

Aggressive angiomyxoma of the uterine corpus: A rare presentation

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ABSTRACT

Aggressive angiomyxoma (AA) is a rare benign soft tissue tumor usually affecting the female pelvis and perineum. A 16-year-old girl presented on June 23, 2016, with vaginal protrusion of 1-year duration. Examination revealed protrusion through the introitus; it was hyperemic with necrosis. Investigation revealed urinalysis that revealed blood (+), leucocytes (+), and others normal; Swab (taken from purulent discharge on the protruded mass) microscopy revealed numerous pus cells; and culture yielded no growth. Abdominopelvic ultrasound (USS) showed left-sided pelvic soft tissue mass with no features of metastasis. Other investigations were normal. She had polypectomy with subtotal abdominal hysterectomy. Histology revealed benign AA. A rare tumor of the genital tract had been presented with a diagnostic dilemma. However, confirmation of diagnosis was easy with histology.

Key words: Angiomyxoma; uterine corpus; vaginal protrusion.

Introduction

Aggressive angiomyxoma (AA) is a rare benign soft tissue tumor affecting the female pelvis and perineum,^[1] and was first described by Steeper and Rosai in 1983. The tumor is benign, nonmetastatic but vascular, infiltrative, and high local recurrence, hence the name “aggressive.” Predominantly, the tumor occurs in women of reproductive age, but may be seen in adolescents and the elderly.^[2] Less commonly, it may be seen in males involving the inguinoscrotal area and perineum with a female: male ratio of 6:1.^[3] The tumor has estrogen and progesterone receptors and may respond to hormone manipulation.^[4]

Case Report

This is a case report of a 16-year-old secondary school student, who was a single nullipara. Her last menstrual period

was on June 18, 2016. She presented to the gynecological outpatient department as a referred case from a peripheral center with vaginal protrusion at menarche a year prior to presentation.

It was initially small but increased to about that of an egg. There was fullness within the vaginal cavity initially but later protruded outside the introitus. There was no protrusion from any other part of the body. It was associated with ulceration, contact bleeding, foul smelling discharge, and fever. There was lower abdominal pain and lower back pain. However, there was no inter-menstrual bleeding or heavy menstrual flow. There was anorexia and weight loss, but there was no

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urinary symptoms, hematochezia or constipation, yellowness of the eyes, cough, or difficulty in breathing. The history of in utero exposure to Diethylstilbestrol could not be ascertained, but there was no family history of similar problem and she was not sexually active. She had polypectomy at a peripheral hospital. The mass recurred to a size larger than it was before and a repeat polypectomy was performed about 9 months after the first surgery at a different hospital. However, histology was not performed. Two weeks after the second surgery, the protrusion was noticed to reoccur again, and she was then referred to our facility.

Examination findings revealed an ill-looking young woman. She was afebrile, not pale, not jaundiced, and cyanosed. She had no peripheral lymphadenopathy and no pedal oedema. The respiratory, cardiovascular, and abdominal examinations were unremarkable. There was an obvious mass protruding through the vaginal introitus; it was hyperemic with some areas of necrosis and foul smelling discharge. It measured about 16 × 15 × 10 centimeters. Further examination was impossible due to tenderness and contact bleeding. An assessment of a suspected case of sarcoma botryoides was made. This is as shown in Figure 1.

The patient and parent were counseled on the condition, management, and its implication. She was admitted, and investigations results were from urinalysis: the urine was clear amber colored; blood (+), leucocytes (+), protein, glucose, nitrite, ketone bodies, urobilinogen, and bilirubin were negative. Full blood count and differentials revealed hematocrit of 35%, platelet count of $235 \times 10^9/L$, and white blood cell count of $6.9 \times 10^9/L$. The differentials were neutrophils of 49%, lymphocyte of 26.1%, eosinophils of 1.2%, basophils of 0.1%, and monocytes of 7.5%. The electrolyte, urea, and creatinine and liver function test were also normal. Retroviral screening was nonreactive.

Swab (taken from purulent discharge on the protruded mass) for microscopy revealed numerous pus cells; however, culture yielded no growth. Chest X-ray was essentially normal and abdominal-pelvic USS showed the liver, spleen, kidneys, and urinary bladder. There was a left-sided pelvic soft tissue mass with no features of metastasis; the uterus and ovaries could not be clearly demonstrated possibly due to distorted anatomy. There were no abdominal or pelvic lymph node enlargement.

An abdominal-pelvic computer tomography scan was requested but was not done due to financial constraint.

She was commenced on antibiotics (intravenous ciprofloxacin and metronidazole) for 72 hours and were converted to oral. The vulva was cleaned with normal saline twice daily.

They were re-counseled on the findings and the need for surgery and its extent which could compromise fertility and menstruation. Consent was obtained and she was then booked for surgery after her general condition had improved.

She had frozen section of the vaginal mass that revealed a polyploidy mass with benign features. She had polypectomy with subtotal abdominal hysterectomy on July 05, 2016.

Intra-operative findings revealed a uterine mass that destroyed the lower uterine segment and detached the cervix from the uterus. There was a small portion of the uterine fundus that was held by the fallopian tubes and round ligament. The mass pulled the uterus through the detached cervical os. The mass was firm and measured about 16 × 16 × 12 centimeters. It had no stalk and bled to contact. The fallopian tubes and ovaries were normal. The estimated blood loss was about 400 milliliters. This is as shown in Figure 2.



Figure 1: Vaginal protrusion



Figure 2: Extent of the vaginal protrusion

Through an abdominal incision, the fallopian tubes and the round ligament were doubly clamped, cut, and ligated, and the remnant of the uterus and attached huge mass were delivered vaginally. The proximal margin of the cervix was excised to increase tumor free margin while the ovaries were preserved.

The post-operative period was uneventful. She was maintained on intravenous fluid, antibiotics, and analgesics for 48 hours. She was changed to oral medication for 5 days and her post-operative packed cell volume on the second day was 32%. They were counseled on the extent of the surgery and stitches were removed on the seventh post-operative day. She was discharged home thereafter in satisfactory condition.

She reported to the gynecological clinic after 2 weeks with a histology report that revealed a mesenchymal neoplasm that is poorly circumscribed, composed of spindle shaped cells widely sparse from each other in a myxoid stroma, within which are numerous clusters of small to moderately sized vascular channels suggestive of a benign AA.

They were reassured and counseled on the findings and prognosis, in addition to fertility option of *in vitro* fertilization and surrogacy. The possibility of adoption and fostering was also discussed when ready for childbearing. She has been on follow-up at the gynecological clinic with no features of recurrence. She will be followed up for life due to high chance of recurrence.

Discussion

The pathogenesis of AA is not clearly known; however, genetic alterations on the 12q chromosome region have been implicated with aberrant expression of high mobility group A (HMGA) protein associated with DNA transcription.^[5] It may arise from special mesenchymal cells of the pelvis and perineum or from perivascular progenitor cells with various degrees of myofibroblastic features.^[6] It is present in the fourth decade in women of reproductive age,^[2] but may be seen in adolescents, as in the case presented, the patient was 16 years old. It is usually misdiagnosed because it is rare. It was also seen as a uterine polyp in a patient from Czech Republic.^[7] The clinical presentation is similar to many other conditions such as fibroid polyp, pelvic organ prolapse, neurofibromatosis, and genital cancers. This could explain why the diagnosis was repeatedly missed in this patient.

Diagnosis is mainly histological. Grossly, it is unencapsulated and may blend imperceptibly with surrounding soft tissue.^[8] It has variable sizes with rubbery consistency and glistening, gelatinous cut surface. The tumor is sparsely cellular

composed of pale eosinophilic stroma embedded with numerous haphazardly arranged blood vessels in a myxoid background.^[9] Soft tissue infiltration is common. The tumor is diffusely immunopositive for vimentin and desmin.^[8] They are also estrogen and progesterone receptor positive. The diagnosis in this patient was confirmed by the histological findings of the mesenchymal neoplasm that was poorly circumscribed, composed of spindle shaped cells widely sparse from each other in a myxoid stroma, within which are numerous clusters of small to moderately sized vascular channels.

Other supportive diagnostic methods include USS, magnetic resonant imaging (MRI), and computed tomography (CT) scanning. On USS, the mass usually appear hypoechoic while MRI and CT delineate the extent of the tumor. On CT, the tumor has attenuation less than that of muscle or it may be mainly cystic with solid components.^[9] This tumor also exhibits avid and heterogeneous enhancement after administration of contrast. In the case presented, USS showed left-sided pelvic soft tissue mass; however, there were no features suggestive of metastasis. CT scan or MRI could not be done because of financial constraint of the patient.

The primary treatment of AA is a wide local excision, although achieving negative resection margin is difficult due to the absence of capsule and its infiltrative nature.^[10] Smaller and superficial tumors of the vulva and vagina may be removed with less difficulty but larger deep-seated tumors of the pelvis may necessitate extensive procedure with partial or complete resection of some pelvic organs.^[10] In this patient, a greater part of the uterus had been destroyed with detachment of the cervix from the uterine corpus. Therefore, polypectomy with subtotal hysterectomy was performed with satisfactory outcome. The tumor has potential for local aggression and recurrence after excision. This may explain why she had two previous excisions in less than a year before being referred to us.

Other treatment modalities include radiotherapy and chemotherapy with poor results. Radiation therapy with tumor embolization can be used to decrease recurrence where tumor is large.^[10] In this index, patient adjuvant therapy was not considered because there was significant free tumor margin on histology.

Conclusion

AA is a rare tumor of which primary uterine corpus type is even rarer and often misdiagnosed. It is mainly seen in women of reproductive age especially in the fourth decade but may also be seen in younger age group. Wide local excision is the treatment of choice in superficial mass; deeper

tumors may require partial or complete resection of pelvic organ. There is need for high index of suspicion when dealing with genital swellings.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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