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Case

Surgical Treatment of a Recurrent Aggressive Angiomyxoma: A Case Report and Review of the Literature

Nüks Agressif Angiomiksomanın Cerrahi Tedavisi: Bir Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ÖZET

Özet: Agressif AngioMiksoma (AAM) temel olarak pélvis ve perinede ortaya çıkan, yumuşak doku kaynaklı nadir benign bir tümördür. Nadir görülmesinden dolayı, 46 yaşında bir kadında 5 yıl sonra nüks eden bir agressif angiomiksoma olgusunu sunuyoruz.

Anahtar Kelimeler: Agressif Angimiksoma, cerrahi tedavi

ABSTRACT

Summary: Aggressive angiomyxoma (AAM) is a rare benign myxoid mesenchyme derived soft tissue tumor which occurs mainly in the female pelvis and perineum. Due to its rarity, we aimed to present a case where aggressive angiomyxoma relapsed after 5 years in pelvis of a 46 year old woman.

Key words: Aggressive angiomyxoma, surgical treatment

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Introduction

Aggressive angiomyxoma (AAM) is a rare benign myxoid mesenchyme derived soft tissue tumor which occurs mainly in the female pelvis and perineum [1]. It was first described by Steeper TA and Rosai J as a distinctive soft tissue tumor in 1983 and the term aroused from the properties of the condition reflecting its locally infiltrative and recurrent behavior, neoplastic nature of the blood vessels involved and myxoid components [2]. AAM is usually seen in fertile ages, peak incidence is in the fourth decade of life [3]. The most evident difference from myxomas is that the myxomas have extremely small number of recognizable vessels [2]. The female to male ratio is reported to be 6,6/1, 7,45/1 and 8,5/1 with a peak incidence in women at 35 to 40 years age [1, 4, 5]. AAM lacks a capsule and grows slowly with local infiltration up to sizes 60 cm in diameter [6]. Although AAM has the property to invade adjacent structures, mostly in the histopathological evaluation nuclear atypia and mitosis are absent [7].

There have been fewer than 350 AAM cases reported in the medical literature up to date. Pre-operative diagnosis is difficult due to its rarity and lack of typical presenting signs and symptoms. Surgical excision without causing significant morbidity with clear surgical margins stays at the first treatment option of this rare soft tissue tumor. The aim of this case report is to present the surgical treatment of a recurrent AAM following initial surgery after 5 years and review of the literature to contribute the knowledge about this rare entity that helps avoiding misdiagnosis.

Case

A 46-year-old woman admitted to our department with the complaint of abnormal uterine bleeding (AUB) for 5 months. Levonorgestrel intrauterine device was installed for AUB three months ago, but the symptoms of the patients had not been improved. In her past medical history, five years ago, a retrorectal mass was removed with laparotomy and the pathological result was aggressive angiomxoma. In her physical examination, a solid mass was palpable at the right vaginal wall through the deep pelvis. Pelvic MRI revealed a 10x8x14 cm non-homogenous hypoechoic mass between right pelvic floor and perineum with contrast enhancement. T2 weighted image of the pelvic mass showed high-density shadows and heterogeneous enhancement (Figure 1). There were no remarkable laboratory test results done for the solid mass. The patient was informed about the laparotomy for the removal of the solid mass and written consent was taken for the operation. During the laparotomy, there was no visible lesion at the pelvis. The only remarkable finding was the omental adhesion on the right side of the pelvis. Omental adhesions were dissected and with opening of the anterior broad ligament peritoneum, the retroperitoneal space was entered. There was still no visible mass at the first retroperitoneal inspection. After opening of the medial paravesical space, a solid whitish unusual mass was seen and a fixative suture was used to follow up the solid mass. The dissection was done caudally deeper in the pelvic floor though the right perineum. A 14 cm

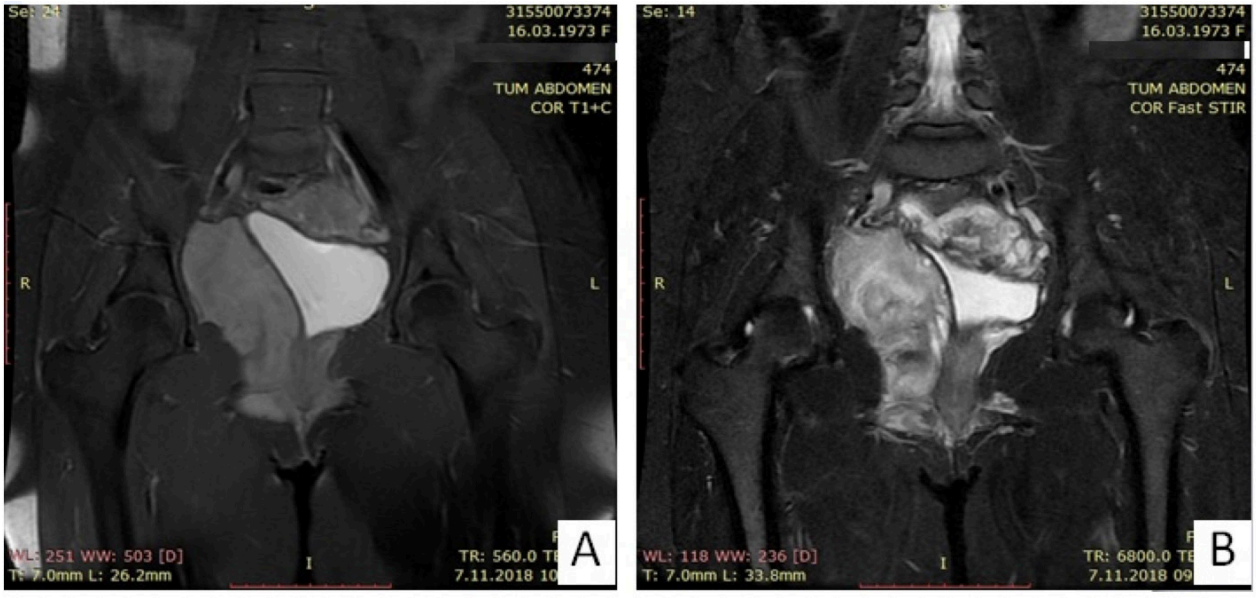

Figure 1. Coronal images of the pelvis with MRI prior to surgical treatment.

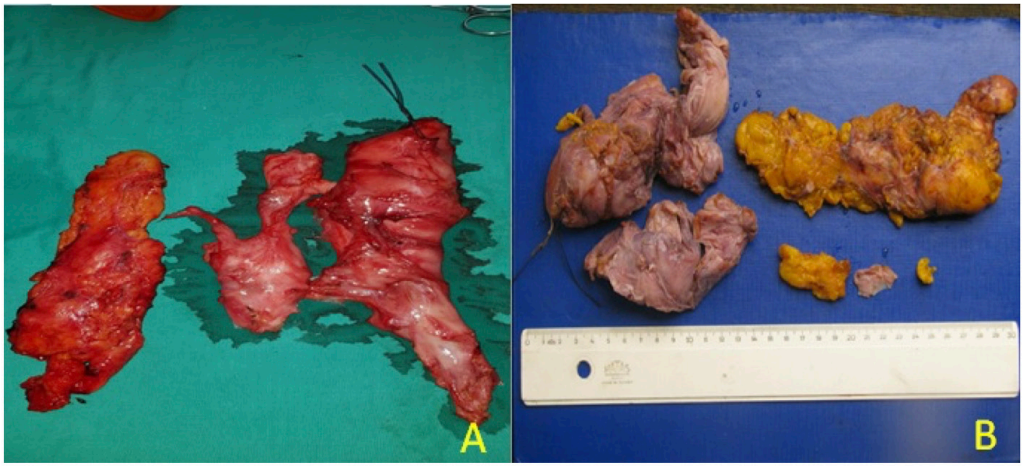

Figure 2. Macroscopic views of the tumor after resection. a-b) Tumor tissues with irregular margins on the left, bright gelatinous appearance in cross-section and invasive fat tissue on the right.

in length solid mass was removed (Figure 2). There was no complication during the operation. The tumor was attached by fibro-fatty-tissues with poorly defined border and soft and firm texture macroscopically. The pathological examination revealed thin-walled blood vessels within hypocellular myxoid tissue and short, spindled, stellar neoplastic cells containing fine collagen fibers and blood vessels that appear to be well within the myxoid edematous stroma when the specimen stained with hematoxylin and eosin. Tumor cells showed strong expression of desmin after using immunohistochemistry. The final pathological result was aggressive angiomixoma (Figure 3). The postoperative course of the patient was uneventful and she was discharged from hospital at the second postoperative day without any complication. The patient has been taken aromatase inhibitor treatment for prevention of recurrence.

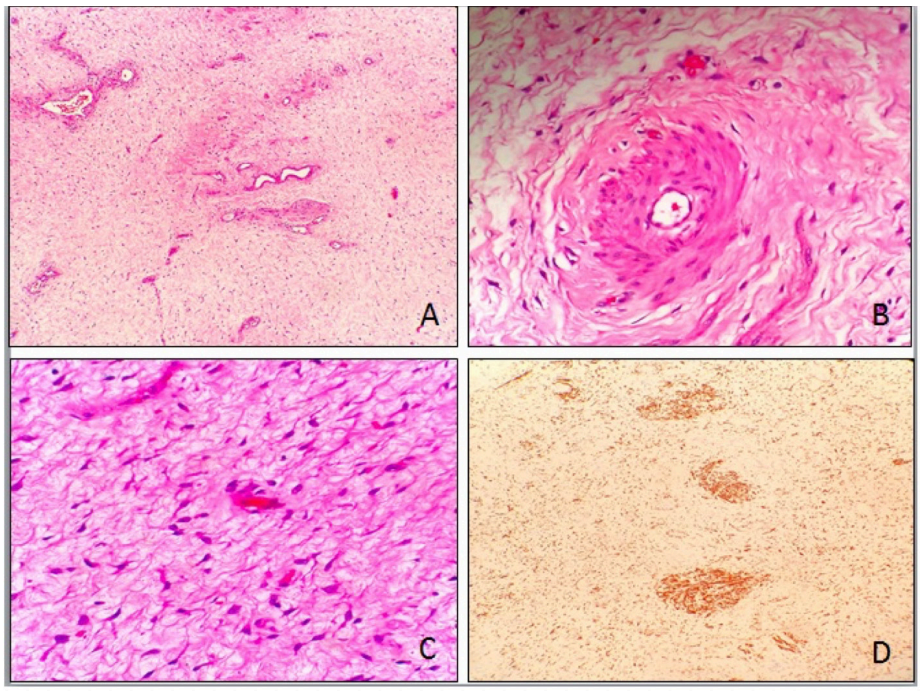

Figure 3. Microscopic finding during pathological examination. A) Thin-walled blood vessels in hypocellularmyxoid tissue (Hematoxylin Eosin x 100); B) Short, spindled or stellar neoplastic cells (Hematoxylin Eosin x 400) with fine collagen fibers and blood vessels, which appear well within the myxoid edematous stroma; C) One of the characteristic microscopic findings for aggressive angiomyxoma is the spread of smooth muscle cells from the vessel wall into the environment. Stroma consisting of fine collagen fibers in the environment (Hematoxylin Eosin x 400); D) Immunhistochemical staining with desmin in tumor (Desmin x 100).

Discussion

AAM is characterized by soft, non-encapsulated mass extending into the surrounding structures with finger like projections [8]. They displace the adjacent tissues rather than invading them[5]. However, after resection high rates of local recurrences are reported within a range of 36% to 83% [1, 4]. Smith et al reported the local recurrence rate was 58.3 in 17 AAM patients who underwent selective marginal resections [5]. Chan et al reported that a 47% local recurrence rate in 106 cases and 71% of recurrences was observed during the first three years after surgery [9]. The recurrent nature of this entity may take course even after 20 years [10]. Although it is reported to be of benign nature and non-metastasizing tumor, to date there are 3 cases reported with a distant lung metastasis [11-13].

The most common localizations of the AAM are pelvis-perineum (58%) and vulva (21%) [14]. Patients are often asymptomatic except a slow growing perineal or vaginal mass or a big mass palpable in abdomen and can easily be mistaken clinically for a Bartholin's cyst, abscess, hernia, lipoma, Gartner duct cyst, liposarcoma[10,15]. Misdiagnosis is very common for this tumor type as high as 82% at initial diagnosis of the patients [16]. For differential diagnosis a wide variety of benign and malign mesenchymal tumors such as myxoid liposarcoma, infiltrating angiolipoma, myxoma and angiomyofibroblastoma must be considered [17].

Due to similarity to that of healthy tissue macroscopically, type R-0 resection of AAM is difficult. The most prominent macroscopic features include smooth surface, appeared to be partially or complete encapsulation, finger like tumor projections. Glistering, gelatinous, semi-translucent blue-gray tumor of homogenous consistency with focal areas of congestion and hemorrhage are seen when the surface of the tumor is cut [2]. Immunohistochemical staining methods including desmin, vimentin, CD34, CD44, S100, Ki-67 and smooth muscle actin (SMA) are used for the diagnosis of AAM [18]. Usually AAM shows diffuse immunoreactivity for vimentin and desmin whereas SMA may be occasionally positive in individual tumor cells [3]. In the study of Jinping et al, the results of 71 immunochemistry examination revealed vimentin, SMA, desmin, CD34 were positive, the positive rate of PR was 70% and that of ER was 65%, S100 and CD68 were negative [4].

Preoperative computerized tomography (CT) or magnetic resonance imaging (MRI) also plays an important role in making of an accurate diagnosis considering its distinctive imaging appearance [19]. After enhancement on CT and MRI images a distinctive appearance of swirling or layered strands can be seen [20]. The tumor is detected hypo dense on CT and hyper intense on T2-weighted MRI images [21]. CT and MRI are also helpful in assessing the extent of the tumor thus can help us on the planning of optimal surgical strategy [19].

Also sonographic evaluation can help us to identify the highly vascularized structure of the tumor by color Doppler imaging technique and detecting the homogeneously hypo echoic mass with echogenic septa which are distinctive characteristics of AAM [7,17]. When suspected for AAM precaution must be taken during excisional biopsy or removal of the tumor, considering the risk of bleeding because of the tumor's vascularization[17].

Surgery is considered as the first line treatment for AAM and complete resection of the tumor is favored. Despite adequate surgical excision, recurrence rates up to 70% are described in the literature [7]. However due to the invasion tendency of this tumor to the surrounding tissues, sometimes a complete resection without causing an important morbidity is a hard task to achieve. Adjacent organs such as vagina, uterus, rectum, anal sphincter, bladder, and urethra are prone to be damaged when aimed to achieve a complete surgical resection sometimes resulting in urinary stress incontinence, faecal incontinence and colostomy [5]. Chan et al. reported that chance of remaining disease free was similar between the patients in whom a complete resection was made and the patients with tumor-involved resection (50% and 40% respectively). They concluded that incomplete resection is permissible, when high operative morbidity is anticipated and preservation of fertility is an issue [6]. Behranwala KA et al. also reported a spontaneous regression of the disease in one patient [22]. When we take into consideration these reports, we can propose that unless the condition gives disturbing symptoms this tumor can be monitored in certain intervals by imaging techniques such as MRI or CT to detect an unusual growth. Since the tumor itself doesn't kill the patient marginal resection can be considered as acceptable.

AAMs often express estrogen (E) and progesterone (P) receptors and can respond to hormonal management, therefore gonadotropin-releasing hormone analogs have been reported as a neoadjuvant therapy in

premenopausal patients before surgery to provide the shrinkage of the tumor for accomplishing an uncomplicated and less radical surgery [1, 19]. Schwartz PE et al. reported a 32 year old woman who had 7 recurrences of AAM managed surgically and the successful use of leuprolide acetate for preventing recurrences [19]. On the other hand, osteoporosis or major depression are significant adverse side effects of prolonged use of GnRh analogues and prolonged use should be considered in certain cases to prevent recurrence of the disease and in the risk of major surgical morbidity.

Also the use of aromatase inhibitors has been proposed as a treatment prior to surgery for decreasing the size of the mass in E-P receptor positive tumors in postmenopausal women [19,23].

Radiation therapy or chemotherapy is unlikely to be considered as an adjunctive therapy because of the tumor's low mitotic activity [24].

Conclusion

When a slow growing perineal mass is detected, especially in women of reproductive age, a high level of suspicion is needed for not to misdiagnose AAM. Because of the high recurrence rates up to 83% and a possibility of recurrence after a long time period, long-term follow-up is at most importance for these patients. A policy of routine surveillance of the patients with annual pelvic MRI should be helpful for the follow-up after surgical resection. Hormonal therapy can also be considered as a valuable treatment option especially in preventing the recurrences.

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