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ANGIOMYXOMA OF THE VAGINA MIMICKING AS EPISIOTOMY SCAR ENDOMETRIOSIS. A CASE REPORT

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ABSTRACT
Angiomyxoma is a very rare mesenchymal tumor. It is usually present in reproductive women and found in lower pelvis. Pain is the common symptom and also presentation differs according to the site of lesion. This is debatably the only case of angiomyxoma presenting clinically as an episiotomy scar endometriosis. Whenever feasible, surgical excision with adequate margin is the treatment of choice.

KEY WORDS: Angiomyxoma, Cutaneous myxoma, Endometriosis.

INTRODUCTION

Angiomyxoma is an extremely rare, slow growing, benign soft tissue tumor of mesenchymal origin.[1,2] It was first reported by Steeper et al in 1983.[3] Angiomyxoma is classified as Superficial and Deep. Deep angiomyxoma is usually characterized by aggressive local growth. It is usually present in woman of reproductive age group and is usually found on the lower pelvis commonly perineum, vulva, vagina or inguinal regions.[1,2] It can extend considerably to the deeper tissues hence cross sectional imaging is important for determining the extent of the tumor so as to optimize the surgical approach and reduce the incidence of recurrence.[2,3] Surgical excision with a clear excision of a margin is the treatment of choice for Angiomyxoma. Here we present a case of Angiomyxoma who presented with a typical history of pain during menstruation over the episiotomy site and was diagnosed as a case of Episiotomy scar endometriosis but later turn out to be Angiomyxoma on Histopathological report.

CASE

A 35 years lady, $P_1A_0L_1$, presented with a complaint of pain over the perineal region at the site of episiotomy during menstruation following the delivery. She also complains of heaviness over the same site. Her last menstrual cycle was 15 days prior to the presentation. And the menstrual cycles were regular. There is no history of use of any contraception. She had normal vaginal delivery with right sided episiotomy 2 years back in our Institute. Post-delivery she developed Episiotomy site infection which was managed with regular dressing. Per abdominal examination was normal. On local examination, there was a small, tender brownish dot of around 0.5×0.5 cm^2 over the right posterior vaginal wall in the episiotomy scar site (figure 1). There was no definite mass and no bleeding or discharge from that site. Per speculum and per vaginal examination were normal. There were no other nodularity and no bluish discoloration at any other sites of vulva and vagina. Per rectal examination was also normal. Since the patient gave typical history of pain during menses at episiotomy site, diagnosis was made of Episiotomy site scar endometriosis. USG showed normal upper and pelvic scan. And all other laboratory reports were normal. She was planned for surgical excision. Wide Local Excision was done and vagina repaired in 3 layers. The specimen was sent for Histopathological evaluation. Post operatively patient had uneventful recovery. Patient followed up with histopathological report and was suggestive of Angiomyxoma (figure 2). After the surgery, she in now asymptomatic during menstruation.

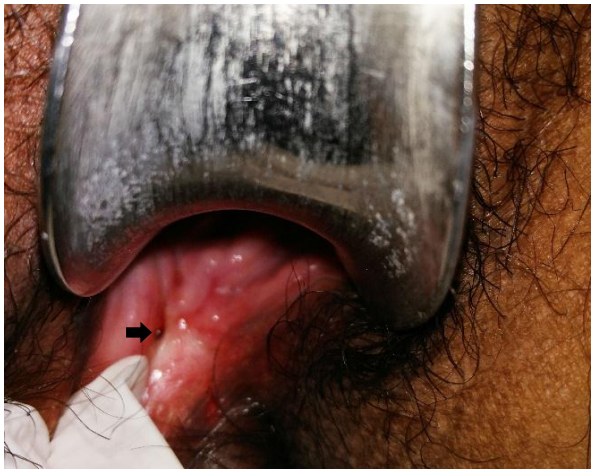

Figure 1: The image of the lesion. Arrow head pointing the lesion.

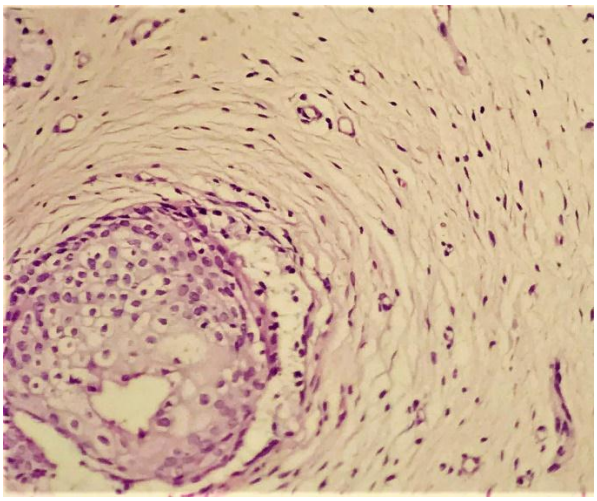

Figure 2: figure showing keratinized stratified squamous epithelium and sub epithelium hypocellular myxoic areas composed of slender spindle to stellate cells, along with numerous delicate thin walled and elongated vessels.

DISCUSSION

Angiomyxoma is an extremely rare, slow growing, benign soft tissue tumor of mesenchymal origin.[1,2] It was first reported by Steeper et al in 1983.[3] This tumor is very rare with only about 150 cases being described in the literature. Angiomyxoma is classified into Superficial and Deep. Superficial angiomyxoma, also known as cutaneous myxoma is predominantly found in male of middle age anywhere in the superficial tissues, but mostly involves trunk, lower extremities, head and neck. It usually appear as slowly growing polypoid cutaneous lesions and are easily confused with a cyst, skin tag, or neurofibroma. Our case was also of superficial type and it presented as episiotomy scar endometriosis. Histologically, there is a Spindle and stellate-shaped cells embedded in a myxoid matrix along with the presence of variable-sized thin-walled capillaries and thick walled vascular channels.[1] Deep angiomyxoma is usually characterized by aggressive local growth with high rate of recurrence rate.[1] It is predominantly found in women of reproductive age group, with a female-to-male ratio of six to one.[2,4] Aggressive Angiomyxoma in male had been reported in the perineum, penis and scrotum. Tumors commonly involve the lower pelvis commonly perineum, vulva, vagina or inguinal regions.[1,2,5] It has also been reported involving the upper abdomen and in transplant kidneys, urinary bladder, parotid gland, oral cavity and larynx.[2,4,6-8]

The clinical presentation vary according to the site of involvement. They can be asymptomatic or present with pain, pressure symptoms, urinary obstructive symptoms, bowel obstruction and dyspareunia. The presentations usually mimic that of the more common disease like Bartholin's gland cyst, abscess, lipoma, simple labial cyst, or other pelvic soft tissue tumors.[3,4] This case may be the only one to mimic as the episiotomy scar endometriosis. On examination, these tumors usually appears fleshy, lobulated, soft to rubbery or as solid mass that is poorly encapsulated with adherence to surrounding tissues.[9] They often display areas of hemorrhage, necrosis and fibrosis. In some cases, just the small portion of the tumor may be visible and palpated from the above and the major portion extend considerably into deeper tissues. So, the extension of tumor deep into the tissue should be appreciated with different imaging techniques.[2] Ultrasonography usually demonstrates a cystic or hypoechoic lesion. However, MRI is currently the best and mainstay investigation. MRI appearance of the lesion is hypointense on T1 weighted, hyperintense on T2 weighted and a characteristic 'whorled' appearance post Gadolinium contrast.[2] The Cross-sectional imaging is important for determining the extent of the tumor. This will help to optimize the surgical approach finally aids in reducing the incidence of recurrence. The treatment of choice is surgical excision with a clear margin; however, it has a high rate of recurrence.[3,9] Up to 72% of cases recur locally within two to four years after surgical excision.[3] Thus, complete resection with microscopic negative margin should be the goal of surgery. Sometimes, in case of large aggressive Angiomyxoma infiltrating the adjacent organs, extensive en-block resection may be required to decrease the recurrence. When the complete resection is not feasible, hormone therapy with gonadotropin releasing hormone agonist such as Leuprolide acetate and Goserelin acetate or Tamoxifen, Raloxifene can be used. These drugs have advantage in downsizing the primary tumor as well as those with recurrence.[1,4] Radiation therapy and chemotherapy have no practical implications due to low mitotic activity. Long-term follow-up includes MRI for detecting recurrences as the most effective imaging modality.[1]

CONCLUSION

Angiomyxoma is a very rare mesenchymal tumor. High index of suspicion is required for diagnosed and sometimes it is only diagnosed after histopathological examination. Whenever feasible, surgical excision with adequate margin is the treatment of choice.

Conflict of interest: none.

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