A unusual pelvic tumor-aggressive angiomyxoma

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ABSTRACT

Soft tissue lesions of the pelvic-perineal region are relatively rare. They often have diagnostic challenges, because of their overlapping morphological features. Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor, most commonly arising from the vulvovaginal region, perineum and pelvis of women. Due to its rarity, it is often initially misdiagnosed as a gynecological malignancy. Here, we present a young female with a pelvic mass, clinically diagnosed as ovarian tumor and histopathologically proven to be an AAM.

Key words: Aggressive angiomyxoma, Angiomyofibroblastoma, Myxoid

INTRODUCTION

Soft tissue lesions of pelvic-perineal region are relatively rare, they can be classified into two categories; those that are relatively site specific and those that occurred in other regions and may also involve the pelvic region.¹ These lesions are often diagnostically challenging, because they may show overlapping morphological features.²⁻³ However, from the practical point of view, deep angiomyxoma is the most important entity, which needs to be recognized and discriminated, because of its propensity for recurrence.³⁻⁴

Case report

Here, we present a case of aggressive angiomyxoma (AAM), presenting as huge abdominal mass in a 40-year-old female. Ultrasonography of the abdomen revealed a huge pelvic mass and the possibility of a malignant ovarian tumor, was suggested by the radiologist. Computed tomography abdomen showed a heterogeneous mass in the pelvis with an attachment to the lateral vaginal wall.

We received the uterus with cervix, one side ovary, both fallopian tubes and a separate huge ovarian mass. Uterus with cervix measured 10 cm × 7 cm × 5 cm. The mass measured about 15 cm × 11 cm × 9 cm. On gross examination, the tumor was solid, lobulated with a soft to a rubbery consistency. The cut surface revealed a glistening, homogeneous mass with focal prominent vascularity. The other ovary was made out separately from the tumor [Figure 1].

Representative bits studied revealed, a paucicellular mesenchymal tumor, composed of fibroblasts within a myxoid background. Vascular proliferation was prominent with a virtual absence of mitoses. These features validated the diagnosis of AAM [Figure 2].

DISCUSSION

The term AAM was originally described by Steeper and Rosai in 1983,⁵ but the term “angiomyxoma” dates back to at least 1952.⁶ About 90% of patients are women, usually of reproductive age. It most commonly arises in the vulvovaginal region, perineum, and pelvis.⁵⁻⁶⁻⁷ A few cases have been described in males, usually in the scrotum.⁹ It presents as a painless, poorly circumscribed gelatinous mass and clinically simulates a Bartholin gland cyst or an inguinal hernia. The tumor grows slowly, and its benign nature is suggested by the histology also and that it shows no tendency to metastasize. However, it is locally aggressive and tends to recur locally (36-72%) after resection.⁸⁻⁹

On gross examination, AAM typically presents as a solid, lobulated, soft to a rubbery mass. The cut surface has a homogeneous, gelatinous appearance. Recurrent tumors show more prominent areas of hemorrhage and fibrosis.⁵⁻⁷⁻⁸ Histologically, it is paucicellular with deceptively infiltrative borders which accounts for its propensity to recur. Blunt spindle-shaped cells with delicate uninuclear or bipolar cytoplasmic processes are set within a myxoid stroma with interspersed medium to large-sized vessels that are often thick-walled and hyalinized. Virtually no mitoses are present.⁴⁻⁷⁻¹¹ The vast majority of cases demonstrates, positivity for desmin in the myxoid areas and/or stromal cells, while actin and CD34 may be variably positive.⁴⁻⁷⁻¹¹ The estrogen and progesterone receptor positivity suggests that the AAM might be hormone-dependent as rapid growth has been observed during pregnancy.¹²⁻¹³

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Fibroepithelial stromal polyps are polypoidal with a circumscribed border. The vascular component is located at the core of the polyp. In contrast, AAM has an infiltrative border, and the vascular component is haphazardly distributed.\textsuperscript{52,53}

Superficial angiomixoma occurs in superficial locations and has bland spindle cells. However, IHC can also be helpful as the spindle cells in angiomixoma are negative for desmin, whereas the spindle cells in AAM are positive for desmin.\textsuperscript{54}

Wide excision is curative, and the prognosis of such is good. Surgery remains the first line of treatment; however, adjuvant treatment, such as gonadotrophin-releasing hormone agonists, has been used for treatment of recurrence.\textsuperscript{6,8,9}

CONCLUSION

Although a rare diagnosis, AAM can present in an unusual location. Detailed radiological examination is helpful in suspecting the problem in advance, but histopathology is the gold standard for its diagnosis. Wide excision is curative of choice and prognosis of this tumor is generally good.

REFERENCES


