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Case Report

Aggressive angiomyxoma: case report and review of the literature

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ABSTRACT

A 47-year-old female presented to clinic with a 5-year history of a left buttock mass. The patient's hemoglobin was low (9.7 g/dL); laboratory analysis was otherwise unremarkable. Ultrasound of the left gluteal region demonstrated a heterogeneous vascular solid lesion. Magnetic resonance and computed tomography imaging showed an enhancing mass extending from the left ischioanal fossa through the levator ani muscle into the pelvis. Biopsy revealed bland-appearing spindle-shaped cells positive for estrogen and progesterone receptors, consistent with an aggressive angiomyxoma. The mass was surgically excised without complication. To date, follow-up imaging has not demonstrated evidence of tumor recurrence.

Introduction

Aggressive angiomyxomas are rare mesenchymal tumors that occur predominantly in females of reproductive age, with peak incidence typically in the 4th to 5th decades of life [1,2]. Patients are often asymptomatic at the time of diagnosis, with perineal or vulvar masses discovered incidentally during physical examination or radiologic imaging [1]. Although rare, aggressive angiomyxomas exhibit typical patterns on ultrasound, CT, and MR imaging: tumors nearly always occur within the perineal and/or pelvic region, and on MRI characteristically demonstrate a "swirled" appearance of relatively low-intensity internal stranding on both T1- and T2-weighted images [3–5]. Here, a case of aggressive angiomyxoma and corresponding review of the literature is presented, with the aim to familiarize clinicians with the clinical and imaging characteristics of these tumors.

Case report

On review of systems during a clinic visit, a 47-year-old female with poor prior medical care but otherwise unremarkable past medical history noted a 5-year history of a left buttock mass

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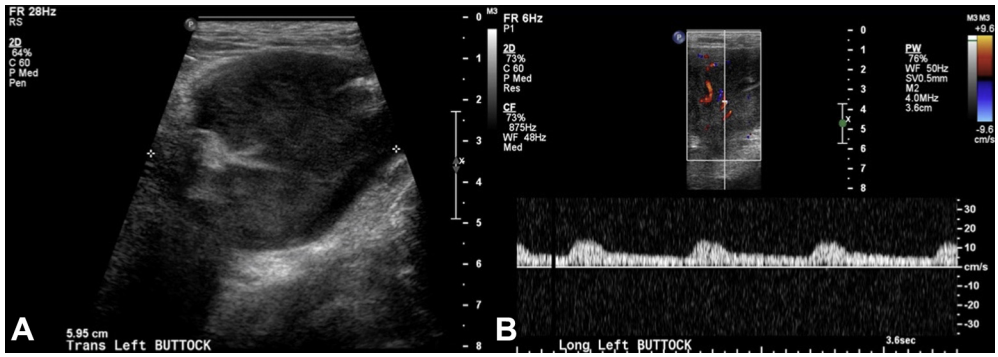

Fig. 1 – Ultrasound of the left gluteal region demonstrates a circumscribed, predominantly hypoechoic mass (A). Color Doppler evaluation of the mass reveals significant internal vascularity (B). The visualized portions of the mass measured approximately 6 cm in transverse dimensions; the intra-pelvic involvement of the mass was not detected sonographically.

that had increased in size over the past 12 months. On physical examination, a firm and nontender mass was palpated in the left gluteal region. The patient's hemoglobin was low (9.7 g/dL); a comprehensive laboratory analysis that included complete blood count and basic metabolic panel and hepatic panel was otherwise unremarkable. Ultrasound of the palpable mass was performed, which demonstrated a predominantly hypoechoic circumscribed solid lesion with internal blood flow (Fig. 1). The patient subsequently underwent CT (Fig. 2) and MRI (Fig. 3), which revealed an enhancing mass within left ischioanal fossa that extended through the pelvic floor musculature into the pelvis. The lesion displaced, but did not infiltrate, the urinary bladder and uterus. On MRI, the mass demonstrated concentric linear bands of T1 and T2 signal in a "swirled" pattern (Fig. 3). Incisional biopsy of the mass demonstrated bland-appearing spindle-shaped cells positive for estrogen and progesterone receptors, consistent with an aggressive angiomyxoma (Fig. 4). The patient underwent surgical excision of the mass approximately 3 weeks after her initial clinic visit (Fig. 4). Follow-up CT imaging has demonstrated disease-free status for 1 year.

Discussion

Aggressive angiomyxomas are rare tumors that typically occur in adult women, with a peak incidence between 30 and 50 years [1,2]. Female to male ratio is 6.6:1 [6]. The tumors most commonly occur within the perineal and/or pelvic region [3]. On sonographic imaging, aggressive angiomyxomas typically appear as a hypoechoic or cystic mass [7]. CT imaging typically demonstrates a mass with well-defined margins, slightly hypodense to muscle [8]. On MRI, these tumors are usually hyperintense on T2-weighted images, likely related to high water content and loose myxoid matrix [4,5,9]. On T1-weighted images, the tumors are isointense to muscle [4,5,9]. Characteristically, the mass will have internal areas of "swirled" linear low-intensity signal on both T1-weighted and T2-weighted images, thought to be related to the fibrovascular stroma [4,5,9]. The "swirled" appearance is less frequently appreciated on contrast-enhanced CT [4,5]. Aggressive angiomyxomas demonstrate significant contrast enhancement, likely due to the high internal vascularity [5].

On gross evaluation, the tumors are tan-gray to pink and have a rubbery consistency with a gelantinous and glistening cut surface [7]. Histologically, aggressive angiomyxomas are sparsely cellular, composed of spindle and stellate-shaped cells on a soft myxoid background [1,6–8]. Internal blood vessels characteristically demonstrate variable caliber and/or wall thickness; some vessels have thick, muscular walls which may be hyalinized [7,10]. Resection margins are often positive due to the tumor's proclivity to infiltrate local tissues [1]. Cells stain positive for estrogen and progesterone receptors as well as

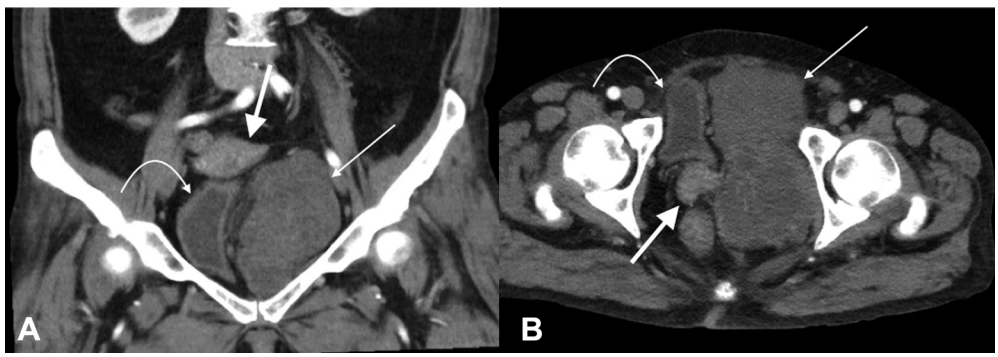

Fig. 2 – Coronal (A) and axial (B) contrast-enhanced CT of the pelvis. The mass (thin straight arrows) is located within the left side of the pelvis and has relatively circumscribed margins. The tumor causes mass effect on, but does not invade, the urinary bladder (curved arrows) and uterus (thick arrows).

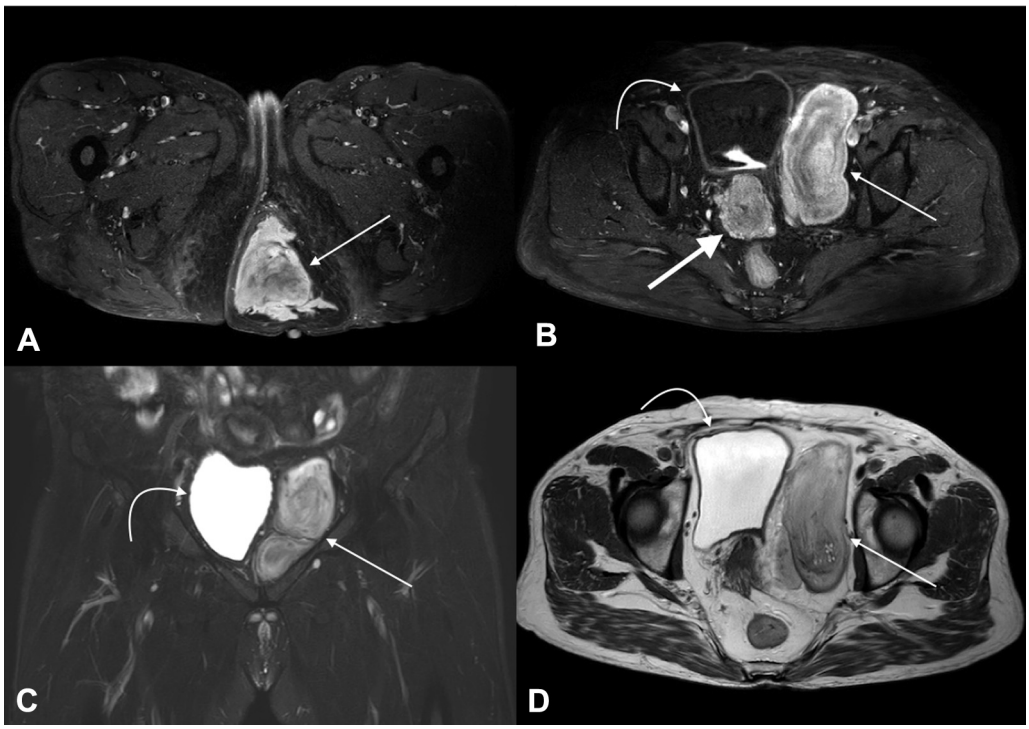

Fig. 3 – Axial contrast-enhanced T1 fat saturated images of the perineum (A) and pelvis (B), coronal T2-weighted fat saturated image (C), and axial proton density weighted image (D) of the pelvis. The tumor is seen extending from the left ischioanal fossa (thin arrow, (A)), through the levator ani muscle, into the pelvis (thin arrows, (B), (C), and (D)). The mass demonstrates marked enhancement and has internal low-intensity "swirled" signal on all image sequences. Adjacent structures, including the urinary bladder (curved arrows) and uterus (thick arrow), are shifted to the right but are not invaded by the tumor.

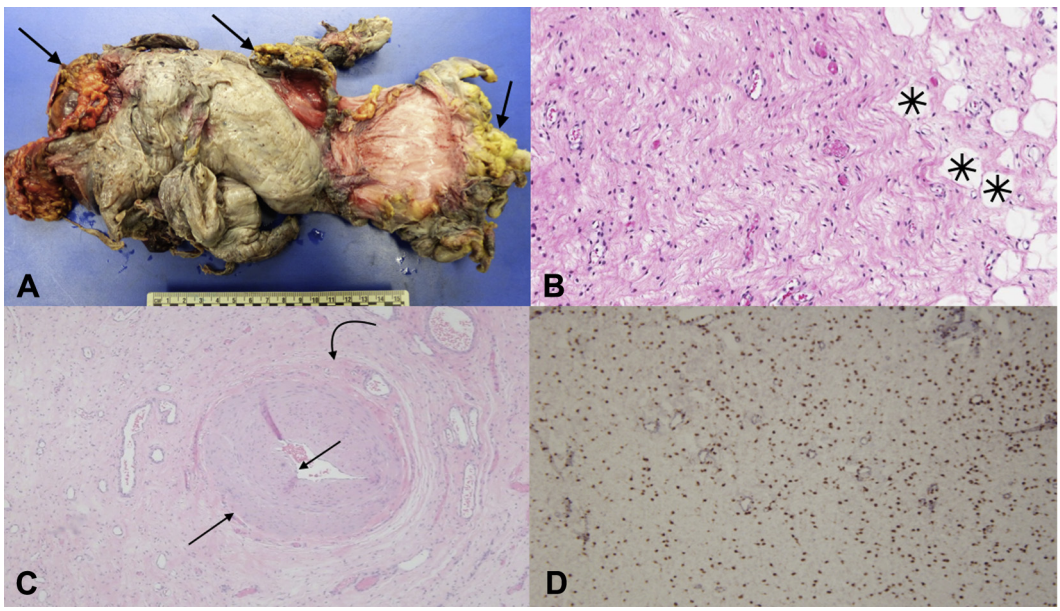

Fig. 4 – Gross pathology (A), H&E stained sections ((B) and (C)), and progesterone stained section (D) of the tumor. Gross evaluation demonstrates a lobulated mass with a tan-gray and pink surface; normal yellow fat (arrows in (A)) is noted along the periphery. Histologically, the tumor is composed of a uniform distribution of bland-appearing spindle-shaped cells (B), with local invasion of the adjacent fat (asterisks). The lesion is highly vascular, with some blood vessels showing characteristic thickened walls (between straight arrows, (C)) and hyalinization surrounding the vessel walls (curved arrow, (C)). The tumor stained positive for both progesterone (D) and estrogen (not shown) receptors.

desmin, vimentin, CD-34, and smooth muscle actin; S-100 and cytokeratins are usually negative [1,2,4,7].

The main differential consideration for aggressive angiomyxomas is angiomyofibroblastoma [11]. The tumor's size is a useful differentiating factor: angiomyofibroblastomas tend to be small and affect only the superficial vulva and vagina, whereas aggressive angiomyxomas are often large masses involving the deep tissues planes at the time of diagnosis [12]. Myxomas and myxoid liposarcomas are also considered as part of the differential. However, myxomas are located intramuscularly, whereas aggressive angiomxyomas may abut, but do not invade, the pelvic and/or perineal musculature [12]. Myxoid liposarcomas more commonly occur within the lower extremities, demonstrate lacy or linear internal fat, and homogenously enhance [12]. Aggressive angiomyxomas, conversely, lack significant internal fat and enhance heterogeneously [5,12].

Treatment of aggressive angiomxyomas involves wide local surgical excision [5,8,13]. Hormonal therapy, including raloxifene, tamoxifen, and gonadotropin-releasing hormone analogs, is also used to both shrink the tumor before excision and treat recurrences [5,12]. Chemotherapy and radiation are generally considered to be poor treatment options due to the tumor's low mitotic activity [8]. In addition, embolization is usually not used as the tumors frequently have numerous feeding vessels [11].

Although aggressive angiomyxomas are benign, local recurrence is quite common; "aggressive" was added as a prefix to the classification of these tumors to emphasize their infiltrative nature and tendency to recur [6,7,14]. In 1 case study of 73 subjects, 34 of 73 (47%) developed recurrent disease, with the rate of recurrence noted to be similar among patients with and without clear resection margins [6]. A few exceptionally rare cases have also been reported of metastatic aggressive angiomxyomas, including cases in which the patients succumbed to the disease [15]. Hence, long-term follow-up is recommended after surgical excision [6]. Nevertheless, prognosis is generally quite good [15].

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