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Rare Case of Aggressive Angiomyxoma of the Vulva: A Case Report

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Abstract

Aggressive Angiomyxoma (AA) is a rare locally aggressive soft tissue tumor with high chance of local recurrence occuring in young women of reproductive age group. These neoplasms are characterized by a mixture of spindle or stellate cells in a loosely myxoid stroma with prominent vascular component containing large, thickwalled blood vessels. A 35 year old multiparous lady presented with right vulval swelling which was rapidly progressing in size. Local examination revealed a soft fleshy mass occupying whole of the right labia majora. In view of the rapidly enlarging lesion over a short duration, we suspected a soft tissue neoplasm and preceded with wide local excision of the mass with adequate margins in all dimensions, eventhough we did not have a preoperative tissue diagnosis. To our surprise, final histopathology was Aggressive Angiomyxoma (AA). The treatment of choice for Aggressive Angiomyxoma is wide local excision with negative margins and the same was done in our case. Recurrence of the tumor may be avoided by wide local excision with adequate margins. It is possibly a hormonally responsive neoplasm. Treatment with Gonadotropin-releasing hormone is an emerging therapy. Overall prognosis of AA is good.

Keywords: Aggressive Angiomyxoma (AA), Vulva, myxoid stroma, vascular, spindle or stellate cells

1. Introduction

Aggressive Angiomyxoma (AA) is a rare locally aggressive soft tissue tumor with high chance of local recurrence. It occurs mainly in the pelvis, vulva, perineum, vagina and urinary bladder (Mandal et al., 2008). It was first described by Steeper and Rosai in 1983(Mandal & Rosai, 1983). It is known as Aggressive Angiomyxoma in view of the neoplastic nature of the blood vessels and locally infiltrative nature. Only few cases have been reported about this rare tumor in the world literature (Fetsch et al., 1996; Begin et al., 1985; White & Chan, 1994). Here we describe a rare case of rapidly growing vulval Aggressive Angiomyxoma in a young multiparous female.

2. Case Report

A 35 year old multiparous lady presented with right vulval swelling of 1 month duration. The swelling was insidious in onset and rapidly progressing in size. The patient had occasional serous and blood stained discharge from the mass. She also had difficulty in walking due to the mechanical effect of the mass. She is a known diabetic on oral hypoglycemic drugs. General and abdominal examination revealed no significant abnormality. Pelvic examination revealed an 11cm * 8 cm soft fleshy mass occupying whole of the right labia majora.Superficial ulceration was present over the summit of the mass. The mass was sessile, but mobile along with the labia majora with few dilated veins over it (Figure 1).

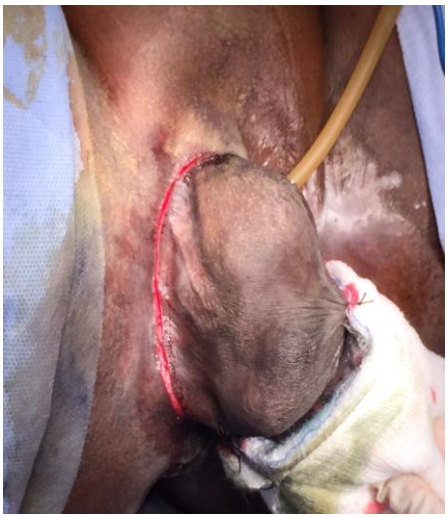

Figure 1. Diagram showing mass in the right labia majora with dilated veins over it. The summit of the mass with ulceration is covered just before resection to prevent tumor spill. The incision marked all around with adequate margins at the commencement of the resection.

Urethra was normal. Rest of the pelvic examination did not reveal any abnormality. On investigation, haematological and biochemical investigations were within normal limits. ChestXray was normal. Ultrasound whole abdomen and pelvis revealed a bulky uterus. Local imaging in the form of MRI pelvis was not done as the lesion was superficial and mobile without any deep extension. Endometrial aspiration was done in view of the bulky uterus, which showed only disordered proliferative pattern of the endometrium. Wedge Biopsy was done from the ulcerated area which revealed only vulval ulceration and granulation tissue. In view of the rapidly enlarging lesion over a short duration, we suspected a soft tissue neoplasm and proceeded with wide local excision of the mass with 1 cm margin in all dimensions, eventhough we did not have a preoperative tissue diagnosis. Urethra was preserved. Primary closure of the wound was done post resection. Macroscopically, the dimensions of the lesion were 11cm*7.5cm*5.4cm. On cut section, the mass was homogenous with a gelatinous appearance. Microscopically, spindle or stellate cells were present in a background of loose collagenous stroma (Figure 2).

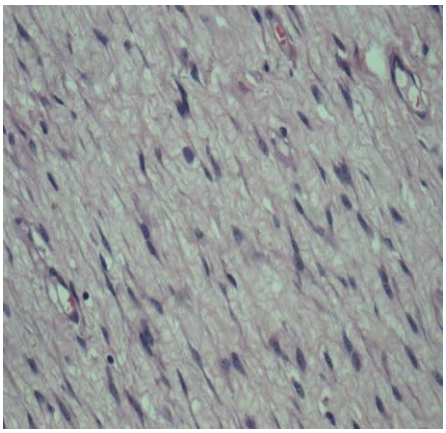

Figure 2. The microscopic appearance showing a mixture of spindle or stellate cells in a loosely myxoid collagenos stroma. The stroma is monotonous and hypocellular with no atypia of the cells and absent mitotic figures.

The stroma with collagen fibers had a prominent vascular component containing large, thickwalled blood vessels. Immunohistochemistry was not done as the microscopy was typical of aggressive angiomyxoma with prominent vascular component (fIgure 3).

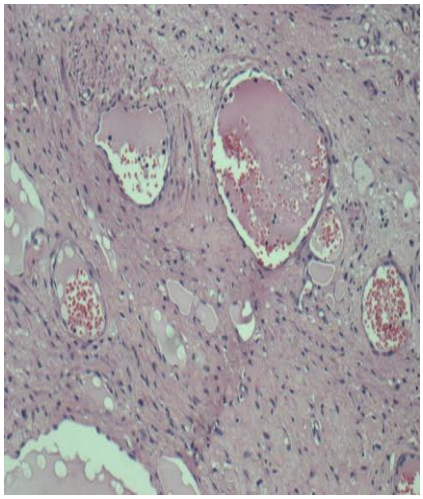

Figure 3. The myxoid stroma with collagen fibres with prominent vascular component containing large, dilated, thick walled blood vessels (Ectatic blood vessels).

Resected edges were free of tumor. The Patient is on regular followup and is disease free for the past 1 year.

3. Discussion

Aggressive Angiomyxoma (AA) is locally aggressive with a high chance of local recurrence (Mandal et al., 2008). Grossly, these tumors are soft, bulky masses with a smooth external surface. This tumor generally occurs as a large, slow growing, polypoid mass or ill defined swelling in the pelvic region with infiltrating margins. The tumor size varies between 3 and 60cm (Fetsch et al., 1996; Begin et al., 1985; White & Chan, 1994). Our case had an 11cm* 8 cm size mass in the right labial region. The histogenesis of the tumor is poorly known (Rabban et al., 2006). The microscopic appearance of these neoplasms is characterized by a mixture of spindle or stellate cells in a loosely myxoid stroma. This stroma contains collagen fibers and a prominent vascular component containing large, thickwalled blood vessels. Hemorrhage and cysts is not a feature of AA, although microcystic changes may be seen (Metin et al.). It is possibly a hormonally responsive neoplasm, inview of it's propensity to occur in females in reproductive age group (Htwe et al., 1995). The differential diagnoses include benign tumors such as myxolipoma, myxoidneurofibroma and myxoid leiomyoma and malignant tumors such as myxofibrosarcoma, myxoid variant of liposarcoma, leiomyosarcoma, malignant fibrous histiocytoma and botryoidrhabdomyosarcoma (Behranwala & Thomas, 2003). The distinctively striking vascular component in aggressive angiomyxoma helps to differentiate from these neoplasms.

Aggressive angiomyxoma is locally invasive and has a tendency to infiltrate into deep pelvic soft tissues. Hence, wide local excision with negative margins is the optimal treatment.It is associated with frequent recurrences, probably due to incomplete removal or multifocal occurrence of these neoplasms (Kaur et al., 2000). This emphasizes the need for long term followup.Meticulous monitoring with clinical examination and imaging techniques is necessary before the possibility of a recurrence can be dismissed. Recurrence of the tumor may be avoided by wide local excision. Wide local excision with adequate negative margins in all dimensions was done in our case and it has helped preventing recurrence in our patient.

Mandal et al, described a case of pedunculated Aggressive Angiomyxoma of the vulva in a young pregnant female (Mandal et al., 2008). Our patient was not pregnant and presented with a rapidly growing sessile mass in the right labia majora. Fetsch et al, reported a clinicopathological study of 29 female patients with pelvic and perineal Aggressive Angiomyxomas. They emphasized the importance of long term followup since the first evidence of recurrence may be many years after the initial resection (Fetsch et al., 1996). Our case is on regular followup for the past one year and is disease free till date.Radiation therapy was given for a case of recurrent Aggressive Angiomyoma which achieved local control by Suleiman et al (Suleiman et al., 2006). Our patient presented with a primary lesion and underwent wide local excision with adequate margins.Hence, radiation therapy was not offered. Deep aggressive angiomyxoma of pelvic soft tissue was described by Sirasagi et al (Sirasagi & Arakeri, 2014). Our case did not have any pelvic extension.

The treatment of choice for Aggressive Angiomyxoma is surgical excision. Treatment with Gonadotropin releasing hormone is an emerging therapy. Overall, the prognosis is good. Metastases are exceedingly rare (Sutton & Laudadio, 2012). Blandamura S et al., described Aggressive angiomyxoma in a young woman with multiple

recurrences and subsequent lung metastases. The patient succumbed to lung metastases. A case of Aggressive angiomyxoma with lung and mediastinal metastases has also been described in a postmenopausal woman (Blandamura et al., 2003). Our Patient has not developed any local recurrence or distant metastasis so far.

4. Conclusion

The diagnosis of Aggressive angiomyxoma should be borne in mind when dealing with a rapidly growing vulval soft tissue tumor in the reproductive age group. Aggressive wide local excision at the initial surgery prevents recurrence of these rare locally invasive soft tissue tumors as in our case. Long term followup is indicated in view of possibility of late recurrences.

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