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Case Report

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AGGRESSIVE ANGIOMYXOMA OF VULVA: A CASE REPORT

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ABSTRACT

Aggressive angiomyxoma is an uncommon slow-growing mesenchymal tumor mostly affects women of reproductive age in the pelvic and perineal region. Diagnosis is difficult not only due to its rarity, but also owing to the resemblance of its clinical and imaging features with other mesenchymal tumors occurring in this region. The authors report a case of vulvar aggressive angiomyxoma in a menopausal woman which was confirmed by histologic examination after wide excision.

KEYWORDS: Mesenchymal tumors; aggressive angiomyxoma; surgery.

INTRODUCTION

Aggressive Angiomyxoma (AA) is a rare mesenchymal neoplasm occurring usually in women of reproductive age. The tumor was named aggressive due to its characteristically slow and insidious growth as well as carrying a high risk of local recurrence after excision with a significant morbidity. We present a case of AA arising in the vulva with the goal of increasing awareness of this uncommon entity among clinicians and pathologists for early diagnosis and appropriate management.

CASE REPORT

A 48-year-old menopausal woman referred to our department for a slow-growing mass on the left labia majora since two years. Three years ago she underwent hysterectomy for uterine polymyoma. Physical examination showed a painless vulvar mass of the left labia majora, measuring 7cm × 8cm, which was not attached to the superficial or the deep tissues. The skin was free of lesion and there was a dilatation of the subcutaneous venous plexus (Figure 1). No inguinal lymph nodes were detected. Abdomino-pelvic Computed Tomography (CT) scan revealed an oval and hypodense mass developed within the left labia majora and measured 5,8cm × 6cm without lymph nodes (Figure 2). Under spinal anesthesia, complete excision of the mass with clear margins was done and sent for histopathological examination. Grossly, the mass was partially polylobulated in shape without capsule. Cut section was solid, grey, without areas of hemorrhage or necrosis. Microscopic examination objectified an abundant myxoid stroma, stellate stromal cells without atypism and alternating small and large vessels (Figure 3). The pathologic report was aggressive angiomyxoma with negative margins. Her postoperative course was uneventful and she still being followed-up with no local recurrence.

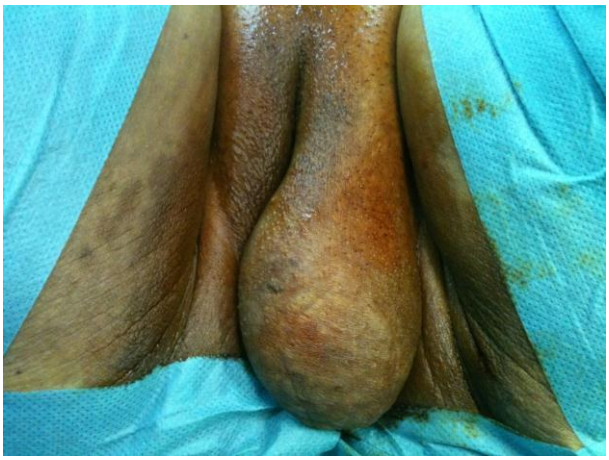

Figure 1: Swelling of the left labia majora.

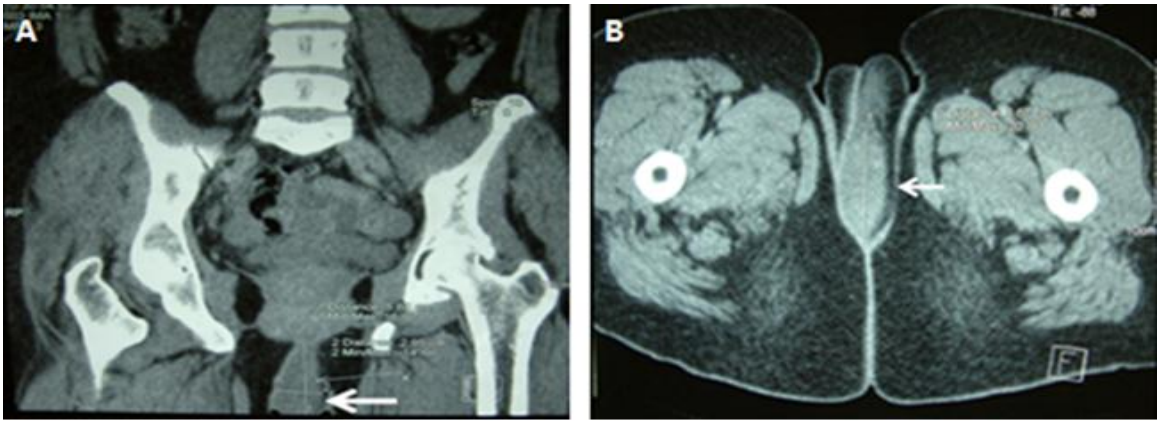

Figure 2: CT scan (A: Coronal section/ B: Axial section) showing an ovoid hypodense mass in the left labia majora (A: white arrow), heterogeneous after contrast-enhanced (B: white arrow).

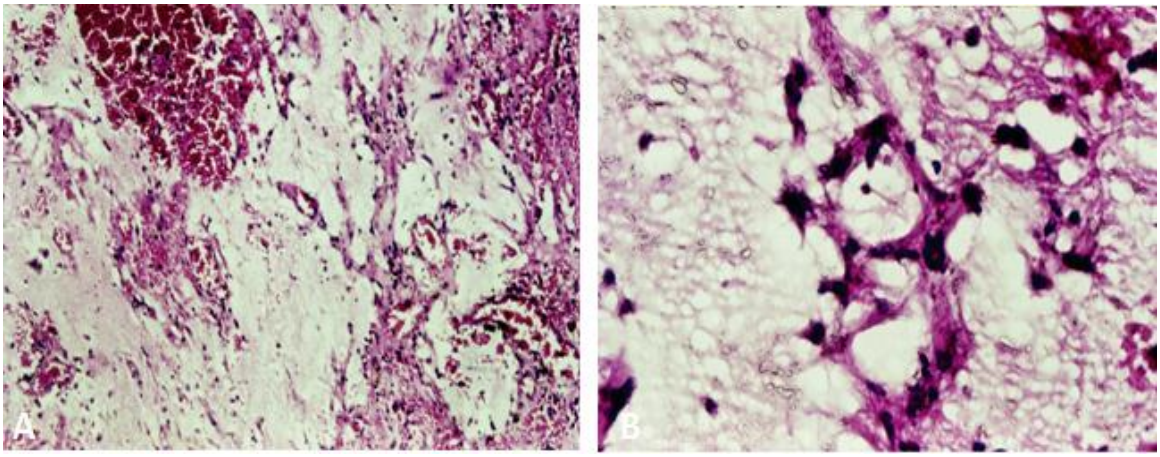

Figure 3: Stellate-shaped cells in a myxoid stroma with congestive vessels (A: HEx40/ B: HEx100).

DISCUSSION

AA is a rare mesenchymal tumor affecting predominantly women in the pelvic and perineal region. Other rarer locations have been also reported including: urinary tract, uterine cavity, perianal area, and inguinoscrotal region in man.[1,2] It mainly affects women between 20 and 60 years, with a higher incidence around 35-40 years. In man, it is seen at later age during the sixth or seventh decade. The tumor can also occur in children, the youngest patient was one year old.[3] There is no consensus regarding the pathogenesis of AA. This hormonally responsive tumor is believed to arise from specialized mesenchymal cells of the pelvi-perineal region or from the multipotent perivascular progenitor cells, which often display variable myofibroblastic and fibroblastic features.[4] Cytogenetic and molecular studies have revealed a variety of genetic alterations involving the specific breakpoint region 12q13-15.[5] A gene in this region called high-mobility group protein isoform I-C (HMGI-C), which encodes proteins involved in the transcriptional regulation, appears to have a role in the pathogenesis of this tumor.[4,5] Clinically, symptoms of AA are nonspecific; thus, it is mostly mistaken for more common entities such as a Bartholin cyst, lipoma, angiomyofibroblastoma, superficial angiomyxoma, cellular angiofibroma and inguinal hernia. The true extent of AA is often underestimated on clinical examination because the visible portion of the tumor represents only a fraction of the more-extensive involvement of the deep soft tissues of the pelvis and retroperitoneum.[6] On CT, the appearance of AA is variable; it may be a well-circumscribed and homogeneous mass that is hypodense relative to muscle, or it may be predominantly cystic with solid components.[7] Its Characteristics on Magnetic Resonance imaging (MRI) include hypointensity on T1-weighted images and hyperintensity on T2-weighted images. Both techniques, CT and MRI, allow staging by determining the extension beyond the pelvic diaphragm which determines the surgical planning. However, MRI is the best tool because of its better soft tissue contrast and resolution in pelvis. In addition, MRI is useful to detect residual tumor and recurrences in the postoperative follow-up which have the same characteristics as the original tumor. Macroscopically, AA is unencapsulated, poorly or vaguely circumscribed, and may blend imperceptibly with surrounding soft tissue. Tumor size is highly variable and ranges from 1 cm to 60 cm.[6] It has a gelatinous consistency and may include cystic, hemorrhage or fibrosis areas. Microscopically, our patient had the typical histologic characteristics of AAs including abundant myxoid stroma, uniform low cellular spindle or stellate stromal cells, and alternating small and large vessels. In

immunohistochemy, most AAs express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth muscle actin, CD34 and CD44; but all are invariably negative for S-100, carcinoembryonic antigen and keratin.[8] Surgical excision is the traditional treatment of AAs. This surgery should be as wide as possible to prevent the risk of local relapse. When fertility is to be preserved or surgery is likely to be extensive and mutilating, incomplete resection is acceptable as local recurrences can be treated with further resection. Unfortunately, relapse may still occur with negative margins, it may occur from months to several years after resection. Recurrence rates from the largest case series range from 25% to 47%, with 85% of those occurring within 5 years of initial surgery.[9,10,11] Radiotherapy and chemotherapy are considered less suitable options due to low mitotic activity of the tumor. Hormonal therapy with Tamoxifen, Raloxifene and gonadotropin-releasing hormone agonist has been shown to reduce the tumor size and may help to make complete excision feasible in large tumors and in the treatment of recurrences.[12] Angiographic embolization described by some authors is difficult to realize, given the numerous vascular anastomosis of these neoplasms. Although exceptional cases of metastasis have been reported[13,14], AA is considered an indolent tumor with locally aggressive behavior. There are no specific guidelines for postoperative management of AA; due to high recurrence rate and potential morbidity associated with undiagnosed recurrences, several authors recommend a periodic evaluation with clinical examination and MRI up to 15 years after excision.[4]

CONCLUSION

AA must be considered in differential diagnosis of vulvoperineal mass. CT and MRI accurately reveal extension of these tumors across the pelvic diaphragm and are thus valuable in determining the surgical approach. Initial resection should be oncologic to avoid and minimize the risk of recurrence.

Conflict of interest: the authors declare no conflict of interest.

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