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Review

Aggressive angiomyxoma: A case series and literature review

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Abstract

Background: Aggressive angiomyxoma was identified as a distinct clinicopathologic entity in 1983 and since then fewer than 250 cases of these rare tumors have been reported in world literature. These tumors usually arise in the pelvis and perineal regions, most often in women of the reproductive age group; however a few cases of its occurrence outside the pelvis have also been reported.
Patients and methods: We report a series of 7 women treated in our institute in the last 8 years. Relevant literature on aggressive angiomyxoma was looked at and various management options reviewed.
Conclusion: Aggressive angiomyxomas are locally aggressive, notorious for local recurrence and extremely rare to metastasize. While surgery remains the mainstay of treatment, there has been a definite shift towards less radical forms of excision, over the years. Various adjuvant treatment modalities have also been tried to reduce tumor recurrence.

Keywords: Management; Pregnancy; Aggressive; Angiomyxoma

Introduction

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoid mesenchymal neoplasm preferentially arising in the pelvic and perineal regions of young adult females, most commonly in the reproductive age group.[1] In men, the tumor involves analogous sites including the scrotum and inguinal area and usually appears at an older age.[2] AA was first described in 1983[3] and since then less than 250 cases have been reported in literature, mostly in the form of small case series or isolated case reports. Though most of these tumors are locally aggressive occasional distant metastasis has also been reported.[4,5]

Diagnosis, more often than not, is at histological examination following surgical resection. Pre-operative diagnosis is difficult because of rarity of these tumors and lack of typical presenting signs and symptoms. The tumor is characterised by a soft, non-encapsulated mass with finger like projections into the adjacent soft tissue.

Surgery is usually the first line of treatment. Incomplete excision is common because of the infiltrating nature of the neoplasm and absence of a definite capsule. Local recurrence is common even after complete excision. Non-surgical interventions like hormonal manipulation, radiotherapy, arterial embolization, etc. have been tried with variable success rates.

Patients and methods

From 2002–2009, 11 women with aggressive angiomyxoma were managed in our institution. Seven of these cases were directly managed in our centre while the others were referred for expert opinion only. Among the seven, one interesting case of AA in a pregnant woman (case 1 – Table 1) has previously been reported separately.[6] We also performed a MEDLINE search from 1983 to July 2009 with the key words "aggressive" and "angiomyxoma" and reviewed relevant literature.

Results

Table 1 summarizes the most relevant details for the 7 patients managed in our institution. In all but one case

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Table 1
Summary of patients with aggressive angiomyxoma (AA).

Case	Original presentation & size	Age at initial presentation	Location	Surgical treatment/ Outcome	Resection Margins	Adjuvant treatment	DFI/number of recurrences
1	Right sided 3 cm vulva swelling	22	Right vulva & vagina extending into right ichiorectal - fossa & levator ani (Fig 2)	3 × WLE/successful pregnancy after third surgery, currently stable size & awaiting	Positive	GnRH analogue after the first and second surgery	5 months after first WLE/3 recurrences
2	8 cm Lt vulval lesion with unsuccessful initial GnRH analogue treatment	47	Left vulva and perineum	WLE + 3 subsequent perianal abscesses drained	Positive	N	26 months/none
3	Bowel habit changes, urinary urgency, bulky uterus, 8-9 cm pelvic mass on exam	58	Left paravesical	Laparotomy & excision of paravesical mass.	Positive	N	11 months/none
4	5 year intermittent swelling in perineum with 8 cm mass on palpation	46	Left paravaginal and perineal area	Abdomino-perineal excision	Positive	N	16 months/none
5	Anterior vaginal wall mass	49	Anterior vaginal wall	WLE followed by VH and second WLE in 2 months	Negative	N	48 months/none
6	Recurrent left Bartholin's cyst diagnosed as AA after a year	44	Left vulva	WLE	Positive	GnRH analogues to shrink recurrent tumor	First recurrence at 24 months, currently 9 cm extending into left ischio-rectal fossa
7	Mistaken as cystocele	48	Vulvo-vaginal mass	WLE	Positive	N	48 months/none

GnRH analogue: gonadotropin-releasing hormone agonist therapy; DFI: disease Free Interval; WLE: Wide Local Excision.

(case no 2), the diagnosis of AA was made only after initial surgery. The average age at diagnosis in our group of patients was just under 45 years (range 22–58 years), with an average disease free interval (DFI) of over 25 months. In 6 out of 7 cases histopathology confirmed involved resection margins. Five women had no recurrence following initial surgery. AA did not recur in the patient with clear margin. Case no 1 did respond well to GnRH-agonist therapy each time however recurred after discontinuation of treatment. Case no 2 was given GnRH-agonist as a neo-adjuvant treatment with the aim of tumor reduction but the treatment was unsatisfactory and the patient had to undergo surgery.

The MEDLINE search yielded 93 single case reports of this rare neoplasm in women if limited to the perineum and pelvis, and thirty-four of them in men. Ten cases of AA are reported in other parts of the body as well as two rare cases of lung metastases. A further 98 patients including both sexes are reported in series ($n = 2–29$ cases) bringing the whole number of cases reported in the literature to under 250 since the first description of AA in 1983.

Discussion

Aggressive angiomyxomas are locally invasive connective tissue tumors presenting in about 90% of cases in women of reproductive age group with a peak incidence in the fourth decade of life.[7] The initial presentation varies from an asymptomatic perineal or vulval nodule/polyp or perineal hernia to a pelvic mass diagnosed on imaging studies. Occasionally, the tumor can be cystic and mistaken for a Bartholin's, labial or Gartner's duct cyst. They can exhibit a slow and insidious growth pattern and hence patients may be asymptomatic for long time until they become conscious of the significant size of the tumor. Tumor recurrences, like the primary ones, are often asymptomatic.

Pre-operative diagnosis is often a problem because of the rarity of these neoplasms and lack of typical features and most cases are diagnosed on histology after primary surgical excision. Accurate diagnosis before surgical excision is important in planning extent of excision and patient counseling.

Imaging characteristics of angiomyxomas

Ultrasound is helpful in this regard and usually demonstrates a hypo echoic soft tissue mass that may even appear cystic. CT appearances are variable and include a well-defined homogenous mass hypo dense relative to muscle, a hypo attenuating solid mass with swirling internal pattern with contrast or a predominantly cystic appearing mass with solid components.[8] With MRI imaging an iso intense or less commonly hypo intense mass (compared to muscle) on T1 and hyper intense on T2 weighted images is characteristic. MRI imaging is accurate in detecting trans-levator spread and also is helpful in surgical planning by defining relation of the mass with anal sphincter, urethra, bladder and pelvic side-wall.[8] Recurrent tumor is also detected consistently in MRI scans and recurrent lesions have similar signal characteristics as that of primary tumor.