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Case Report

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A Rare Lesion in Perianal Area, Angiomyxoma: Case Report

Perianal Bölgenin Nadir Görülen Bir Lezyonu, Anjiyomiksoma: Olgu Sunumu

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ABSTRACT

Angiomyxoma is a soft-tissue lesion found particularly in women who are of childbearing age. It is mostly encountered in perineal, pelvic, and gluteal area. Due to its recurring nature, surgical intervention is necessary. In our case, a 39-year-old female patient who was operated with the perianal hemangioma pre-diagnosis and whose pathologic evaluation was reported as angiomyxoma was presented.

Keywords: Angiomyxoma; perianal; surgery.

ÖZET

Anjiomiksoma özellikle doğurganlık çağındaki kadınlarda nadir görülen bir yumuşak doku lezyonudur. Çoğunlukla perine, pelvis, gluteal bölgede ortaya çıkar. Yüksek oranda rekürrens göstermesi nedeniyle cerrahisi önem taşımaktadır. Olgumuzda perineal hemanjiyom ön tanısıyla opere edilen, patolojik değerlendirmesi anjiyomiksoma olarak rapor edilen 39 yaşındaki kadın hasta sunulmuştur.

Anahtar sözcükler: Anjyomiksoma; perianal; cerrahi.

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Angiomyxoma is a local, infiltrative, and mesenchymal tumor that is usually seen in perianal and gluteal areas in women in their 40s.[1] Although it is a benign tumor, its rates of local recurrence are high. It is frequently misdiagnosed as lipoma, Bartholin's cyst, hemangioma, abscess, or hernia. These tumors usually appear as slow growing, deeply located pelvic masses.[2] In the literature, very few cases of extrapelvic masses are reported.[3] As radiological and clinical findings are non-specific, it is difficult to make a definitive pre-operative diagnosis. In this study, we planned to examine the process of diagnosis and treatment of angiomyxoma in the case of a 39-year-old female patient who experienced an aggressive angiomyxoma resulting in a swelling in her left pelvic area which has been present of 5 years and has been steadily growing in the past year.

Case Report

A 39-year-old female patient consulted our polyclinic with the presenting complaint that she experienced painless swelling on her left gluteal area. The patients' history revealed that she had this complained for 5 years, with an increase in swelling in the past year. She had no comorbidities and no operation history in the pelvic area. The superficial tissue ultrasonography revealed a lesion with lobular contours displaying vascular structures, extending into the subcutaneous and deep tissue areas in the left gluteal and pelvivulvar areas which were interpreted to be hemangioma. In addition, the patient had a pelvic magnetic rezonans (MR) imaging done. Pelvic MR imaging revealed a lesion measuring approximately 12.5×5 cm in the

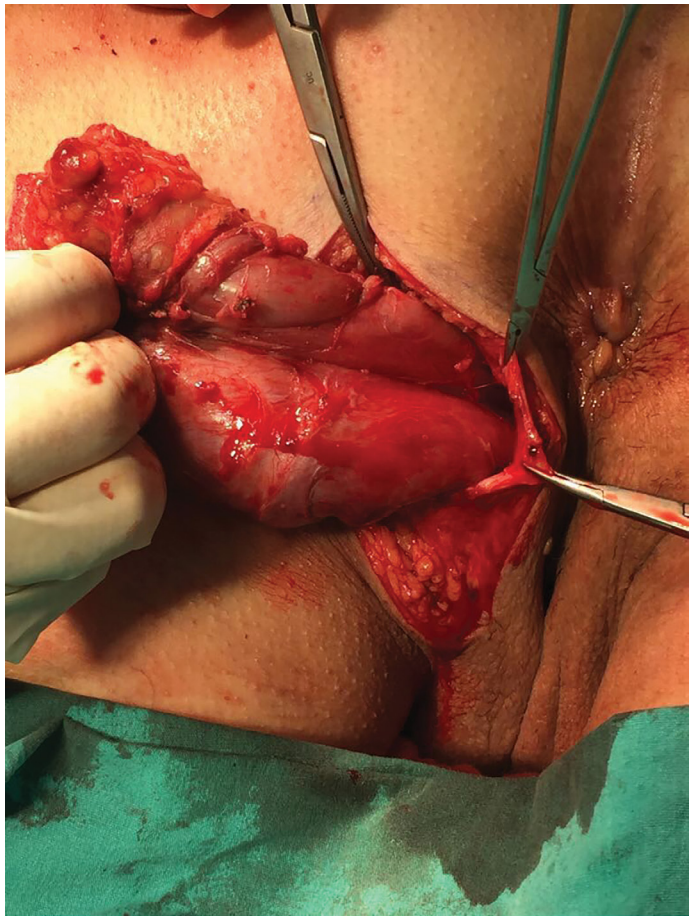

Figure 1. Angiomyxoma excised by perianal region.

left vulvar area extending into the posteroinferior, which was thought to be hemangioma. The patient was operated with the pre-diagnosis of hemangioma. The mass was excised by means of an ellipsoid incision performed immediately around it (Fig. 1). The pathologic evaluation of the mass was reported as angiomyxoma (Fig. 2). The surgical margins were clean and efficacious. The patient was discharged without complications on the 4th day after the operation. Patient's 1-year follow-up procedures revealed no relapses.

Discussion

Angiomyxoma is a rare benign tumor predominantly presenting in pelviperineal areas of adult patients. It was first identified in 1983 by Steeper and Rosai. It is mostly seen in women of reproductive age.[4] The female to male ratio is 6/1.[2] It can often be misdiagnosed as a labial hernia, lipoma, hemangioma, levator hernia, and pelvic hernia.[5] Although it is a benign and slow growing tumor, angiomyxoma's local recurrence rate in a 2-year period can be as high as 70%.

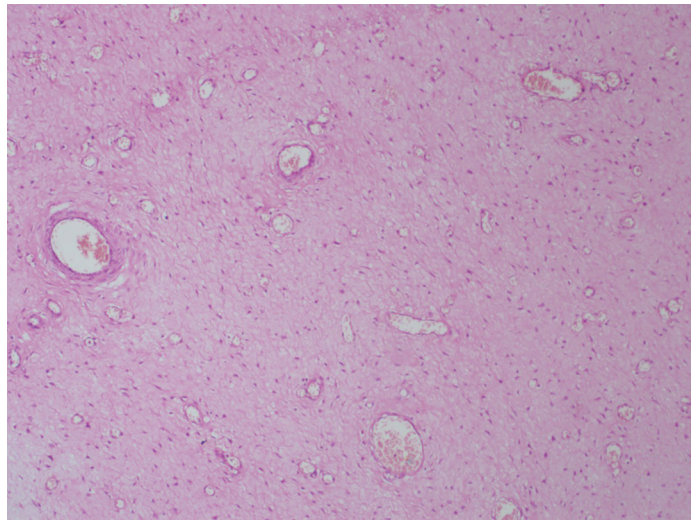

Figure 2. Histological analysis of angiomyxoma.

When the tumor is small in size, it is usually asymptomatic. It becomes symptomatic when it reaches large dimensions. The diagnosis of the tumor when it is small can only be made by incidental detection in imaging methods performed for other reasons for that region. Pre-operational CT and MR scans can provide detailed information on the tumor dissemination and the pelvic and adjacent organ involvement. These data can also help to plan the surgical approach. In the case, we presented that both USG and MR screenings were made. The relationship of the previously diagnosed hemangioma lesion with the surrounding soft tissue could be revealed more clearly by MR.

Although it is possible to make a diagnosis with a pre-operative biopsy, the definitive tissue diagnosis can be obtained postoperatively.[6] Since the pre-operative pre-diagnosis in our case was hemangioma, biopsy was not considered for initial diagnosis.

Main treatment is surgical resection. Achieving a clean surgical margin can be difficult due to the lack of a real cyst.[7] In patients with asymptomatic and small tumor size, for whom surgery may be anatomically risky, follow-up may also be possible. Hormonal therapy, including raloxifene, tamoxifen, and gonadotropin-releasing hormone analogs, can be used to shrink the tumor before excision and treat recurrences. The reason for this is the estrogen and progesterone receptors histologically owned by the tumor. However, the studies with larger numbers of patients are needed to show definite benefit. In addition, since all patients diagnosed with angiomyxoma are not women and are not in the premenopausal period, its efficacy in all patients is controversial. Nevertheless, it can be used in tumor recurrences

and as post-operative adjuvant therapy, depending on patient characteristics.[8] Chemotherapy and radiotherapy are not considered as treatment options due to the low mitotic efficacy of the tumor.[9] However, Rhomberg et al.[10] found that adjuvant radiotherapy combined with the sensitizer razoxane stopped the spread of the disease in a woman with a fourth relapse for an unknown time.

The tumor originates from the blood vessel walls. Histopathologically, the tumor appears as medium-sized structures of blood vessels within a myxoid stroma with loosened lumens, and randomly distributed fusiform mesenchymal cells.[11] In immunohistochemical studies, vimentin, desmin, actin, and CD34 were detected positively. In addition, estrogen and progesterone receptors frequently showed.[12] Although these properties of angiomyxoma are non-specific, they are still useful for diagnosis.

Despite the fact that it is a benign tumor, there are cases resulting in death in the literature.[13] In any case, it is a tumor with good prognosis. The main problem in this tumor is its high rates of local recurrence. In a retrospective case review of 106 cases, Chan et al.[14] discovered a recurrence rate of 71% that occurred in the first 3 years. In addition, it was determined that there was no variation of recurrence rates between positive surgical margin cases and clean surgical margin cases.

Conclusion

Angiomyxoma is a benign tumor with high rates of recurrence predominantly seen in female patients of childbearing age. It should be considered as a differential diagnosis in female patients with complaints of palpable mass in the pelvic area. The gold standard treatment is surgical resection with clean margins. The need and type of adjuvant treatment vary according to the patient. Post-operative follow-up is important due to the high rate of recurrence.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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