

Aggressive Angiomixoma of the Vulva: Case Report

Vulvanın Agresif Anjiyomiksomu

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Geliş Tarihi/Received: 20.11.2013
Kabul Tarihi/Accepted: 16.08.2014

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ABSTRACT Aggressive angiomixoma (AA) is an uncommon mesenchymal tumour of epidermis with local recurrence and rare metastasis. AA is treated with total excision and often lately diagnosed or misdiagnosed because of slow growth and similar features as the lipoma, vaginal cysts and bartholin cysts. Total excision of AA is the main treatment and provides curative therapy. In this case report, we present a 38-year-old woman with a vulvar mass diagnosed as aggressive angiomixoma and underwent total excision procedure.

Key Words: Myxoma; vulva

ÖZET Agresif anjiyomiksoma (AA) nadir görülen, lokal rekürrens ve nadir metastaz özelliği olan benign mezenkimal tümör olup epidermis kökenlidir. AA'nın tedavisi total eksizyondur; yavaş büyümesi, lipom, bartholin kisti ve vajinal kistlerle benzer görünümde olması nedeniyle genellikle yanlış ya da geç tanı almaktadır. AA'nın gerçek tedavisi total eksizyon olup bu prosedür genellikle kür sağlamaktadır. Bu yazıda, 38 yaşında vulvadan köken alan ve histopatolojik olarak anjiyomiksoma tanısı konulan ve total eksizyon yapılan bir olgu sunulmuştur.

Anahtar Kelimeler: Miksoma; vulva

Türkiye Klinikleri J Case Rep 2015;23(3):326-8

Aggressive angiomixoma (AA) is a slow-growing, painless, rare mesenchymal tumour originating from the epidermis.¹ One of the lower genital tract diseases, angiomixoma, is more frequent in the reproductive age than postmenopausal period.² The aim of this paper was presentation of a case with histopathologically diagnosed AA originating from the vulva.

CASE REPORT

A 38-year-old woman was referred to our clinic with the diagnosis of a vulvar mass. Her medical history included two vaginal deliveries. She complained of painless swelling in the right labium majus region that worsened progressively for the last two years. On clinical examination, there was a 5x6 centimeter (cm), soft viscous mass with a thin peduncle in the right labium

doi: 10.5336/caserep.2013-38163

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majus. There was no other pathology in pelvic examination and ultrasonographic evaluation of the patient. Total excision procedure was performed with uneventful intraoperative and postoperative course. Macroscopically, the lesion was 5x6 cm in size and brown-colored with homogenous appearance without necrosis or hemorrhage on the surface (Figure 1). Histopathological examination revealed loose myxoid stroma and spindle or star-shaped stromal cells with scattered small blood vessels using hematoxylin and eosin (HE) (Figure 2). The mass was diagnosed as vulvar angiomyxoma. Immunohistochemical evaluation for diagnosis was not performed, because there was no suspicion about the diagnosis of vulvar angiomyxoma. The surgical margin for tumor was detected as negative. Patient was taken into a follow-up program at regular intervals because of the local recurrence risk. There was no recurrence of tumor during one year of follow-up. Written informed consent was obtained from the patient for presentation of case report.

DISCUSSION

Aggressive angiomyxoma is a type of benign mesenchymal tumor with a slow-growing mass frequent local recurrence rate and rare metastasis. It was described in 1983 for the first time by Rosai and Steeper.¹ AA usually appears aged between 16 and 70 years (median 34 years).³ Because of the similar appearance with the lipoma, vaginal cysts and bartholin cysts and slow growth, AA is often diagnosed lately or misdiagnosed. Despite slow-



FIGURE 1: The macroscopic image of tumor after excision.

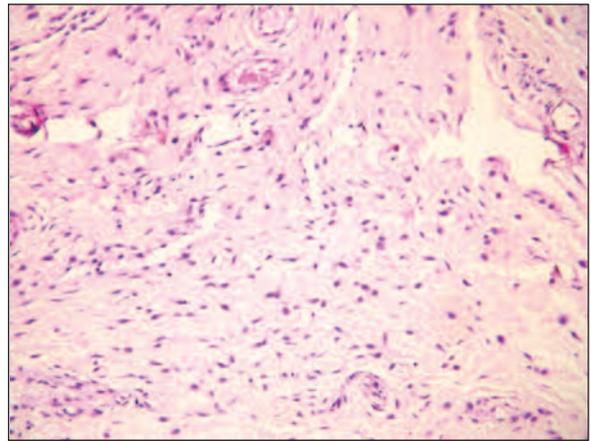


FIGURE 2: The microscopic view of tumor. Myxoid stroma and star-shaped stromal cells with scattered small blood vessels (HE, x30).

growth, AA could reach to a great size (up to 60 cm), but usually the largest diameter is about 10-20 cm.^{4,5}

Ultrasound, computerized tomography (CT) and magnetic resonance imaging (MRI) may be used for diagnosis of AA which reaches into the deep pelvic region. Ultrasound shows large perineal mass with predominantly echogenic low resistance type of arterial blood flow at different places. CT scan shows well-defined margins and MRI shows high signal intensity related to loose myxoid matrix and high water content of angiomyxoma.⁶ In our case, there was a slow-growing and longstanding vulvar mass. As differential diagnosis, bartholin cysts, vaginal cysts and vulvar lipoma were excluded through histopathological examination.

Microscopically, AA is characterized by widely scattered bland spindle cells with ill-defined cytoplasm within a myxoid background, plenty of collagen fibers, loose connective tissue, the stromal spindle cell septa, blood vessels using HE.⁷ Mitotic figures and nuclear atypia are almost always absent.⁸ In this case, microscopic view and staining properties were found similar to the literature data. On the other hand, AA mostly express different combinations of oestrogen and progesterone receptors, vimentin, desmin, smooth-muscle actin, muscle-specific action, CD34, and CD44, but all are invariably negative for S-100, CEA, and keratin, immunohistochemically.^{4,9}

Total excision of AA is the main treatment and provides curative therapy. Hormone therapy such as gonadotropin-releasing hormone agonist and tamoxifene or angiographic embolization are used commonly as an alternative therapy to radical excision or for treating recurrent tumor.^{10,11} Radiation therapy and chemotherapy are not used for treatment due to the low mitotic activity of this tumour. In the present case, total excision of the mass with the peduncle was performed and the patient was informed about the risk for local recurrence. Because total excision was performed and the patient

was closely followed up, radical adjuvant treatment was not planned. AA has a high risk of local infiltration and recurrence (50% to 70% of patients experiencing a relapse after surgical resection) after complete excision.¹² Recurrences generally occur in the first 5 years after primary surgery, but late recurrences up to 20 years have been reported.¹³

Consequently, the possibility of aggressive angiomyxoma should be kept in mind in large vulvar masses with or without a peduncle and close follow-up following total excision should not be neglected to prevent local recurrence.

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