Case report

Deep (aggressive) angiomyxoma of the vagina misdiagnosed as Gartner cyst: A case report

Suskan Djusad a, Yulia Margaretta Sari a,*, Hartono Tjahjadi b

a Urogynecology Division, Department of Obstetrics and Gynecology, Faculty of Medicine, University of Indonesia, Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia
b Anatomic Pathology Department, Faculty of Medicine, University of Indonesia, Dr. Cipto Mangunkusumo Hospital, Jakarta, Indonesia

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ABSTRACT

Introduction and importance: Aggressive angiomyxoma is a rare soft tissue tumor. Aggressive angiomyxoma is a slow-growing vulvovaginal mesenchymal neoplasm with a marked tendency for local recurrence, but with a low tendency to metastasize. As it has a predilection for the pelvic and perineal regions, Aggressive angiomyxoma is often misdiagnosed.

This case report documented rare case of misdiagnosed Aggressive Angiomyxoma as Gartner duct cyst. Presentation of case: This article report a case of 31 year old women who complained mass came out from vagina without any urinary symptom and trauma. Physical examination and ultrasound finding suggested that the mass was Gartner Duct cyst. Management in this case was excision of the vaginal cyst. Histopathology examination revealed Deep (aggressive) angiomyxoma.

Discussion: The rarity of Deep (Aggressive) Angiomyxoma makes the preoperative diagnosis fairly difficult. Aggressive angiomyxoma is often misdiagnosed as it may have similar clinical presentation to common lesions such as Bartholin cyst or prolapse vaginal wall, Gartner cyst or levator hernia. Aggressive Angiomyxoma should be considered as differential diagnosis in patient with vaginal cyst.

Conclusion: Aggressive Angiomyxoma is rare condition. Preoperative diagnosis and management are challenging. Long term follow up and evaluation should be done due to high rate of recurrence.

1. Introduction and importance

Aggressive angiomyxoma is a rare, benign, locally infiltrative mesenchymal tumor found usually in women during reproductive age, which was first described by Steeper and Rosai in 1983. It is a slow-growing, low-grade neoplasm involving the pelvis and perineum with a high risk for local recurrence, which often occurs after many years [1]. The term aggressive was introduced to emphasize the locally aggressive behaviour and the high potential for local recurrence, it does not reflect a high probability for metastasis. The name angiomyxoma was chosen because of the similarity to myxoma and the notable vascular component [2].

Aggressive angiomyxoma is most often found in or in proximity to the lower pelvis, more specifically perineum, vulva, vagina or inguinal regions [3]. The majority of patients present with a slow-growing mass which is otherwise asymptomatic and this is frequently the only symptom/sign [4]. Clinically, Aggressive angiomyxoma may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia or sarcoma [4,5].

Recurrences are common, though, reported to be between 9 and 72%. These numbers are uncertain because late recurrences may develop several years after the primary tumor was found, and long-term follow-up of the patients is therefore very important [2].

We present a rare case of Aggressive angiomyxoma in 31 years old female which was misdiagnosed as Gartner duct cyst. Patient presented with vaginal cyst and underwent excision of the cyst. Final histopathology result was Deep (Aggressive) Angiomyxoma, a rare soft tissue tumor in female pelvis. Misdiagnosis in this case should be reported so that it becomes a consideration in diagnosing benign vaginal tumor.

This case report has been reported in line with the SCARE 2020 criteria [6].

2. Patient information

A 31 years old, Javanese woman was referred from Obgyn to Urogynecology Outpatient Clinic of Doctor Cipto Mangunkusumo Central
General Hospital with chief complain a lump came out from vagina since a year ago. The lump feels bigger two weeks before admission. The mass often came out during activity. Patient had no complaint of pain and vaginal discharge. There were no history of trauma, any associated itchiness, and urinary symptoms. There are no lumps in other part of her body. Patient is unmarried and has never had sexual intercourse. Patient had never been treated before for this disease. There is no family history of the same disease. Patient is private employee. She has no history of smoking, alcohol and drug consumption.

3. Clinical finding

Physical examination revealed cystic mass size $5 \times 2 \times 4$ cm, smooth surface, fluctuant, freely mobile, nontender, originated from left paraurethral region. Sondage was inserted through urethra meatus, there was no connection between cystic mass and urethra (Fig 1). Rectal examination revealed normal shape of uterus and there was no mass palpated in adnexa. Blood investigation and pelvic ultrasound were normal.

4. Diagnostic assessment

Translabial ultrasound showed cystic mass size $5.4 \times 2.33 \times 2.53$ cm, homogenous, hypoechoic with echo interna, clear border, monocellular. Mass is not connected with urethra (Fig. 2).

5. Therapeutic intervention

The operation was performed by experienced Urogynecologist in Dr. Cipto Mangunkusumo Central General Hospital. Excision of the cyst was performed to restore normal anatomy (Fig. 3).

6. Follow up and outcome

Histopathology result was in accordance with Aggressive angio- myxoma (Fig. 4).

Postoperative period was uneventful and patient was discharge in satisfactory condition. Patient was counselled on the possibility of recurrence. She is being followed up and to date (a month post-excision), there has been no sign of recurrence and no other disability (Fig. 5). Since there is possibility of recurrence in the future the patient was advised to control annually undergo routine gynecology examination and MRI if necessary to assess recurrence.

7. Discussion

Aggressive angiomyxoma is a rare soft tissue tumor, first named and described by Steeper and Rosai in their quintessential case series in 1983 (Steeper and Rosai, 1983) [1]. It typically occurs in the pelvic and perineal region in females, with six times the incidence in females compared to males. The World Health Organization defines Aggressive angiomyxoma as a "tumor of uncertain differentiation". It usually occurs in reproductive age with peak incidence between the third and fourth decades of life, with higher (95%) preponderance in women [5]. Aggressive angiomyxoma is a rare, locally aggressive myxoid mesenchymal tumor, arising in the deep soft tissues of the vulva, vagina, perineum and pelvis of young adult women [7]. Prevalence in the population is unknown due to its rarity, making management and counseling difficult. Most reports of this disease in the form of case reports. However, it seems that Aggressive angiomyxoma has a tendency for recurrence and can be a locally aggressive and invasive tumor [5]. The term “aggressive” emphasises neoplastic nature of blood vessels, its locally infiltrative nature and high risk of local recurrences and does not indicate its malignant nature [7].

Aggressive angiomyxoma is a neoplasm. It is defined as benign but has infiltrative potential into skeletal muscle and fat. The disease is therefore considered locally aggressive although it does not infiltrate surrounding viscus. The majority of Aggressive angiomyxoma are large, often more than 10 cm in largest diameter. These tumors are macroscopically lobulated and may adhere to surrounding soft tissue [2,8]. Microscopically, cells with a spindled or stellate morphology are seen, embedded in a loose matrix consisting of wavy collagen and oedema. Cellularity is generally low to moderate. Infiltration into fat, muscle, and nerves is seen. The hallmark of Aggressive angiomyxoma is vessels of varying calibre haphazardly scattered throughout the tumor parenchyma, whereas mitotic figures are scarce. Immunohistochemically, most Aggressive Angiomyxoma are positive for desmin, smooth muscle

Fig. 1. Preoperative examination.

Fig. 2. Ultrasonography examination.
actin, muscle-specific actin, vimentin, oestrogen receptor, and progesterone receptor. Some tumours are positive for CD34, whereas S100 is invariably negative. Most Aggressive angiomyxoma express oestrogen and progesterone receptors and are likely to have a hormone-dependent growth. Because of this, treatment with GnRH agonists has been administered to Aggressive angiomyxoma patients, and some case reports with dramatic responses to such GnRH agonists have been reported [2].

This case was initially diagnosed as Gartner duct cyst. Diagnostic method that performed in this patient were physical examination and ultrasonography. Multiple imaging modalities have been used to evaluate preoperatively, including CT (as in the described case), magnetic resonance imaging (MRI), and ultrasonography. Appearance on imaging may be variable, but has been noted to have a distinct “swirling” pattern on MRI, related to its myxoid composition. On CT, it may be hypodense to muscle, or have both cystic and solid components. It has been suggested that MRI may be preferred, due to characteristic swirling pattern observed, better depiction of tissue, and avoidance of radiation exposure [5]. But due to consideration of high cost of its examination, ultrasound imaging could be use as diagnostic tool.

The rarity of this condition makes the preoperative diagnosis fairly difficult [9]. Aggressive angiomyxoma is often misdiagnosed as it may have similar clinical presentation to common lesions such as Bartholin cyst or prolapse vaginal wall, Gartner cyst or levator hernia. Misdiagnosis rate may be as high as 70 to 100% [7]. Although Aggressive Angiomyxoma are not common, they should be thought as differential diagnoses of vulval tumours that appear to be benign on clinical examination [7,8].

Management in this case was excision of the cyst. Longitudinal incision was made in vaginal mucosa, continue with sharp dissection to separate the wall of the cyst until reaching the base of the cyst. Finally the base of the cyst was cut and vaginal mucosa was stitched. Surgical excision with wide margins remain treatment of choice [9,10]. It is difficult to remove it completely by surgery because it infiltrate the surrounding tissue. Because of this reason, on ischiorectal and retroperitoneal spaces tumor recurrence occurs commonly [9]. Recurrence may occur either due to intentional or unintentional subtotal removal, as its borders are difficult to identify grossly, or despite adequate resection. Surgeons may consider medical management of residual disease with hormone modulating therapy or alternatively simply with close observation, which has been shown to be a viable option with long follow up and no subsequent recurrence of tumor. Recurrence rates range from 25% to 47%, with 85% recurring within 5 years of initial surgery [5]. Additionally, there is not a well-established role for chemotherapy or
radiation therapy in the treatment of Aggressive Angiomyxoma, though there are a few cases described where radiation was utilized pre-or-post-operatively to decrease tumor size or minimize risk of recurrence. Medical treatment using hormonal agents, in particular gonadotropin releasing hormone (GNRH) agonists, sometimes in combination with selective oestrogen receptor modulators (SERM), has been described in treatment of recurrent tumours (Palomba et al., 2011). Use of hormone-modulating therapies has shown success in both neoadjuvant and adjuvant modality as it has been used to preoperatively shrink tumours to minimize surgical morbidity, and post-operatively to treat recurrent disease. Both post-operative radiation and GNRH-agonists have demonstrated complete resolution of residual or recurrent tumor growth, and will continue to be valuable treatment tools to be explored in recurrent and possibly metastatic disease [5]. Long term follow up by clinical examination and MRI essential for early detection of local recurrence [7].

8. Conclusion

This case report misdiagnosed Aggressive Angiomyxoma as Gartner duct cyst. The rarity of this condition makes the preoperative diagnosis fairly difficult. Although it is a rare case, Aggressive Angiomyxoma should be considered as differential diagnosis of vulvar tumor. Long term follow up should be done to this due to high rate of recurrence.

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Author contribution
Suskhan Djusad: concept, operator, data analysis, revising, final approval.
Yulia Margareta Sari: data collection, data analysis, drafting, writing the paper.
Hartono Tjahjadi: histopathology analysis.

Guarantor
Suskhan Djusad MD.
Yulia Margareta Sari MD.
Hartono Tjahjadi MD.

Declaration of competing interest
None declared.

Appendix A. Supplementary data
Supplementary data to this article can be found online at https://doi.org/10.1016/j.ijscr.2021.105948.

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