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Aggressive Angiomyxoma Of Scrotun-A Rare Entity

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Abstract: ***Aggressive Angiomyxoma is a rare mesenchymal benign myxoid tumor of the pelvis and perineum which occurs almost exclusively in adult females. It rarely occurs in males and involves scrotum. A 62years old male patient presented with a slowly growing scrotal swelling since 6 months.Ultra sound examination revealed diffuse sub cutaneous scrotal wall thickening with hydrocele. Excision of the scrotal mass was done and sent for histopathological examination. Gross examination revealed skin covered soft tissue mass. On cut section there was partially circumscribed, lobulated mass with myxoid areas. Microscopic examination revealed lobulated lesion containing variable sized blood vessels, hypocellular and myxoid areas. About 150 cases were reported worldwide (few cases were reported in males) Aggressive Angiomyxoma in the scrotal region may present as a scrotal mass, often mistaken for a hernia or hydrocele.***

KEY WORDS: AGGRESSIVE ANGIOMYXOMA, SCROTAL MASS, RARE ENTITY.

I. INTRODUCTION

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983.[1] Aggressive Angiomyxoma (AAM) is a rare benign mesenchymal myxoid tumor of the pelvis and perineum which occurs almost exclusively in adult females. Overall, its incidence is about 6-folds higher in females.[2] It usually arises from the soft tissues of the pelvic region, perineum, vulva and buttock. The adjective "aggressive" emphasizes the neoplastic character of the blood vessels, its locally infiltrative nature and the high risk of local recurrence after surgical excision.[3] It is called angiomyxoma because of its myxoid nature and prominent vascularity.[4] Rarely, this tumor appears in males simulating inguinal hernia, testicular neoplasm, spermatic cord neoplasm, hydrocele or spermatocele.[2]

II. CASE REPORT

A 62 years old patient presented with a slowly growing scrotal swelling since 6 months. Clinical examination revealed diffuse scrotal swelling measuring about 13×10cm. Hernial site examination revealed no abnormalities. Ultrasound examination report was diffuse subcutaneous scrotal wall thickening with hydrocele. Testis and epididymis on both sides showed no abnormalities. A Subtotal excision of

the scrotal swelling was done and sent for histopathological examination.

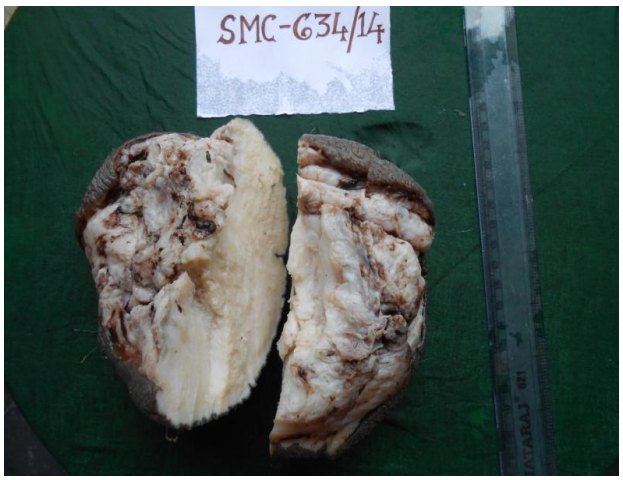
A. Gross Appearance

received skin covered soft tissue mass measuring 15x 12 x 8 cm.(fig.1).

Fig.1:

Cut section shows partially circumscribed, lobulated mass with infiltrating margins. The mass is gray white with slimy areas (myxoid areas). No testes were identified.(fig.2)

Fig.2:



B. Microscopic appearance

Sections studied show ill defined lobules made up of predominantly hypocellular myxoid areas with scattered spindle to stellate cells and bundles of smooth muscle cells. There are numerous blood vessels of varying caliber. Focal areas show perivascular mononuclear inflammatory infiltrate.(Fig.3)

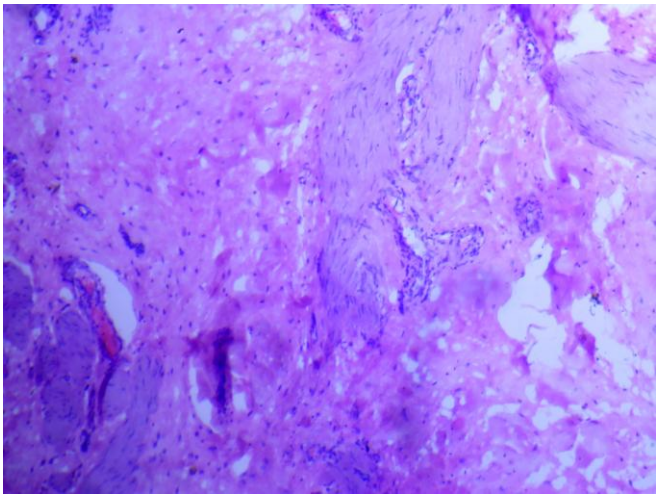


Figure 3: Microscopic aspect of the tumour (200X H&E)

III. Discussion:

Aggressive angiomyxoma was first described by Steeper and Rosai in 1983. Around 150 cases have been described in literature since then.[4] It is a morphologically distinctive, slowly growing vulvo-vaginal mesenchymal neoplasm that occurs chiefly in adult women with a marked tendency for local recurrence. It usually does not metastasize but few cases have been reported in literature where metastasis have occurred.[5,6] Due to its locally infiltrative course and frequent recurrences it has been termed aggressive.

Occurrence of aggressive angiomyxoma in men is extremely rare and in men, aggressive angiomyxoma is usually derived from the pelviperineal interstitial tissue involving the scrotum (38%), spermatic cord (33%), perineal region (13%), and intrapelvic organs (8%).[3] Aggressive angiomyxoma in the scrotal region may present as a scrotal mass, often mistaken for a hernia or hydrocele.[4] Detailed radiological work-up may be helpful in detection, but histology is the gold standard for establishing the diagnosis.[4]

In both males and females, the gross and microscopic appearances and clinical course are similar. On gross examination the tumors are lobulated, soft to rubbery, solid masses. The cut surface reveals a glistening, soft homogeneous appearance. Recurrent tumors show more prominent areas of hemorrhage and fibrosis.[7] Microscopically, the tumor is composed of widely scattered spindled to stellate-shaped cells with ill-

defined cytoplasm and variably sized, thin- or thick-walled vascular channels in a myxoid stroma that is rich in collagen fibers. Cellularity is usually low but focal areas show increased cellularity, particularly around large vessels. Mitotic figures are rare or absent. The stroma is characterized by prominent myxoid change with fine collagen fibrils, often with areas of erythrocyte extravasation. A characteristic feature is the presence of variably sized vessels that range from small thin-walled capillaries to large vessels with secondary changes including perivascular hyalinization and medial hypertrophy.[8] Diffrential diagnosis of this tumor includes myxoid neurofibroma, intramuscular myxoma, myxoid lipoma and angiomyofibroblastoma.[9] Tumour cells are immunoreactive for desmin, muscle specific actin and vimentin. Estrogen and progesterone receptor may be positive. The tumour cells are immunonegative for S100 protien, factor VIII related antigen, carcino embryonic antigen and cyto keratin.[2]

IV. CONCLUSION

Aggressive angiomyxoma being a rare lesion and it must be considered in the differential diagnosis of scrotal swelling. careful pre operative workup should be carried out in suspicious cases. Rejection with clear margins is necessary to prevent recurrence.

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