complete resection (wide excision with tumour free margin); incomplete or partial resection is acceptable

when high morbidity is anticipated. Because most tumours are large, grow infiltrative and blends with adjacent soft tissue, and are located in close proximity to vital organs such as bladder and rectum, wide excision is not always possible and/or may cause significant morbidity[6]. Several reported attempts using chemotherapy and radiotherapy as part of the treatment for AA have been disappointing, probably due to the low mitotic activity/growth fraction of cells. Most AA express oestrogen and progesterone receptors and are likely to have a hormone-dependent growth. Because of this, treatment with GnRH agonists and Tamoxifen has been administered to AA patients, and some case reports with dramatic responses to such GnRH agonists have been reported. Recurrences are common, though, reported to be between 9 and 72 %[7].

Case Report :

Mrs. Nurjahan, a 40 years old housewife hailing from Harishpur, Jinaidaha admitted on 18-06-12 presented with the history of something coming down through her vagina for 1 year. It was slow growing and was associated with mild dragging lower abdominal pain. The mass was gradually enlarging but she did not seek medical advice until she felt difficulty in walking and sitting. There was no difficulty in micturition and daefecation. She also had menorrhagia for 6 months. But she had normal menstrual cycle and average duration. She had no history of loss of appetite or weight loss. She received 3 units of blood transfusion. On examination patient was anxious, having average body built and nutrition. She was moderately anaemic, not icteric. There was no lymphadenopathy. Per vaginal examination revealed that Uterus was bulky, 10 weeks in size, there was multiple fibroid. Cervix could not be seen externally and found posteriorly. A brownish, soft, multilocular, pedunculated nontender solid mass was found in the left side of the vaginal wall measuring about (8X6) cm^2 with restricted mobility (Figure 2).

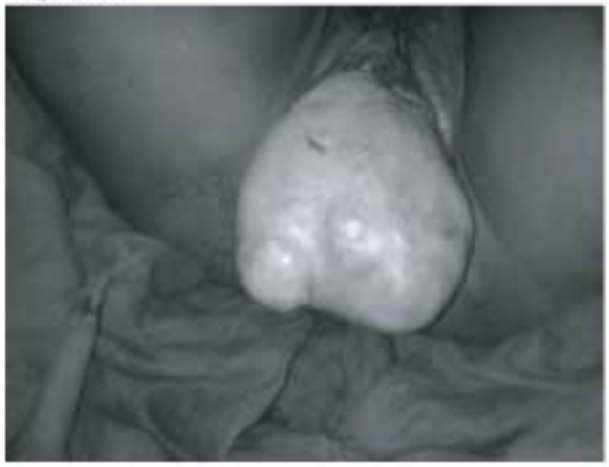

Figure 2 : Vaginal mass

Ultrasonogram revealed that uterus was bulky and multiple fibroid was seen. The cervix was broad and a hypodense mass was present displacing rather than invading the pelvic organs.

So decision of doing laparotomy was taken. Then with all aseptic precaution abdomen was opened by pfennenstiel incision. Uterus was found bulky 10 weeks size and multiple fibroids were present. Total abdominal hysterectomy was done. Both sided ovary was preserved. But no continuation of the vaginal mass was found abdominally. Then after maintaining proper haemostasis abdomen was closed in layers and then we went to vaginal route. The vaginal mass was present on anterolateral side of left vaginal wall. The mass measuring about (8x6 cm) was excised completely at vaginal route (Figure 3). A vaginal pack was kept in situ for 24 hours and the tissue was sent for histopathology.

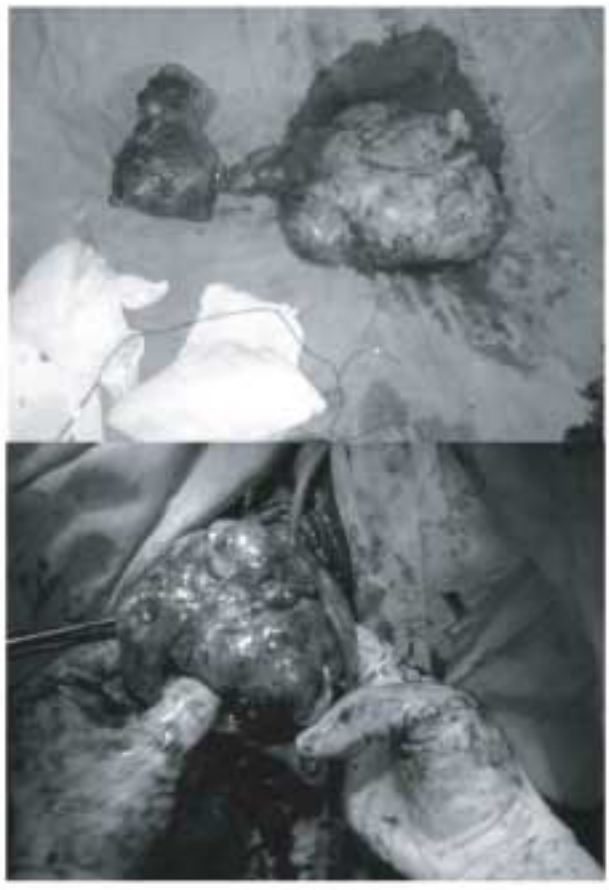

Figure 3 : Excised uterus and Vaginal mass

Histopathology revealed endometrium in proliferative phase, myometrium shows multiple benign leiomyoma, Cervix has chronic cervicitis with squamous metaplasia and vaginal mass was an aggressive angiomyxoma.

Discussion :

Angiomyxoma usually occurs in 4th decade[2]. Our patient also presented at her 40 years of age. It is more common around the perineal region. Our patient also presented as a vaginal mass. The majority of patients present with a slow-growing mass which is otherwise asymptomatic and this is frequently the only symptom/sign. Observed accompanying symptoms and signs are regional pain, a feeling of local pressure, or dyspareunia. In our case patient had only dragging pain. As the mass protrude externally through vagina, it did not produce any compression in urethra and rectum. So there was no associated bowel and bladder disturbance. There was history of dyspareunia. She had associated menorrhagia which was supposed to be due to uterine leiomyoma. Excision is the gold standard of treatment and so was done in this case. Due to financial constrain no immunohistochemistry was done and tamoxifen was not given. She was kept under follow-up to observe recurrence.

Conclusion :

Angiomyxoma is a tumor of low malignant potential. But there is frequent relapse. Even after 6 month of follow-up our patient didn't have any sign of recurrence.

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