Extragenital Aggressive Angiomyxoma of the Axilla and the Chest Wall

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ABSTRACT
Aggressive angiomyxomas are uncommon mesenchymal tumours which most often arise in the perineal and the pelvic regions in women. Extragenital aggressive angiomyxomas are extremely rare. We are reporting a young male with an aggressive angiomyxoma which involved the axillary region and extended into the anterior chest wall, which demonstrated its characteristic histomorphological features. The diagnosis was confirmed by immunohistochemistry. A careful histological examination, along with immunohistochemistry, aids in diagnosing this lesion and differentiating it from tumours which have similar histologies.

Key Words: Aggressive angiomyxoma, Extragenital, Chest wall, Axilla

INTRODUCTION
An aggressive angiomyxoma is a rare myxoid soft tissue neoplasm which originates from fibroblasts/myofibroblasts which are composed of variably sized blood vessels, which range from thin walled capillaries to thick hyalinized blood vessels with a predominant myxoid stroma. These tumours primarily affect the pelvis and the perineum [1]. Only two cases of aggressive angiomyxomas which arose from the soft tissues of other sites have been reported in the literature [2,3]. This tumour has a marked preponderance for females who are in the reproductive age group [1,2]. Aggressive angiomyxomas have been reported in males to affect the inguino-scrotal region and the perineum, but the reports are very few in number [1,4,5]. To the best of our knowledge, this is the first case of extragenital aggressive angiomyxoma which involved the axilla and the anterior chest wall, which occurred in a male patient.

CASE REPORT
A 27-years-old male presented with a progressively enlarging mass in the left axillary region, which was there since 8 months. The initial size of the mass was 6x8 cm, which increased to 12x10cm and it extended into the anterior chest wall. There were no systemic complaints. On examination, the tumour mass was found to be firm and non tender and it involved the left axilla and the anterior chest wall. MRI revealed a relatively well defined mass with its anterior border overlapping the lateral border of the pectoralis major muscle, its posterior border extending beyond the mid axillary line, its superior border in the left axilla and its inferior border reaching up to the 5th rib, deep into the pectoralis major. No other lesions were detected in the abdomen or the pelvis. A clinical diagnosis of a schwannoma was given. The preoperative biopsy showed a hypocellular lesion with loosely dispersed spindle cells, with wavy buckled nuclei and hyalinized blood vessels. Based on the clinical suspicion and the histopathological features, a presumptive diagnosis of a schwannoma was rendered. A wide excision of the tumour was done and it was sent for a histopathological examination. The intraoperative findings revealed a soft, well circumscribed, lobulated mass which measured 12x7cm, which occupied the left axilla and the left side of the thorax. No adhesions to the adjacent muscle or the neurovascular bundle were detected grossly. The cut surface of the tumour showed a grey brown multinodular growth with gelatinous and myxoid areas [Table/Fig-1]. Histopathology revealed a myxohyaline vascular tumour which was composed of loose bundles of stellate to spindle cells with oval nuclei, which showed nuclear indentations. Few had prominent nuclei and a moderate eosinophilic cytoplasm which was characteristic of myofibroblasts. The surrounding collagenous stroma showed numerous thick walled blood vessels with vessel wall hyalinization. Few showed fibrin thrombi and few thin walled congested vessels. Extensive myxoid degeneration, oedema and eosinophilic, lymphoplasmacytic and mast cell infiltrates, along with extravasated RBCs, were also noted [Table/Fig-2]. The tumour showed infiltration into the adjacent skeletal muscle fibres. Immunohistochemistry revealed a diffuse smooth muscle actin [Table/Fig-3] and a focal S100 positivity. CD34 and the oestrogen receptor were negative. Based on the histopathological features and the immunohistochemistry, a diagnosis of an extragenital aggressive angiomyxoma was rendered. Six months post surgery, the patient remains asymptomatic, with no evidence of a recurrence.

[Table/Fig-1]: A well circumscribed tumor. Inset: cut section shows grey brown multinodular growth with gelatinous and myxoid areas
Aggressive angiomyxomas need to be differentiated microscopically from superficial angiomyxomas, fibroepithelial polyps, angiofibromas, myxoid neurolemmomas, myxoid malignant fibrous histiocytomas, myxoid lipomatous tumours, myoid leiomyomas and cellular angiofibromas. The clinicopathological features, along with the immunoreactivity, help in the diagnosis [1,3,5]. The presence of medium sized thick walled blood vessels, the deep location of the tumours and the absence of stromal neutrophils in this case, excluded the possibility of a superficial angiomyxoma, a fibroepithelial polyp and an angiofibroblastoma. Since a majority of the tumour cells were S100 negative, a myoid neurolemmoma was ruled out. A myoid MFH shows the features of anaplasia, namely an increase in the mitosis or the presence of an abnormal mitosis or a nuclear atypia. These features were absent in the present case. The absence of significant proportions of smooth muscle or a lipomatous component, excluded the possibility of myxoid lipomatous or smooth muscle tumours. Both cellular angiofibromas and aggressive angiomyxomas show medium to large sized hyalinized blood vessels with stromal mast cells; however, cellular angiofibromas are more cellular, more superficial in location with circumscribed margins and intraläsional fat and they usually show positivity for CD34 [9].

The high mobility group A (hMGAl) gene rearrangements on chromosome 12q13-15 have been implicated in aggressive angiomyxomas. This transcription factor is present during the embryonic life and it is absent in the adult tissue. A strong hMGA2 [2] nuclear immunoreactivity aids in differentiating these tumours from other lesions, which include angiomyofibroblastomas and cellular angiofibromas [1].

The aggressive angiomyxomas are thought to be benign, locally infiltrative and non metastasizing [8]. However, two cases of metastases of AAMs which were reported, raised the possibility of the AAMs being classified as tumours of intermediate malignancies [1,10,11].

A complete surgical resection of the tumour is the first line of treatment. Owing to the infiltrating nature of AAMs, these are associated with a high recurrence. Hence, a postoperative follow up is necessary. Hormonal therapy with gonadotropin-releasing hormones has been used as an alternative treatment for the resolution of these tumours [1,5].

CONCLUSION
Aggressive angiomyxomas are rare slow growing tumours with a high recurrence rate and they occur almost exclusively in the pelvis or perineal region. The present case is unique in being located in the axilla and the anterior chest wall. A histopathological examination, combined with immunohistochemistry, help in distinguishing these lesions from the more commonly occurring similar entities, when they are located in sites other than the pelvis or the perineum.

REFERENCES


