INTRODUCTION

Deep Aggressive Angiomyxoma (DAA) is a slow-growing and locally invasive benign mesenchymal tumor derived from myxoid cells, a rare pelvic connective tissue. It was first reported in 1983 and about 250 cases were detected around the world. This tumor is commonly found in women of reproductive age, with the highest incidence in the 3rd and 4th decades of life. DAA is most often found in the pelvic, genital, inguinal region, or perineum.

This tumor has a moderate to high risk of relapse and can grow to a considerable size, which is disturbing for patients, especially those of reproductive age. The nature of the tumor should be evaluated carefully using radiological and pathological approaches. A wide excision can remove the tumor completely, but control and follow-up are mandatory to evaluate the completeness of the procedure and monitor for any relapse.

CASE REPORT

A nulligravida woman, 52 years old, with a vulvar mass that was felt to slowly enlarge to a size of ±50 cm without any other complaints. The patient was successfully managed with wide excision without injection of a gonadotropin-releasing hormone (GnRH) agonist. After one year of postoperative evaluation, no recurrence occurred.
the uterus was normal size with no abnormalities in the cervix and vagina.

Supporting Examination

On blood laboratory examination, hemoglobin levels were 10.4 g/dL, total leukocytes were 28,000, and platelets were 268,000. Blood sugar examination was 116 mg/dL, and liver function was within normal limits. Kidney function increased with urea levels of 169 mg/dL and creatinine of 2.1 mg/dL—coagulation factors within normal limits.

Gynecological ultrasound examination revealed that the uterus and both adnexa were within normal limits, and there was no free intra-abdominal fluid. On ultrasound examination of the abdomen, there was a solid mass with calcification in the vulvar region (the size was not accessible to the probe), no nodules or suspicion of metastases were seen, mild left hydroureter, and increased echogenicity of the right and left renal cortex (Brenbridge I), suspicious of a chronic process of both kidneys.

On Magnetic Resonance Imaging (MRI) examination of the pelvis, it was found that the right vulvar region showed a well-defined lesion (which visualized the size of ± AP 14.95 x LL 31.80 x CC 15.3 cm) with the signal intensity being hypointense on T1WI, hyperintense on T2WI and FatSat, restricted on DWI, after contrast injection no enhancement was seen. The lesion appeared attached and difficult to separate from the right labia majora, with Multiple lymphadenopathies in the right and left femoral and inguinal regions (largest size ± 2.10 x 1.02 cm in the right femoral region).

Treatment

The patient improved his general condition and with written informed consent, it was planned to do a wide excision of the right vulva. The procedure was performed in the lithotomy position under general anesthesia. A wide dissection of the right vulva was performed, and the mass was completely separated. A drain was placed on the right vulva, followed by a stent in the left ureter.

In post-excision measurements, the right vulvar mass size was 50x42x10cm, weighing 14.8kg. An open cut was made on the mass to obtain a springy, purplish-white, and shiny mass.

On histopathological examination (Figure 4) with Hematosilin-Eosin staining, the stroma of fibromyxoid connective tissue was swollen, hyperemic, and lightly polluted with PMN-leukocin, containing a distribution of small, spindle-stellate-shaped cells, mature fat cells, many medium-sized blood vessels with thick walls. The necrotic area is approximately 20% and there are no signs of malignancy according to the picture of Deep (Aggressive) Angiomyxoma. Blood vessels, 40x (a), erythrocytes out of blood vessels, 400x (b), thick walled vessels with spindle cells, 400x (c), lymphoplasmacytic cell infiltration, 400x (d), Ectatic blood vessel within myxoid stroma, 40x (e), spindle cells (black arrow) between lymphoid cells (red arrow), 400x (f).

Postoperatively, the patient recovered well. Drain production was minimal and was removed on the second postoperative day. Change dressing and vaginal toilet were conducted every day. The patient was discharged on the third postoperative day in good condition while still using a urinary catheter. Patients are recommended for control every three days for evaluation. On day 6, the urinary catheter was removed. On day 9, surgical sutures were removed. (Figure 5)

DISCUSSION

Aggressive Angiomyxoma (AAM), first described by Steeper and Rosai in 1983, defines as a slow-growing mass with a myxoid and vascular section of the tissue that commonly occurs in the pelvic and genital area of women of reproductive age. Patient generally complain of mass growth in the genital region without pain or disturbances in micturition and defecation.

In appearance, the mass can vary in size; in some literature, it is stated that the mass starts from a small size of ±3 cm to 60cm and has unclear boundaries. Due to the rarity of these cases, many misdiagnoses such as Bartholin’s cyst occur. Although the mass has indistinct borders, it is not a malignancy, so the main treatment for this diagnosis is the excision of the tumor with wide excision of ±1cm from the identified tumor margin.

Molecularly, the cellular origin of AAM is still unclear. However, chromosome alteration in the 12q13-15 region and aberrant HMGIC (DNA architectural factor gene) expression induced by

Figure 1. Patient condition before operation. A large supple mass can be observed originating from the patient’s vulva. No pain and hyperemia was observed, but inguinal lymphadenopathy was reported.

Figure 2. The pelvic MRI of the patient showing the location and boundaries of the tumor.
chromosomal translocation t(8;12) are found in several cases. Moreover, positive expression of vimentin, SMA, MSA, desmin, CD34, F8, ER, PR, and negative expression for S-100, CK, and CD68 was also reported, suggesting that AAM is characterized by differentiation into fibroblasts and muscle fibroblasts.

To effectively identify this tumor, a CT scan or MRI is often helpful. MRI examination shows a picture of hyperintensity with a “swirled” picture with indistinct boundaries, which is a strong characteristic of this tumor. Since this character cannot be obtained by CT scan, MRI often becomes the preferred supporting examination and can be used as an examination modality for recurrence events.11–13

From the histopathological picture, AAM’s hallmarks are an indistinct border, a vascular component composed of larger blood vessels with thicker walls, and a myxoid appearance on histopathological readings. It is consistent with the morphological picture when the mass was split into purplish-white tissue, shiny, and on histopathological examination found a swollen fibromyxoid connective tissue stroma accompanied by many medium-sized blood vessels with thick walls.4,6,14,15

Blood flow from the pelvis and reproductive organs is a direct branch of the internal iliac artery, a continuation of the abdominal aorta. With the supply of large blood vessels, the high rate of morphological enlargement of the mass and the recurrence rate of this disease is also high. The literature found that recurrence at 3-15 years after wide excision of AAM and recurrence at <6 months were found in patients with non-maximal excision due to the large AAM mass.1,3,9

No evidence-based recommendations are available for the postoperative management of AAM. Because of the high local recurrence rate and the possibility of metastasis, patients are advised to undergo long-term follow-up up to 15 years after primary excision. The recurrence of AAM is not only due to the high vascularity of the mass but also because AAM has estrogen and progesterone receptors, as evidenced by immunohistochemical examination of ER (Estrogen Receptor) and PR (Progesterone Receptor) in almost all cases.
examined. An immunohistochemical examination was not performed on the patient due to cost limitations. Based on the literature on patients of reproductive age, there can be a high recurrence rate (36-72%). Due to the high recurrence rate and the high positivity of ER and PR in this tumor, it can be considered for treatment using GnRH agonist to minimize the possibility of recurrence. However, long term use is not advised due to its adverse effects (Menopausal Symptoms and Osteoporosis).

CONCLUSION

Aggressive Angiomyxoma is a rare localized aggressive neoplasm, so the possibility of differential diagnosis should be considered if growths are found in the vulvovaginal, perineal, or pelvic area. Because of the high recurrence rate, appropriate operative and adjuvant diagnosis and management have an important role in the treatment of patients with Aggressive Angiomyxoma.

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CONFLICT OF INTEREST

The author reports no conflicts of interest in this work.

REFERENCES