DOI: 10.7860/JCDR/2023/62130.17959

Aggressive Angiomyxoma: An Unusual Scrotal Mass

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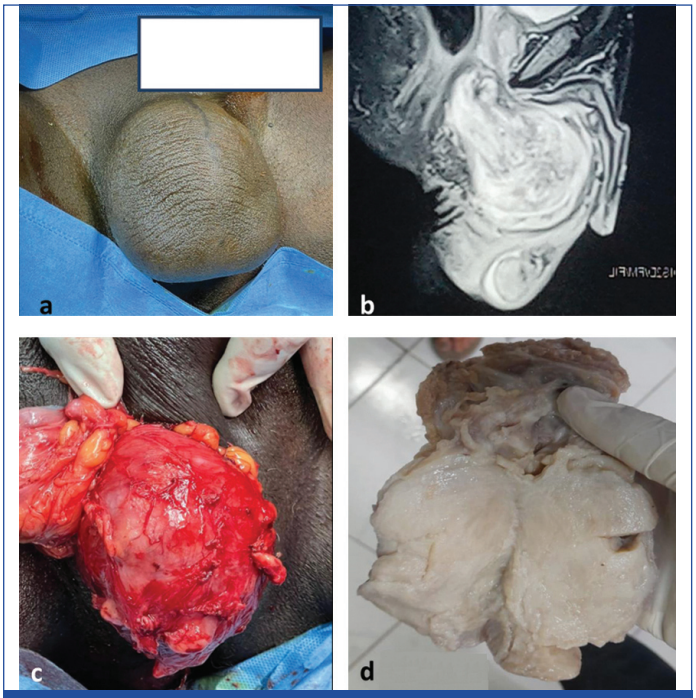
Keywords: Benign, Mesenchymal tumour, Perineum, Spindle cell

Scrotal masses are a frequent complaint in general practice. The treating doctor must distinguish a benign from a malignant scrotal lump to perform precise surgery and provide ongoing care.

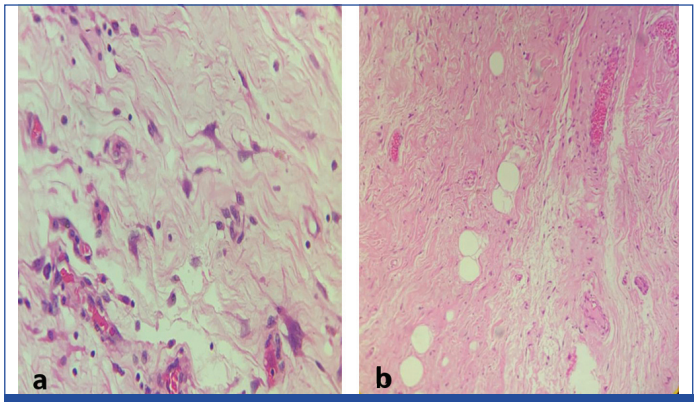
A 61-year-old male presented with a painless scrotal mass for six months. On clinical examination, the scrotal mass was firm and fixed to the perineal membrane [Table/Fig-1a]. Magnetic Resonance Imaging (MRI) scan revealed a 13×8×5 cm heterogeneous soft tissue mass [Table/Fig-1b] in the peranal/perineum lesion abutting the anterior wall of the anal canal, root of the penis, and scrotum. The possibility of myxoid/spindle cell neoplasm of the perineum was mentioned, and the remark of a locally aggressive neoplasm, likely of malignancy, was made in view of presence of infiltrating margins. Trucut biopsy was reported as a benign soft tissue tumour. Trans-scrotal excision was performed, and the postoperative resected specimen was sent for histopathological examination [Table/Fig-1c].

Gross appearance of the external surface was grey white to grey brown. Cut surface was homogenous grey white soft in consistency with solid region at one pole measuring 7×5×0.5 cm [Table/Fig-1d]. No areas of necrosis or haemorrhage noted. Tissue bits from representative sites were given. The microscopic examination revealed an unencapsulated infiltrative hypocellular lesion composed of spindle to stellate shaped tumour cells with delicate cytoplasmic processes and bland nuclear chromatin. No atypia or increase in mitosis was noted. The tumour cells were seen to be embedded in a myxoid stroma with scattered delicate collagen fibers [Table/Fig-2a].



[Table/Fig-1]: a) Clinical picture of scrotal mass; b) MRI scan revealed a 13×8×5 cm heterogeneous soft tissue mass in the perineum extending into the scrotum; c) Intraoperative picture of scrotal mass that was firm and fixed to perineal membrane. d) Macroscopic appearance shows grey white homogenous mass with ill-defined margin and gelatinous cut-surface.

Entrapment of mature adipose tissue was noted at foci [Table/Fig-2b]. Numerous small and medium sized thick walled hyalinised blood vessels were also noted and it was reported as Aggressive Angiomyxoma (AAM). The patient did not have recurrence at three and six month postoperative follow-up period.



[Table/Fig-2]: Microscopic appearance: a) Hypocellular tumour composed of spindle to stellate shaped cells, with delicate cytoplasmic processes, bland nuclei are seen lying within myxoid stroma along with few dilated capillaries (Haematoxylin and Eosin 40x); b) At the periphery of the lesion, entrapment of adipocytes and distinct myiod bundles wrapping around vessel was noted (Haematoxylin and Eosin 10x).

Scrotal masses account for 1% of cases in the surgical Out Patient Department (OPD). Although some types of scrotal masses are more prevalent at particular ages, they can appear at any age [1]. AAM is a very rare benign spindle cell mesenchymal tumour that usually occurs in the pelviperineal region, more commonly in females than males (M:F is 1:7) in their fourth decade [2]. It was first described in 1983, by Steeper and Rosai. So far only 36 cases of scrotal AAM are reported in the literature. AAM usually present as a painless mass. About 30% of these tumours recur locally, as it shows infiltrative nature hence the term 'Aggressive', although distant metastasis is extremely uncommon, MRI aids in determining the tumour's extent [3].

Its aetiology is not clearly established yet. Few studies hypothesised that AAM might develop from stem cells with the capacity for multiple differentiation near arteries that eventually give rise to fibroblasts and myofibroblasts [4,5].

Most AAM patients noted to have a long history of asymptomatic mass or swelling in the scrotum. The tumour size range from 1.5 cm to as big as 25 cm. The genital location and subtle clinical presentation of the tumour delay the treatment requirement by patients, which leads to a progressively growing tumour size with a high rate of clinical misdiagnosis. It is challenging to get an accurate preoperative diagnosis, and the differential diagnosis includes non neoplastic and neoplastic lesions [5].

On macroscopic examination, AAM is typically a soft, lobulated, grayish-white, fairly delineated tumour with vague infiltrative margins and myxoid appearance. The essential diagnostic criteria of AAM include the histological features of ill marginated lesion with uniformly dispersed tumour cells in abundant myxoid stroma,

vascular components and invasive proliferation. The bland tumour cells may be spindled, star or oval-shaped with bland nuclei and sparse mitoses. The vessels may vary from thick hyalinised or thin-walled walls of different diameters. The aberrant HMGA2 expression is considered as desirable for diagnosis however is not essential, as its sensitive but not specific marker for AAM. It is useful especially to define tumour margin, as the non tumourous mesenchyme lacks positivity to HMGA2 [6].

AAM in the scrotum is typically treated with a large local excision with tumour-free surgical margins [7]. When surgical complications are common or fertility retention becomes an issue, some specialists think incomplete or partial resection is permissible. The likelihood of local AAM recurrence is significant and differs by gender, particularly within 2-3 years.

Myxoid liposarcoma, angiomyofibroblastoma, myxoid neurofibroma, and myxoma are the tumours in the main differential diagnosis of AAM. Malignant myxoid neoplasms should be recognised from AAM since the management protocol varies for the both. Although it is not usually necessary, immunohistochemistry aids in distinguishing AAM from other mesenchymal tumours. Desmin, Vimentin, Smooth Muscle Actin (SMA), and CD34 are all positive in AAM, while S-100, CK, and CD68 are all negative [6].

Due to its infiltrative boundaries, AAM might radiographically evoke suspicion of malignancy, as in the present case. Owing to the rarity of this lesion and lack of distinct imaging findings, an accurate preoperative diagnosis is frequently challenging. From the experience with above described case and literature reviews, it was observed that the preoperative imaging techniques and intraoperative pathology investigations can partly aid with the diagnosis. However, the gold standard for the diagnosis of AAM relies on the thorough histological evaluation of postoperative specimens. Despite being an uncommon phenomenon in the male genital region, the surgeon should consider it as one of their differential diagnosis to prevent over diagnosing malignancy and subsequent treatment.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Dec 13, 2022
- Manual Googling: Apr 13, 2023
- iThenticate Software: Apr 20, 2023 (9%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Dec 08, 2022**
Date of Peer Review: **Jan 16, 2023**
Date of Acceptance: **Mar 23, 2023**
Date of Publishing: **May 01, 2023**