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Angiomyxoma in Vagina: A Rare Case

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Abstract: ***Angiomyxomas (AA) are a rare type of locally infiltrative tumour with a high tendency for recurrence. We present a 35 year old woman who presented with a painless swelling in the vaginal area which was progressively increasing in size. Her gynecological examination and laboratory investigations were unremarkable. She underwent a wide local excision of the tumour. Histopathology examination of the tumour revealed an angiomyxoma. The principle treatment of angiomyxoma is the complete surgical excision with tumor-free margins. Long-term follow-up is required as this tumour has a high tendency of local recurrence. AA should be considered in the differential diagnosis of any painless swelling located in the genitofemoral region, particularly in women of reproductive age. Very few cases of angiomyxoma of the vagina have been reported so far and we describe one such case in this report.***

Keywords: Angiomyxoma, tumour

1. Introduction

Angiomyxomas are a rare type of soft tissue tumour. They are not thought of as cancer because they usually grow quite slowly and they don't usually spread to other parts of the body. These tumours develop from a type of cell called myxoid cells. They are one type of cell found in the body's connective tissue. These tissues hold organs and other body structures in place. There are two types of angiomyxoma - superficial angiomyxoma and aggressive angiomyxoma. Superficial angiomyxoma is generally seen on the skin or just below the surface of the skin. It can affect anywhere on the trunk or lower part of the body, the genitals (penis and vagina), and head and neck. Aggressive angiomyxoma tends to grow deeper into the tissue. They can also grow into the tissues around them. But they are very unlikely to spread to other parts of the body. They mostly develop on the perineum or in the pelvis.Aggressive angiomyxoma mostly affects women who are of childbearing age. Angiomyxoma in the vulva and vagina are especially a rare case. Approximately around 350 cases have been documented in the scientific literature so far[1].A 35 year-old Asian female with a palpable lesion is presented herein, which turned out as Angiomyxoma.

2. Case Presentation

A 35 year-old woman, P2L2 presented in the gynecology OPD with complaints of lump in the vagina since 6 months. It was not associated with pain. There were no complaints of abnormal discharge or dyspareunia. The patient noted a progressive increase in size of the swelling and hence decided to consult a gynecologist for the same.

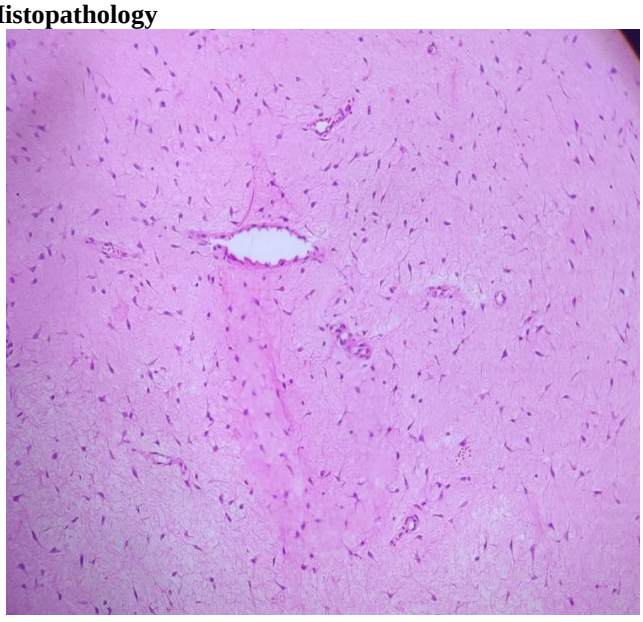
Past medical and family history was unremarkable. On general examination, the patient was moderately built and afebrile. There was no evidence of jaundice, anemia, cyanosis, lymphadenopathy, clubbing, weight loss, or any bowel and bladder function alterations. Her menstrual cycles were regular with normal flow.

During physical examination, around 3x4 cm mass was noted in the right upper fold of labia majora.On palpation, the mass was nontender, nonreducible, and cystic in consistency. The cervix and vagina was healthy and the uterus was normal in size with both fornices free. Her laboratory investigations were within normal limits and ultrasonography of the abdomen was within normal limit.

The initial differential diagnosis included the bartholins cyst or a paravaginal cyst

The patient underwent a wide local excision of the tumour which was sent for histopathology examination.

Histopathology



H and E stained section from the vulvovaginal mass shows a neoplasm composed of cells with small bland ovoid to short spindle nuclei and scanty cytoplasm in abundant myxoid stroma with delicate collagen fibre. In between these cells numerous variable sized blood vessels are seen. Mitotic activity is negligible.

3. Discussion

Angiomyxomas are recognized as superficial or aggressive. Superficial angiomyxomas occur more commonly in middle-aged patients presenting as a single nodule or a polypoidal mass in the head and neck region, trunk, and lower extremities.

Aggressive Angiomyxoma (AA) is reported almost exclusively in women of reproductive age, with occasional cases seen in perimenopausal females and children[2.] This may be attributed to the hormone-responsive nature of the tumor, as its growth is stimulated by estrogen and progesterone. It usually presents as a painless mass in the vulvoperineal region.

The tumour has a high tendency for local invasion and local recurrences (as high as 30–72%) The tendency of the tumour to metastasize is low.

The diagnosis of AA is very difficult because it is often asymptomatic until the tumor reaches large sizes (as in our case). AA commonly presents as a painless swelling located around the genitofemoral region. For this reason, it is often misdiagnosed as a vulvar abscess or Bartholin's gland cyst.

Preoperative imaging includes USG, MRI and CT scan. It should be used to see the extent of the tumor and plan surgical excision accordingly as the size of the tumor is often underestimated by clinical examination.

A study conducted by Chen Yang Zhao et al[3] on USG findings of angiomyxoma showed that AA lesions were irregular and hypoechoic masses with relatively well-defined margins. The lesions had intermediate to high echogenicity with a layered or swirled arrangement. Finger-like or tongue-like growth patterns were visible in some cases, as a result of infiltrative growth into the gaps of surrounding soft tissues. Prominent deformity of the lesions was shown on real-time ultrasonic imaging after the masses were compressed by probes, which verified the soft texture of AAs. Abundant blood flow was detected inside and around the masses on colour Doppler ultrasound.

A study was conducted by Wu H et al[4]to evaluate the value of CT and MRI in aggressive angiomyxoma of the pelvis. The authors concluded that both CT and MRI show characteristic imaging pattern and the diagnosis should be considered in any young woman with a well-defined mass arising from the pelvis or perineum. They elucidated that both CT and MRI precisely predict the extent of the tumor. However, MRI is more specific and is superior to CT when ascertaining the tumor's relation to the surrounding structures.

Grzegorz Marek Karwacki et al[5]suggested that typical MRI features of AA are swirled strands aligned with the craniocaudal due to the stretching of the fibrovascular stroma during protrusion through the pelvic diaphragm.

Wide local excision with negative margins and long-term follow-up remains the best course of management but often results in high morbidity. Adjuvant therapy in the form of gonadotropin-releasing hormone (GnRH) agonists has shown promising results to prevent recurrences.

Many treatment modalities have been tried with varying success. Therapies of AA include surgery, GnRH-a, arterial embolization, and radiation therapy. However, radiation therapy and chemical therapy are used in rare cases, because proliferation and division of tumor cells are not significant, limiting responses to radiation and chemical therapy.

Radical surgical excision with negative margins is the conventional treatment of choice. However, it is not always possible to achieve negative resection margins as the tumor is locally infiltrative, leading to high operative morbidity. Therefore, less radical surgery is recommended nowadays. Adjuvant therapy in raloxifene, tamoxifen, or GnRH agonists like leuprolide acetate and goserelin have proven beneficial where the tumor is estrogen and progesterone receptor sensitive. In a case report by Fine et al[7], recurrent AA of the vulva was treated solely by 3 months of GnRH agonist without needing any other medical therapy or surgery metastasis has been reported as an exceedingly rare event in literature. Siassi *et al.*[8] reported a death due to multiorgan metastasis invading the peritoneum, lungs, and lymph nodes

All adjuvant treatment modalities remain controversial. Chemotherapy yields no beneficial results for adjuvant therapy because of low mitotic activity of the tumor. Several beneficial results with tamoxifen or gonadotropin-releasing hormone (GnRH) agonist have been described. However, long-term use of these drugs is associated with side effects such as menopausal symptoms and bone loss. Moreover, the optimal duration of therapy is unknown. The immunohistochemical findings of the present tumor confirmed positivity for both estrogen and progesterone receptors

Our patient required frequent follow ups till the complete recovery post surgery. This highlights the importance of counselling the patients regarding high intra op and post op morbidity associated with the tumour.

4. Conclusion

Despite being a very rare diagnosis, AA should be considered in the differential diagnosis of any painless swelling located in the genital region, especially in women of reproductive age.

Preoperative modalities such as CT and/or MRI should be used not only for the diagnosis but to help us alienate the margins for better excision of the tumour.

The treatment should be complete surgical excision with tumor-free margins. The patient should be counselled regarding the need of regular and long follow up not only due to the high morbidity of the surgical intervention but also due to its high tendency of local recurrence.

Many hormonal treatments such as Injection Leuprolide and oral Tamoxifen have been tried for postoperative usage with an intention to prevent the recurrence of the tumour. But the literature for the same is very limited.

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