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DEEP AGGRESSIVE ANGIOMYXOMA OF INGUINAL REGION IN A MALE- A RARE CASE REPORT

Histopathology

Dr. Dharmashri Kadam Consultant, Department of Surgery, V. N. Desai Hospital, Santacruz (East), Mumbai, Maharashtra, India.

Dr. Abhijit Bhanji Consultant, Head of Department, Department of Pathology, V. N. Desai Hospital, Santacruz (East), Mumbai, Maharashtra, India.

Dr. Renuka Kulkarni* Senior Resident, Department of Pathology, V. N. Desai Hospital, Santacruz (East), Mumbai, Maharashtra, India. *Corresponding Author

Dr. Sattwik Bhalerao Junior Resident, Department of Surgery, V. N. Desai Hospital, Santacruz (East), Mumbai, Maharashtra, India.

ABSTRACT

Deep Aggressive Angiomyxoma is a rare mesenchymal tumour demonstrating local infiltration, but rarely metastasises. It is observed usually in females in the pelvic or perineal region however, few cases have been reported in men. We present a case of a 69 years old man with swelling in the inguinal region since 4 years, gradually increasing in size. Differential diagnosis of teratoma, lipoma and sebaceous cysts were made clinically. He was operated for wide local resection. Gross examination of tumour revealed a soft to firm bosselated encapsulated mass measuring 10.5x8.9x7.5cm. Microscopy findings showed hyper and hypocellular areas composed of spindle cells, varying sized hyalinised vessels with myxoid and collagenous areas; characteristic of angiomyxoma. There was sparse mitoses and was devoid of atypia or necrosis. Immunohistochemistry confirmed the diagnosis with expression of SMA, ER and PR. Patient was healthy after a year follow-up. Although rare in the male population, aggressive angiomyxoma should be considered as a critical differential diagnosis in patients presenting with pelvic or perineal swellings. These entities must be looked for by clinicians even in males in uncommon locations so as to distinguish them from benign tumours with low recurrence and malignant myxoid tumours with high rates of metastasis and provide appropriate management.

KEYWORDS

Deep aggressive angiomyxoma, Pelvic tumours, mesenchymal tumours, locally infiltrating tumours, mesenchymal tumours

INTRODUCTION:

Deep aggressive angiomyxoma is a locally infiltrative, non-metastasising rare mesenchymal tumour of unknown histogenesis. It is commonly seen in women of reproductive age group, but has also been reported in men.[1] Differentiating from other mesenchymal tumours of pelvic/perineal region is necessary as it has high local recurrence rate and good prognosis. These lesions are slow-growing, soft, un-encapsulated with finger like projections in the surrounding structures and with or without pressure symptoms.We report a case of Deep aggressive angiomyxoma of the inguinal region in a male patient.

Case Report:

A 69 year-old man presented with swelling in left inguinal region since 4 years which gradually increased in size. There was no history of trauma, previous surgery or any other co-morbidities. Clinically, differential diagnosis of teratoma, lipoma or sebaceous cyst were made. CECT Pelvis showed a well-defined mass measuring 12x8cm in left inguinal region in subcutaneous plane with foci of fat seen within, suggestive of Teratoma. (Fig. 1). Patient was operated for wide local resection. Intra-operatively, a 12x10 cm subcutaneous mass with a large feeder vessel at base was noted.

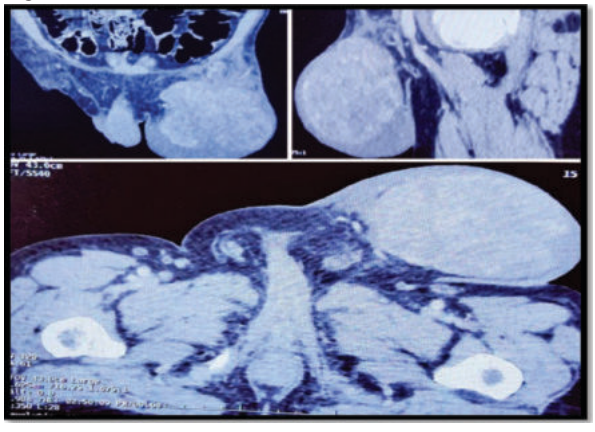

Fig. 1: CECT showing A well-defined mass in left inguinal region in subcutaneous plane with foci of fat within. **a)** Coronal view, **b)** Sagittal view, **c)** Axial view

A Skin bearing tissue piece measuring 14x12x9cm was received. Externally, it was bosselated, soft to firm, measuring 10.5x8.9x7.5cm. Cut surface showed a well-circumscribed tumour, yellowish in colour with few congested areas. Microscopy revealed a tumour with thin fibrous pseudo-capsule which showed hyper and hypocellular areas composed of spindle cells exhibiting spindled blunt ended nuclei and indistinct eosinophilic cytoplasm. Varying sized non-anastomosing thin and thick-walled hyalinised vessels throughout the tumour parenchyma were seen with myxoid and collagenous stroma. Stroma showed an inflammatory infiltrate composed of plasma-cells, mast-cells and lymphocytes. Mitosis was sparse while necrosis or cellular atypia was not absent. (Fig. 2&3)

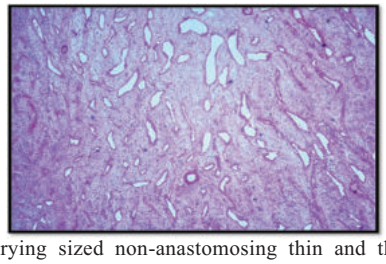

Fig. 2: Varying sized non-anastomosing thin and thick-walled hyalinised vessels throughout the tumour parenchyma. (H & E stained, 10x)

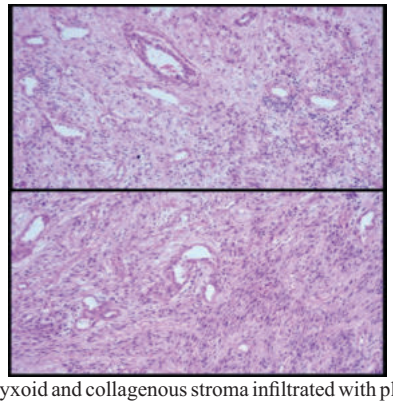

Fig. 3: a) Myxoid and collagenous stroma infiltrated with plasma cells,

mast cells and lymphocytes. (H & E stained, 40x) **b)** Hyper and hypocellular areas composed of spindle cells exhibiting spindled blunt ended nuclei and indistinct eosinophilic cytoplasm. (H & E stained, 40x)

A diagnosis of Aggressive Angiomyxoma (AAM) was made. Immunohistochemistry revealed that tumour expressed SMA, BCL2, CD31, ER and PR while negative for Desmin, S-100, CD34, H-Caldesmon and CDX. Mib1 was 1-2%. Thus, a final diagnosis of angiomyxoma was given. After a month, follow-up MRI revealed no residual disease. On one year regular follow up, the patient is healthy with no complaints.

DISCUSSION:

Deep AAM is a rare mesenchymal tumor arising from soft tissues of pelvis or perineum. It occurs mainly in reproductive age group usually in females. In 1983, based on clinical and pathological features of 9 patients, Steeper and Rosai named this tumor "aggressive angiomyxoma" for first time. (Steeper TA et al., 1983) [1]

AAM is a locally aggressive soft tissue tumor, which primarily occurs in pelvic–perineal regions. The term "deep" denotes its propensity for local aggression and recurrence after excision. Approximately 70% - 80 % of cases have recurrence. Local recurrences are treated with reoperation with wide local excision. (Kura MM et al., 2012) [2] The incidence in females is significantly higher than males & female-to-male ratio of 6:1 with few cases reported in men. (Chen H et al., 2017) [3] It is mostly seen in premenopausal women with a peak incidence in the third to fifth decades of life.

Upto 350 cases of AAM have been documented in the scientific literature till date with around 85 cases reported in male patients as reviewed by Chen et al in an extensive review of literature study (Chen Y et al., 2022). [4] Only 3 cases of aggressive angiomyxoma in the inguinal region in males have been studied in the literature as per our knowledge. The average age in men is 46 years and ranges from 1 to 82 years (Idrees MT et al., 2006) [5].

The main site of disease is perineum, followed by pelvic cavity and vagina. (Chen H et al., 2017) [3] The sites that are involved in males in limited case-reports till date are scrotum, spermatic cord, inguinal region, prostate and epididymis (Hsieh F et al., 2018) [6] Van Rogen et al in their case series of 11 cases presented with one case of 36 year old male with AAM in the inguinal region and Kondo in his case report of a 63 year old male patient presented with slow growing AAM in inguinal region. (Van Roggen et al., 2005) (Kondo T et al., 2010) [7,8] Preoperative diagnosis is difficult because of the rarity of these neoplasms and lack of specific features clinically and on imaging studies. Hence, a histopathological diagnosis along with immunohistochemistry is the gold-standard diagnostic modality. (Idrees MT et al., 2006) [5]

Clinically, AAM are mistaken for lipomas, Bartholin duct cysts, rectoceles, hernias or even malignant pelvic floor tumours. (Chen H et al., 2017) [3] Similarly, in our case clinical differential diagnosis as teratoma, sebaceous cyst and lipoma were made. Histological differential diagnosis range from benign myxoid tumours like myxolipoma, myxoid neurofibroma and angiomyofibroblastoma and from metastatic tumours like myxoid liposarcoma, myxoid malignant fibrous histiocytoma (MFH) and embryonal rhabdomyosarcoma. (Idrees MT et al., 2006) [5]

The characteristic microscopic appearance of vascular component with vessels of various calibers with a bland myxoid stroma and minimal mitotic figures helps in distinguishing majority of the above neoplasms. Myxoid neurofibroma is devoid of the prominent vascular component unlike AAM whereas myxoid liposarcoma shows presence of lipoblasts and straight vascular channels without thick-walled hyalinised blood vessels. Angiomyofibroblastoma shows characteristically well-circumscribed borders, plumper stromal cells with higher cellularity and is a desmin positive tumour while myxoid MFH shows small branching vascular channels and pleomorphic tumour cells scattered in between. Embryonal rhabdomyosarcoma has a higher cellularity, presence of rhabdomyoblasts with nuclear pleomorphism presenting exclusively in early years of life. The bland histology of AAM with prominent vascular features and minimal mitotic activity distinguishes itself from the rest tumours whereas immunohistochemistry ascertains the diagnosis in all cases.

Research study has shown that in women, tumor cells are characteristically positive for ER and PR, suggesting a hormonal role in development of tumor & therapeutic advantage using hormone agonists in tumours with difficult resection. Idres et al., in their case series of 4 cases studied that male AAM demonstrated ER and PR expression similar to their female counterparts (Idrees MT et al., 2006) [5]. Our present case showed ER, PR expression as well. Hormonal antagonists like tamoxifen and goserelin have been used widely in some cases, however they were not used in the treatment of this case. (Kondo T et al., 2010) [8]

Recently, a translocation on chromosome 12 with consequent aberrant expression of DNA factor HMGA2 (HMGIC protein) involved in DNA transcription has been found which may be useful in diagnosis of challenging cases and used as a sensitive novel marker for AAM diagnosis. (Chen H et al., 2017) [3]

Surgical local resection with clear margins is the mainstay treatment in these cases. Although complete surgical resection is ideal outcome, partial removal is allowed when operative morbidity is expected or when fertility preservation is concerned (Steeper TA et al., 1983) [1] Two cases of metastasis are reported in literature in 2 females while none reported in males so far. Metastasis had been reported in literature as follows: 1. Pulmonary and mediastinal metastasis with disseminated AAM who died of the disease 2. Metastasis to the lung from a vulvar AAM leading to death. (Idrees MT et al., 2006) [5]

A long-term follow-up with imaging studies is recommended owing to high recurrence rates. Recurrences has been common with 30 -70 % which poses as an unique feature of AAM. (Idrees MT et al., 2006) [5]

To conclude, clinicopathologic characteristics of AAM in men & women are comparable. Rarity of the tumour in males with typical histopathology and locally infiltrative nature, these distinct characteristics should be kept in mind. Differentiation from benign tumours with low recurrence risk and malignant myxoid tumours with capacity to metastasise is crucial.

Abbreviations:

AAM- Aggressive angiomyxoma
SMA- Smooth Muscle Actin
ER- Estrogen Receptor
PR- Progesterone Receptor
MFH- Malignant Fibrous Histiocytoma

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