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Case Report

# Aggressive angiomyxoma of the vagina: A case report of a rare lateral vaginal wall soft tissue mass

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## Abstract

Aggressive angiomyxoma of the vagina is a rare disease. We present a 27-year-old woman, para 2002 with a vaginal soft tissue mass size 10 x 5 x 5 cm that protruded from the right lateral wall and extended to the introitus. This protruding mass was connected to a large pelvic mass. The vaginal soft tissue mass biopsy and histology showed aggressive angiomyxoma. After biopsy, the patient was operated to debulk the pelvic mass and reconstruct the vaginal wall. The final pathology was unchanged. Her second child was delivered by cesarean delivery in about 1 year later due to her previous cesarean section. She was still free of disease for 4 years after operation.

## Key Words:

Aggressive angiomyxoma, vagina, protrusion, mass

## Introduction

Aggressive angiomyxoma is a very rare soft tissue slow growing tumor that mainly affects the pelvis and perineal area. Its behavior is locally invasive and usually has local recurrence [1]. Aggressive angiomyxoma was first described in 1983 by Steeper and Rosai with over 100 cases reported in the literature since then [2,3]. The frequent problem of this rare disease has been a misdiagnosis as Bartholin's gland cyst or abscess, simple labial cyst if it originated at the vulva region or vaginal polyp. Gartner's duct cyst or peduncu-

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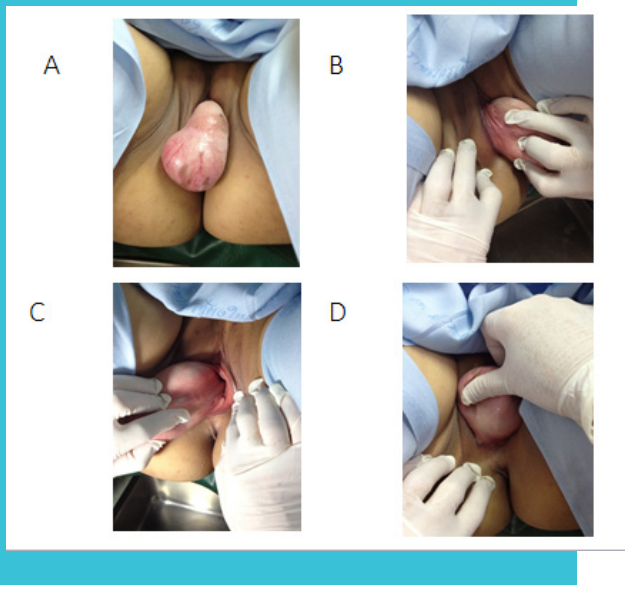
lated soft tissue tumors was the misdiagnosis if the location occurred at the vaginal wall [4]. The diagnosis is usually confirmed from the tissue pathology after excision of the tumor. We presented one patient with this rare disease for the aim to share the clinical experience from our institute.

## Case Presentation

A 27-year-old woman, presented with right lateral soft tissue vaginal wall mass size 10 x 5 x 5 cm. The mass was slow growing and protruded beyond her vulva for one year without any symptoms (Figure 1). She was referred to our institute on September 20, 2012. Her parity was 1-0-0-1 with a cesarean delivery six years prior. The patient also underwent a previous pelvic surgery to remove the pelvic mass two years after delivery at her provincial hospital with unknown definite histology. The physical and pelvic examination was unremarkable except for the right lateral vaginal wall mass. The whole abdom-

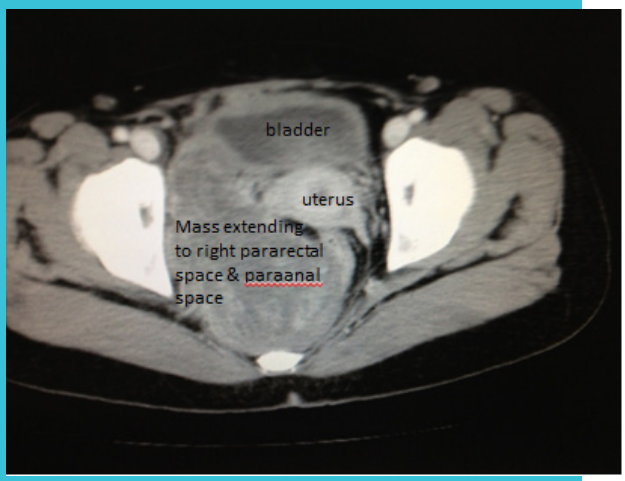
inal computerized tomography scan (CT-scan) showed a large poorly-defined, lobulated inhomogeneous enhancing mass located in the presacral region, extending into right pararectal and paraanal space and downwardly prolapsed through the external genitalia. It also involved a left laterally displaced rectum and rectosigmoid colon (Figure 2).

**Figure 1.**




*Vaginal soft tissue mass protruding from the right lateral vaginal wall*

**Figure 2.**



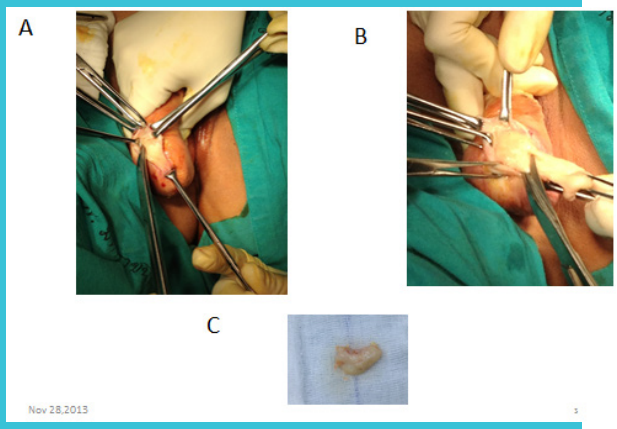

*CT-scan: large poorly-defined, lobulated inhomogeneous enhancing mass located in the presacral region, extending into right pararectal and paraanal space and downwardly prolapsed through the external genitalia. It also involved and displaced rectum and left lateral rectosigmoid colon.*

The biopsy of the vaginal mass was done on October 30, 2012. After excising the vaginal mucosa, the rubbery mass was found and was biopsied (Figure 3). The pathology was aggressive angiomyxoma. Because the CT-scan finding showed the origin of the vaginal mass coming from a huge retroperitoneal mass, we decided to remove that pelvic mass by an exploratory laparotomy in December, 2013. The operative finding identified the retroperitoneal mass occupied in the right cul-de sac that protruded to the vaginal region (Figure 4). The mass was rubbery and did not invade the adjacent organs. After removal of the pelvic mass by gentle pulling, the vaginal mass collapsed. Thus, the vaginal reconstruction was performed by excising the stretched vagina and suturing the remaining vaginal rim together (Figure 5). The greatest diameter of the gross specimen removed was 38 cm (Figure 6). The patient recovered completely. The final pathological diagnosis of the pelvic tumor was aggressive angiomyxoma, similar to the specimen from vaginal wall. Histologically, the tumor was hypocellular and was composed of abundant loose myxoid or edematous stroma which contained delicate collagen fibrils and many vascular channels of varying sizes. The stromal cells show diffuse positive immunoreaction for desmin (Figure 7) and negativity for smooth muscle actin. The stains for hormonal receptors showed diffuse positivity in stromal cells for estrogen receptor (70% of cells) and progesterone receptor (80% of cells). Unfortunately, the lower surgical margin was positive for neoplastic tissue. The patient was routinely followed-up and delivered her second healthy child by cesarean section in January, 2014. Her last follow up in January, 2016 she had no evidence of disease recurrence.

## Discussion

Aggressive angiomyxoma is a rare tumor that presents as a soft tissue mass at the pelvis, cervix, vagina and vulva [4]. It seldom occurs in males with the ratio of female to male of 6:1 [5]. The location in males also occurred at the perineum, scrotum, groin and spermatic cord [5]. The peak incidence was 35-40 years old, however, our presented case was only 27 years old. The first case of vaginal angiomyxoma was published in 2000 with brief details was a 38-year-old woman presenting with a left vagina cystic mass 10 x 7 cm covered by mucosa between the left labia minora and introitus. The authors excised the vaginal mass first and found that the mass extended towards the pararectal regions, so they subsequently explored the abdomen to totally remove the mass [6].

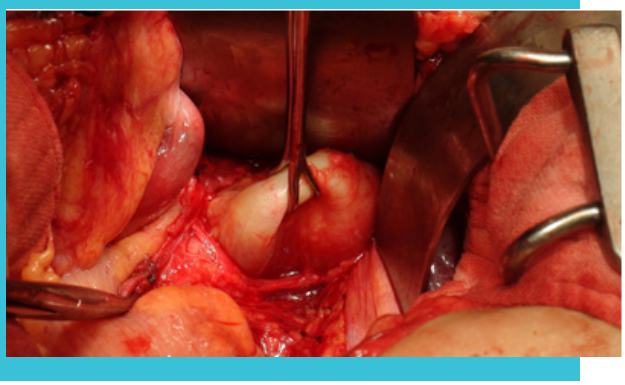
**Figure 3.**




*Vaginal mass was biopsied and showed a rubbery mass*

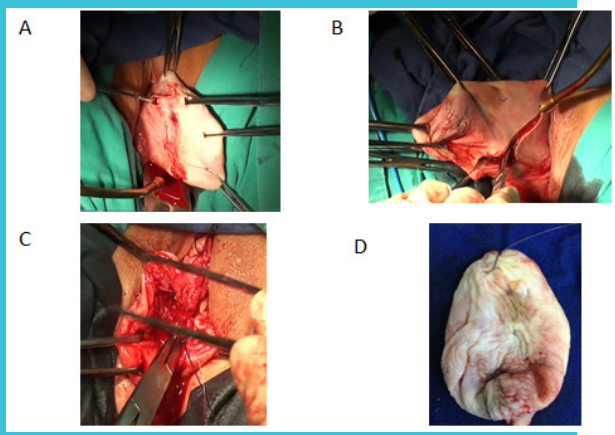
Aggressive angiomyxoma requires preoperative imaging including CT-scan or Magnetic Resonance Imaging (MRI) to identify the extent of disease that is usually underestimated by the physical examination [5]. The principle management of this tumor was complete tumor excision. The gross tumor is characterized by a soft edematous mass with gelatin-like consistency. Because there was no encapsulation and infiltrative property, the complete wide local resection was very difficult especially at the initial surgical time [4]. Histologically, aggressive angiomyxoma is a hypocellular tumor composed of small stellate-shaped to fusiform cells

**Figure 4.**



*The operative finding identified the retroperitoneal mass occupied the right cul-de-sac*

**Figure 5.**




*Reconstruction of the vagina. A&B: the stretched vaginal was excision. C) the remain vaginal rim was sutured together. D) The excised vaginal wall*

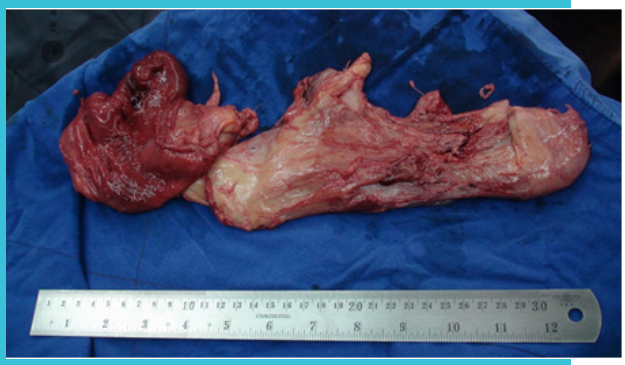
with the cytoplasmic process. These cells are scattered in a loose myxoid matrix with delicate wavy collagen fibrils. The vascular component varied from tiny capillary-like vascular channels to thick-walled muscular vessels. Nuclear atypia is usually absent or minimal [5]. Typically, the tumor

cells show diffuse immunoreaction for vimentin, desmin, progesterone receptor, and progesterone receptor, although these are not specific immunomarkers for aggressive angiomyxoma [9]. An important histologic differential diagnosis in the present case is angiomyofibroblastoma, which occurs in a similar age group and shares a similar immunohistochemical profile with aggressive angiomyxoma. However, angiomyofibroblastoma is typically a circumscribed and more superficially located tumor. Histologically, it is composed of hypocellular and hypercellular zones of a more heterogenous stromal cell population which also includes epithelioid or plasmacytoid cells and multinucleated cells [9]. Myxoid liposarcoma of the retroperitoneum may also be included in the differential diagnosis. The absence of arborizing vascular network and negative immunoreaction for S-100 protein support an exclusion of liposarcoma [9]. The main problem of aggressive angiomxoma is the high recurrence rate. Our case underwent a previous operation to remove a pelvic mass four years before developing the vaginal mass. Unfortunately, the histology of that pelvic mass was unknown. However, it was possibly an aggressive angiomyxoma. Recurrence times of 1-14 years have been reported after the initial excision [4]. In the recurrent setting, especially in patients with strongly positive estrogen and progesterone receptor, there was a report of successful treatment with gonadotrophin releasing hormone agonist [10]. However, our case was still free of disease after nearly four years despite a positive lower surgical margin showing a good prognosis of this disease. In a recent review of 111 cases of aggressive angiomyxoma, the 10-year disease-free survival rate was 50% in the patients with positive resection margin, which is only slightly higher than those with negative margin (40%). The finding suggests that incomplete resection is acceptable when the morbidity risk for radical resection is high or when fertility preservation is desired. [10 ]

**Acknowledgement**
None

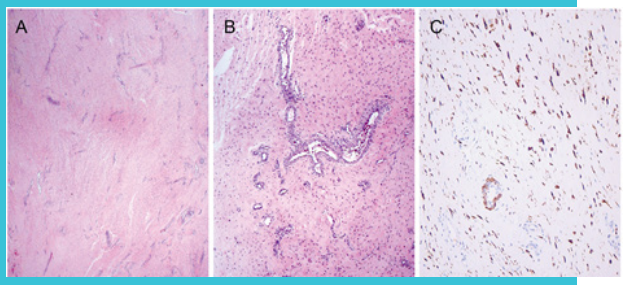
**Declaration of Interest**
None

**Figure 6.**



*Gross specimen removed from the pelvis*

**Figure 7.**




*Histology of aggressive angiomyxoma. A): Hypocellular tumor composed of myxoid stroma containing collagen fibrils and vascular channels. B) Thin- and thick-walled vessels in the stroma with scattered spindled to stellate-shaped cells. C) The stromal cells show diffuse positive immunoreaction for desmin.*

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