

# An Unusual Case of Scrotal Aggressive Angiomyxoma (AAM) Masquerading as Non-Reducible Inguinal Hernia – A Rare Case Report and Literature Review

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## Abstract

## Case Report

This case report details a rare instance of scrotal Aggressive Angiomyxoma (AAM) in a 62-year-old male, presenting with a gradually enlarging mass masquerading as a non-reducible inguinal hernia. AAM, predominantly found in adult females, is an exceptionally uncommon occurrence in men. The diagnostic challenge lay in the rarity of male cases and the tumor's mimicry of common conditions like inguinal hernia. Upon physical examination, a non-reducible, non-tender swelling extending from the right inguinal region to the scrotal sac was noted. Radiological investigations initially suggested a scrotal mass or non-reducible inguinal hernia. Intraoperative exploration revealed a large encapsulated cystic mass, measuring 12 x 8 cm, originating from the inguinal ligament. Histopathological examination confirmed the diagnosis of aggressive angiomyxoma, characterized by spindle-shaped tumor cells, thick-walled blood vessels, and myxoid texture. The discussion underscores the diagnostic complexities of AAM, emphasizing the necessity of histopathological examination for conclusive diagnosis. A review of male AAM cases in the literature highlights similarities with female cases, supporting the expression of estrogen (ER) and progesterone receptors (PR). Surgical removal with clear margins is the primary treatment, although recurrence rates vary. The potential role of hormonal therapy, especially in ER and PR positive cases, is noted but requires further exploration. In conclusion, this rare case sheds light on the clinical and pathological characteristics of scrotal AAM in men. The diagnostic journey underscores the importance of considering AAM in the differential diagnosis of scrotal masses. Long-term follow-up is crucial due to the tumor's propensity for local recurrence, making this case a valuable addition to the limited literature on male AAM.

**Keywords:** Aggressive Angiomyxoma, Myxoid Tumor, Scrotal Mass, Non-reducible Inguinal Hernia, Mesenchymal Tumor, Locally invasive Scrotal Mass, Inguinal Mass, Estrogen receptor Tumor, Progesterone receptor Tumor.

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## INTRODUCTION

Aggressive Angiomyxoma is a rare mesenchymal benign my old tumour of the pelvis and perineum which occurs almost exclusively in adult females and the occurrence of AAM in men is extremely rare. However in men, AAM is usually derived from the pelviperineal interstitial tissue involving the scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic organs (8%). AAM in the scrotal region may present as a scrotal mass, often mistaken for a hernia or hydrocele which was a diagnostic challenge in this case as well. Detailed radiological workup perhaps would have been helpful in detection, but histology of

excisional biopsy sample is the gold standard for establishing the diagnosis [1].

## CASE REPORT

A 62-year-old male was admitted to our General Surgery Department with a history of a gradually enlarging mass in the Right inguinal region extending into the scrotum for 5 years. On Physical examination a soft, non reducible, non-tender, swelling of size 12x6cm as seen in Image 1, extending from right inguinal region to the right scrotal sac. It was not possible to get about the swelling and B/L testis were free from the swelling and palpable. The mass seemed to originate from the ligament/ cord due to which non reduce-able left inguinal hernia was one of the initial differential

diagnosis. However cough impulse and other relevant examinations remained negative and posed a question in our minds and further assistance was taken from radiological investigations. No other similar swelling was found elsewhere in the body. All other systems appear within normal limits. [1, 2]. On USG imaging, an echogenic lesion seen in scrotum separate from the testis with definitive communication with abdomen as shown in image; Query indirect inguinal hernia or Lipoma and subsequently an inguinal approach inguinal and scrotal exploration was performed keeping in mind the clinical examination as well as Imaging modalities with Scrotal mass/ or non-reducible Inguinal Hernia as the preoperative diagnosis [3, 4]. A large encapsulated cystic mass possibly 12 x 8 cm large was then excised as shown in (Figure 2) and dissected up to the inguinal ligament (Figure 3) which was perhaps the originating location.

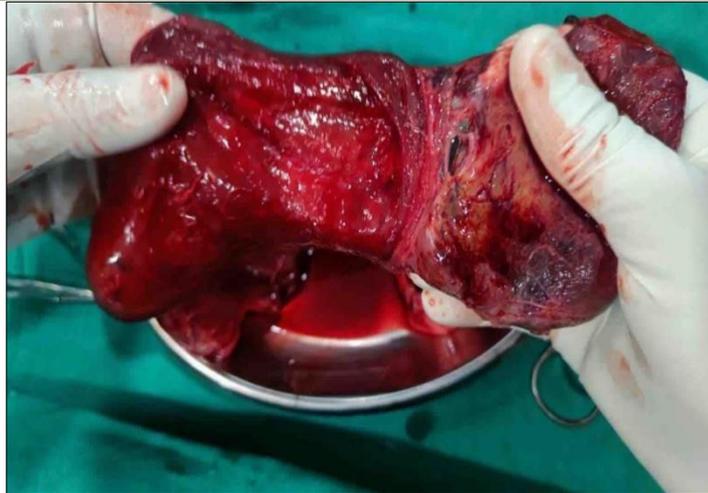
As seen in Figure 4, the tumour was grossly dissected so as to find it composed of myxoid texture and then histopathological examination of the specimen revealed features of intramuscular myxoma. Showing a sparse population of spindle-shaped Tumor cells without cytological atypia or mitosis. Foci of thick-walled blood vessels of various sizes were identified and surrounded by edematous stromal tissue. At the periphery of the tumor, residual skeletal muscle was focally presented. The tumor cells were positive for CD34 (most probably myofibroblast cells Figure 5). Focal staining was present for Alpha-SMA and Desmin (supporting myoid origin of these cells). The morphological picture and the special stains were compatible with aggressive angiomyxoma of the scrotum and peritoneum. Although a relatively rare diagnosis, myxoma should be considered as a differential diagnosis [2-4].



**Figure 1**



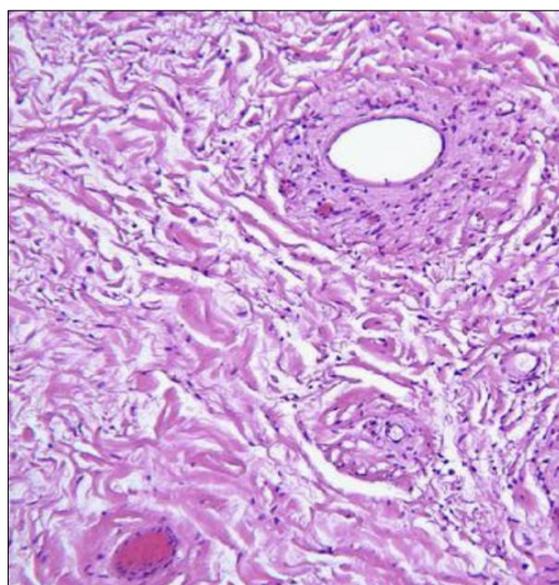
**Figure 2: Large encapsulated cystic mass/tumor excised**



**Figure 3: Intra operative Images**



**Figure 4: Gross dissection of excised mass showing myxoid nature of swelling**



**Figure 5: Histologic slide showing thick-walled vessels, no mitosis and collagen fibrils (H&E)**

## DISCUSSION

AAM was first described as a separate histopathological entity by Steeper and Rosai in 1983. Considering its benign nature, the term “aggressive” was modified to “deep” in the fourth edition of the World Health Organization Classification of Soft Tissue Tumors in 2013. In men, only case reports or case series have been reported in literature. Most of these patients were usually asymptomatic, whereas a small number of patients presented with inguinal hernia and testicular tumors. In the present case, the tumor developed in the scrotum and presented as a gradually increasing mass with no typical clinical symptoms. Most tumors in the reviewed cases were >10 cm in size as AAMs are not easily detected early. Most tumors are ill defined, making complete resection difficult and resulting in frequent local recurrence. However, few tumors demonstrate partial or complete encapsulation [5, 6].

Misdiagnosis in AAM is common because it can mimic other diseases, including hydrocele, inguinal hernia, or paratesticular neoplasia. Preoperative diagnosis is often difficult and challenging because of the rarity of these tumors and lack of specific imaging features. AAM is diagnosed based on the histopathological examination of postoperative specimens. The histopathological features of males with AAM reported in literature are similar to those reported in classical female cases. Microscopically, AAM comprises small-sized spindle cells or stellate cells embedded in a loose myxoid matrix with abundant collagen fibers and variably sized vessels. Blood vessels ranging from capillary-like to thick-walled vessels, which are the most prominent feature of AAM, can be observed. In the present case, no evidence of atypical mitotic activity or nuclear atypia was noted. Immunohistochemical staining plays a crucial role in the diagnosis of AAM, although there is no specific immunohistochemical marker of AAM. The neoplastic cells of AAM are generally positive for desmin, vimentin, SMA, CD34, ER, and PR but negative for S-100 and CK in female patients [6].

AAM should be distinguished from angiomyofibroblastoma, myxoid liposarcoma, myxoma, superficial angiomyxoma, and myxoid neurofibroma. Angiomyofibroblastoma is a benign tumor that has recently been described as histologically similar to AAM, with myofibroblastic cells clustered in abundant myxoid stroma. This tumor contains several areas of hypo- and hypercellular cells, often clustered around blood vessels. Myxoid liposarcoma must be considered as a differential diagnosis when tumor cells infiltrate adipose tissues. It can be easily distinguished from AAM as myxoid liposarcoma is marked with adipocytes set in abundant thin-walled vessels.

The surgical removal of AAM with clear margins is the traditional treatment to prevent local

recurrence. However, it is not clear whether the recurrence rate is associated with the surgical margin status. In women, the recurrence rates of 71%, 85%, and 94% have been observed within the first 3, 5, and 7 years of local excision, respectively possible reasons for the lower local recurrence rate in male patients are sample limitations or lower hormone expression. It is generally known that AAM has no metastatic tendency; Moreover, owing to the low proliferative activity of AAM, the role of radiotherapy and chemotherapy is unclear and limited. In recent years, hormonal therapy has been considered an adjunctive treatment for ER and/or PR positive female patients with primary large mass or local relapse that is not amenable to surgery [6]. Unfortunately, it is not clear whether the relapse rate is higher when hormone therapy is discontinued. In the present case, only local resection was performed, and although the patient was both ER and PR positive, hormonal therapy was not advised as few data are available on hormonal therapy in male patients with AAM. Long-term follow-up surveillance is required because of the aggression and relapse characteristics of this tumor. At present, the patient has been followed-up for 6 months without any recurrence or metastasis [7, 8].

## CONCLUSION

In summary, a rare case of scrotal AAM was reported and previously reported male cases with AAM were summarized. In particular, the review revealed that the clinicopathological characteristics of AAM in men are similar to those of AAM in women, including the expression of ER and PR [6]. This provides an opportunity to treat such male patients with hormone therapy. Moreover, the literature review revealed a low recurrence rate (4.7%) in males after the surgical excision of the tumor; however, more data are needed to confirm this observation. Finally, AAM should be distinguished from myxoid neoplasms in male genital areas. Differential diagnosis of this tumor includes angiomyofibroblastoma, cellular angiofibroma, cutaneous myxoma, myxoid neurofibroma, intramuscular myxoma, and myxoid lipoma [8].

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