Aggressive Angiomyxoma of Scrotum Presenting as Scrotal Lymphedema in a Case of Postoperative Carcinoma Penis

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Abstract Aggressive angiomyxoma tumor is a rare tumor in males. We report a case of incidentally diagnosed aggressive angiomyxoma of scrotum presenting as scrotal lymphedema in a patient who underwent partial penectomy and radiotherapy for carcinoma penis.

Keywords Aggressive angiomyxoma · Locally infiltrative · High recurrence · Good prognosis

Introduction

Aggressive angiomyxoma is a locally infiltrative mesenchymal tumor which is common in the perineal region in young females and is rare in males. Sites frequently involved in males are the scrotum, spermatic cord, inguinal region, and perineum [1].

Case Report

A 62-year-old gentleman presented with a painless scrotal swelling of 6 months duration that gradually progressed in size (Fig. 1). He underwent partial penectomy, inguinal lymphnode dissection, and radiotherapy for carcinoma penis 18 months back and was in regular follow-up. No history of trauma, fever, weight loss, and abdominal lump was elicited. Clinically, a 15×12 cm-sized soft, non fluctuant, non-transilluminant, and non-tender, irreducible swelling

conforming to a scrotal lymphedema was seen and no inguinal lymphadenopathy was found.

USG scrotum was done which suggested mild hydrocoele in both scrotal sacs with diffuse subcutaneous thickening in the entire scrotal wall, both testis and epididymis were normal. Microfilarial test was negative. All other laboratory investigations were within normal limits. At this point, the swelling was thought to be post radiotherapy lymphedema. Subtotal excision of scrotum with wide margins was done to relieve the patient of discomfort from the swelling. Scrotum was reconstructed. Specimen was sent for biopsy.

Surface was smooth and had a gelatinous cut surface. Grossly, it was encapsulated with a pleura-like membrane. Histopathological examination revealed spindle and stellate-shaped cells proliferating around small- to medium-sized vessels within a fibromyxoid matrix. These cells had eosinophilic cytoplasm and lacked significant nuclear pleomorphism and mitosis (Fig. 2).

Based on these histological features, a diagnosis of aggressive angiomyxoma was made. Post operative period was uneventful; patient is disease free for 3 months and is on regular follow-up.

Discussion

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983. It is a locally infiltrative mesenchymal tumor which is common in the perineal region in young females and is rare in males [1]. The adjective “aggressive” emphasizes the neoplastic character of the blood vessels, its locally infiltrative nature, and the high risk of local recurrence, not indicating a malignant potential of the lesion [2]. In men, aggressive angiomyxoma is usually derived from the pelviperineal interstitial tissue involving the scrotum (38 %), spermatic cord

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Fig. 1 Clinical photograph of scrotal swelling

(33 %), perineal region (13 %), and intrapelvic organs such as the bladder (8 %) [3].

Grossly, these tumors are soft, partly circumscribed, polypoidal lesions with gelatinous appearance on cut section. Microscopically, these lesions are composed of many thick-walled vessels of varying sizes in a loose myxoid and collagenous stroma with spindle and stellate-shaped neoplastic cells.

Immunohistochemically, these tumors express ER and PR, thus suggesting that they may be hormone dependent [4]. IHC was not done in this case. Chromosomal translocation of the

Fig. 2 Histopathology image showing aggressive angiomyxoma comprising of benign spindle cells [black arrow head] amidst myxoid change interspersed by numerous rounded thick- and thin-walled vessels

12q13-15 band involving the HMGA2 gene has been described [5].

On CT scan, these tumors have a well-defined margin with attenuation less than that of the muscle. On MRI, these tumors show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma [6]. In this particular case as it was a retrospective diagnosis, CT and MRI were not done preoperatively.

Surgical excision is the treatment of choice, although treatment with gonadotropin-releasing hormone agonists (Leuprolide) is an emerging therapy [7].

The tumor is usually locally infiltrative and has a high rate of local recurrence after surgical excision [1]. The excision of these tumors is difficult as they have the same consistency as that of normal connective tissue and therefore have a propensity for local recurrence (36–72 %) Hence, long-term follow-up is necessary and magnetic resonance imaging is the preferred method for detecting recurrences [8]. Metastases are exceedingly rare, and overall, the prognosis is good [5].

Conflict of Interest Dr. K. Siva Sankara Rao, Dr. K. Appa Rao, and Dr. S. Praveen Declare that they have no conflict of interest.

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