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Research Paper

A Rare Case of Periurethral Aggressive Angiomyxoma

Medical Science

KEYWORDS : urethra; vagina; angiomyxoma

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INTRODUCTION

Aggressive angiomyxoma is a rare soft tissue tumor of the pelvic region developing from myxoid cells which are a type of cells found in body's connective tissue. Aggressive angiomyxoma is a rare neoplasm of the soft tissues affecting mostly female patients in the third decade of life. Male to female ratio is 1:6. It affects the genital and pelvic area and has a propensity for local recurrences. It is unique in being structurally benign and behaviorally locally malignant. This tumor was first described and named aggressive angiomyxoma by Steeper & Rosai in 1983.The diagnosis of aggressive angiomyxoma is usually made by the pathologist. Its differential diagnosis includes myxoma, myxoid liposarcoma, sarcoma botryoides, myxoid variant of malignant fibrous histiocytoma, nerve sheath myxoma and other soft tissue tumors with secondary myxoid changes.

The standard treatment for Aggressive angiomyxoma is complete wide excision with adequate margins and reconstruction whenever mandatory.

We report a rare case of periurethral aggressive angiomyxoma with review of relevant literature.

CASE HISTORY

A 31 years, married woman with two children, presented to surgery OPD with history of difficulty in passing urine for last two months.The difficulty was progressively increasing in severity causing her to strain at micturition and the urine stream has become weak. There was no history of trauma to perineum, no urethral catheterisation or any instrumentation, no fever with chills or haematuria. Clinical examination, General and Systemic, revealed no abnormality. On per vaginal examination there was a soft, well circumscribed, globular mass situated between urethra and anterior wall of vagina.

Routine investigations were within normal limits. The differential diagnosis included: Aggressive angiomyxoma, Leiomyoma of the urethra, Lipofibroma, soft tissue tumour and dermoid cyst. Transvaginal ultrasound examination revealed a well defined, globular, 4 cm in diameter, hypoechoic mass situated between urethra and anterior wall of vagina.

After preparation patient was posted for surgical operation. Under Spinal anaesthesia, urethro-cystoscopy was done using 21-F sheath and 0 & 30 degree lens. Urethral mucosa and interior of bladder were normal. Urethral catheter was kept in situ to facilitate the surgery and to prevent trauma to urethra. The tumour was surgically excised and the periurethral tissues were closed in layers using 2-0 Vicryl.(fig.1,2) Specimen was subjected to histopathological evaluation. Post operative course was uneventful and patient was discharged on eighth day. Four years follow up revealed no recurrence and patient is asymptomatic.

Microscopically, the tumour was composed of moderate cellularity with scattered spindle and stellate shaped cells. The cytoplasm was ill-defined. The blood vessels were variable in size and the walls were thin and thick. Myxoid stroma noted (Fig.3,4) Some areas show increased cellularity. The cells had small round to oval hyperchromatic nuclei with small centrally located nucleoli.

DISCUSSION

The term aggressive angiomyxoma was coined by Steeper and Rosai in 1983 for a morphologically distinctive, slow-growing myxoid neoplasm that occurs chiefly in the genital, perineal and pelvic regions of adult women. Male to female ratio is 1:6.It is of two types: (1) superficial which grows near the surface and (2) aggressive which involves the deeper structures. Size ranges from a few centimeters to more than 20 cm Superficial angiomyxoma grows near the surface measuring 3-4 cm in size as a subcutaneous nodule and mostly involves the genitalia, trunk and head and neck. It affects mostly middle-aged adult females, i.e., between fourth to sixth decade. It is polypoid and is mostly associated with Carney's complex which is a triad of spotty pigmentation, cardiac myxomas, and endocrinal over activity.[1,2]

The standerd treatment for Aggressive angiomyxoma is complete wide excision with adequate margins and reconstruction whenever mandetory. Use of GnRH agonists may be of value in managing cases of aggressive angiomyxoma, either primary or recurrent, which are not amenable to surgical excision.[3]

Aggressive angiomyxoma is an uncommon soft tissue tumour which preferentially involves pelvic and vulvoperineal regions, typically characterised by gelatinous appearance and locally infiltrative nature without evidence of nuclear atypia or mitosis. It has a strong predilection for adult females in the third through sixth decades of life with a peak incidence in the fourth decade. Most of these lesions clinically simulate Bartholin's gland cyst. They have marked tendency for local recurrence (30-40%),but usually do not metastasise.[4,5] The tumour has also been described rarely in males with a median age at presentation in the sixth decade. [6]

Aggressive angiomyxomas display unusual growth pattern with high signal density in T2 weighted MRI.CT & MRI are useful in diagnosis and help in complete removal of tumour particularly arising from perineum, vulva and bladder. There is a case reported that required debulking of large Aggressive angiomyxoma of vagina followed by radiotherapy and then followed by definitive radical resection. [7]

A different clinical entity detected in vulvoperineal region is Smooth muscle tumors of undermined malignant potential (STUMP). The majority of these tumors are of uterine origin. They can recur in the form of STUMP or leiomyosarcoma. It is difficult to predict the clinical outcome for this patient, as the clinical courses of uterine STUMP are widely variable. In another retrospective review of 41 cases of uterine STUMP, there was an overall 7 percent recurrence rate in the form of both STUMP and leiomyosarcoma [8]

Neville D Perera et al reported leiomyoma of the urethra, a rare clinical entity. The patient was a 16-year old female who presented with a labial mass. The tumour (8 x 10 cm) was completely excised with reconstruction of the bladder and urethra. Histology confirmed cellular leiomyoma.[9].The most common site of Leiomyoma is the posterior wall of the urethra. The common clinical presentations include periurethral or vaginal mass, dysuria, dyspareunia, haematuria, and rarely obstructive urinary symptoms[10]

Lucio F. Gonzaga and others reported a rare case of a neoplasia mimicking a urethral tumor in a 32-year-old female patient. The extensive lesion was treated by anterior pelvic exanteration, vulvectomy, and bilateral inguinal lymphadenectomy and with autologous myocutaneuos flaps reconstruction and urinary diversion (sigmoid) and colostomy (Hartman's procedure). The pathological study established the diagnosis of aggressive vaginal angiomyxoma. The immunohistochemistry examination showed tumor positivity for CD-34 appointed strongly for angiomyxoma.[11]

CONCLUSION

A general surgeon may encounter unexpected, relatively rare disease in his clinical practice. Clinical scrutiny, a high index of suspicion for operative and postoperative complications and a common sense approach may improve the final outcome. It is important to be prepared to identify them and apply the appropriate treatment. Aggressive angiomyxoma requires individualized attention to decide how to manage and careful planning for long term follow up.

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Fig 01 : Bulging seen between urethra and anterior wall of vagina.

Fig 02 : Intra operative findings.

Fig 03 : Microscopic findings (H&E 10x)

Fig 04 : Microscopic findings (H&E 40x)

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