CASE REPORT

Aggressive Angiomyxoma of Inguinoscrotal Region Mimicking Inguinal Hernia: a Case Report

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Abstract Aggressive angiomyxoma (AAM) is a rare mesenchymal tumour of pelvis and perineum, almost exclusively occurring in females. We are reporting a case of a 53-year-old gentleman who presented with a long standing inguinoscrotal swelling misdiagnosed as inguinal hernia, for which he underwent incomplete excision at local hospital and then was referred to our centre for completion surgery.

Keywords Aggressive angiomyxoma · Inguinoscrotal swelling · Myxoid tumours · Aggressive angiomyxoma in males

Introduction

First described by Steeper and Rosai in 1983 [1], aggressive angiomyxoma is a rare mesenchymal neoplasm arising primarily from soft tissue of pelvis and perineum of adults. There is a strong female preponderance with reported female to male ratio of 6.1:1 in a

series of 100 cases [2]. Usual site of occurrence in male is inguinoscrotal region and perineum. The term aggressive emphasizes the often infiltrative nature of the tumour and high local recurrence rate. Because of its rarity, it may be misdiagnosed as hydrocele, hernia, or inguinal node leading to incomplete excision and hence high risk of local recurrence.

Case Report

A 53-year-old patient presented with a slowly growing painless inguinal swelling of 8-year duration with rapid progression for 2 months, for which he underwent excision through inguinal incision at local hospital. Histopathological examination revealed aggressive angiomyxoma with involvement of resected margins and patient was referred to our centre for further evaluation and management. Physical examination revealed a non-tender, non-reducible, firm, scrotal swelling just above the right testis with no other swelling palpable in bilateral inguinoscrotal region. Ultrasonography demonstrated a heterogeneously hypoechoic area above the right testis with suspicion of inguinal hernia, both testes were normal in size and appearance. CT scan showed no evidence of disease elsewhere (Fig. 1a). On exploration of inguinoscrotal region, a 4 × 3 cm solid swelling was found in the right hemiscrotum. The tumour was discrete and without adhesions to the nearby tissues. It was excised with a layer of enveloping normal looking fascia (Fig. 1b). Right testis and cord structures were found to be normal and hence preserved. Histopathology

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Fig. 1 **a** CECT: hypodense lesion present in inguinoscrotal region alongside spermatic cord. **b** Surgical specimen: 4 × 3 cm well-encapsulated tumour mass excised along with overlying scar of previous surgery

showed scattered large spindle cells with well-developed myxoid features and stroma showing myxoid change with plenty of vessels of varying calibre, suggestive of aggressive angiomyxoma (Fig. 2).

Discussion

Aggressive angiomyxoma is a rare neoplasm with around 350 cases reported so far, and only 14 cases reported in males [3]. The majority of patients present with a slow-growing asymptomatic mass, sometimes with accompanying symptoms like regional pain, a feeling of local pressure or dyspareunia. It is often mistaken for more common clinical entities like Bartholin cyst, vaginal cyst, abscess, leiomyoma, lipoma, fibroepithelial polyp, inguinal or perineal hernia. Though this entity is more common in females, it should be considered as differential diagnosis of tumours in the analogous regions of men like scrotum, spermatic cord and perineum. Though distant metastasis is very rare, they have high propensity to occur locally with series reporting between 25 and 47% [2, 4]. High local recurrence rate is attributable to locally infiltrative nature to adjacent soft tissue. Radiographically, on computed tomography, AAM may appear as well-defined homogenous mass which is hypodense relative to muscle or predominantly cystic with solid component. It is isointense or has low signal intensity on T1-weighted and has a whorled pattern of high signal intensity on T2-weighted MRI. These imaging findings are thought to be due to abundant myxoid matrix and high water content of the tumour [5]. Grossly, tumour size has been reported to vary from 1 to 60 cm. Microscopically, these are

hypocellular and highly vascular tumours with myxoid stroma containing cytologically bland stellate and spindle cells. The hallmark of AAM is vessels of varying calibre haphazardly scattered throughout the tumour parenchyma. There is no specific immunohistochemical marker but these tumours generally are positive for desmin, smooth muscle actin, muscle-specific actin, vimentin, oestrogen receptor and progesterone receptor. Some tumours are positive for CD34, whereas S100 is invariably negative [3, 4, 6].

Surgical excision with wide margins forms the first line of management. However, wider margins may not always be possible because of infiltrative nature of tumour and often proximity to pelvic organs like bladder and rectum. A review of 111 cases of AAM showed no significant difference in local recurrence between groups of patients with positive or resected negative margin (40 and 50% in 10 years, respectively) [1]. There is no well-defined role of chemotherapy or radiation therapy in management of AAM. A newer approach is use of hormonal therapy as most of these tumours occur in premenopausal females and are positive for oestrogen and progesterone receptors. Successful treatment of AAM with gonadotropin releasing hormone agonist has been reported in several case reports [7]. However, surgery remains the standard of care with GnRH agonist as alternative therapy in primary or adjuvant setting.

In summary, aggressive angiomyxoma though locally recurrent has otherwise good prognosis. These tumours in men often mimic benign conditions like hernia or hydrocele, leading to misdiagnosis and incomplete excision with positive or close margin. Thus, diagnosing AAM preoperatively or intraoperatively helps in achieving excision with wider margins.

Fig. 2 (H&E-200X) **a** Thick and thin walled vessels of varying calibre haphazardly scattered with adjacent spindle and stellate shape cells in myxoid stroma. **b** (H&E-200X) Spindly and stellate cells seen infiltrating into fat showing myxoid stroma

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

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