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AGGRESSIVE ANGIOMYXOMA OF ARM WITH IMMUNOHISTOCHEMISTRY STUDY - A CASE REPORT

Pathology

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ABSTRACT

Aggressive angiomyxoma (AA) is a soft-tissue tumor of mesenchymal origin. It is rare type of tumor occurs commonly in genital, perineal or pelvic region in reproductive age female. It's occurrence in male is very rare and acral site being extremely rare. It exhibits marked tendency for local recurrence and hence the name aggressive. It has risk of distant metastasis though it is extremely low. We report a case of AA in a 34-year-old male presenting with right arm swelling. Fine needle aspiration cytology of the growth was not of much significance. The tumor was excised and submitted for histopathological examination. A diagnosis of aggressive angiomyxoma was made based on characteristic histological features. Recently, the terminology has been changed to deep angiomyxoma instead of aggressive as the recurrences occur many years after the initial excision. In fact, it has become evident that these neoplasms have a favorable, less aggressive course if initially completely excised with negative margins.Wide local excision with 1-cm margin is considered optimal. Inspite of rarity of angiomyxoma incidence, it is extremely rare in males and site as well.

KEYWORDS

angiomyxoma, myxoid, soft tissue tumor

CASE REPORT:

A 34 year old male came to surgery outpatient department presenting with swelling over deltoid region of right arm since 10 years. Swelling was painless and slowly progressive. A clinical diagnosis of soft tissue tumor was made. Ultrasonography showed ill-defined lesion with vascularity within and was suggestive of neoplastic etiology. Magnetic resonance imaging (MRI) showed well defined lesion in deltoid muscle with myxoid component showing heterogenous enhancement suggesting possibility of low-grade soft tissue neoplasm.

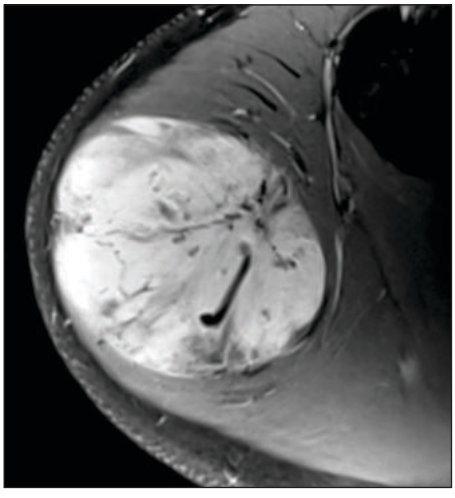

Fig 1: MRI Showing Mass In Deltoid Muscle With Myxoid Component

Fine needle aspiration cytology was diagnostically inconclusive showing only fibrocollagenous and adipose tissue with myxoid change and proliferating capillaries. Wide local excision of mass was done and sent to histopathology section. We received single globular mass of size 10x 8.5x 6 cm which was capsulated. On cut section, homogenous mass with glistening surface was seen. Microscopically, tumor was poorly circumscribed having variable sized thick and thin blood vessels with muscular hypertrophy which were separated by loose myxoid stroma having stellate cells and collagen fibers. Tumor was infiltrating in underlying muscle and fat. There was no evidence of any pleomorphism, hyperchromasia, increased mitosis or necrosis. Immunohistochemical studies further showed positive staining of tumor cells for CD34, vimentin and smooth muscle actin (SMA) and negative for desmin, estrogen receptor (ER) and progesterone receptor (PR). The typical microscopic features supported by immunohisto chemical findings suggested an aggressive angiomyxoma as the diagnosis.

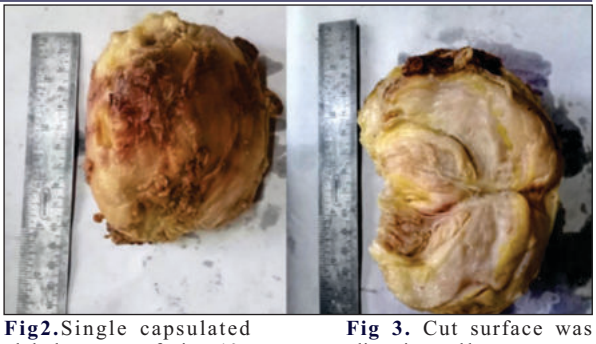

Fig2. Single capsulated globular mass of size 10x 8.5x 6 cm.

Fig 3. Cut surface was glistening and homogeneous

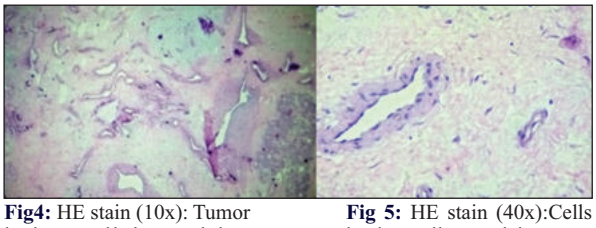

Fig4: HE stain (10x): Tumor is hypocellular and has prominent thick and thin walled blood vessels.

Fig 5: HE stain (40x):Cells having stellate nuclei.

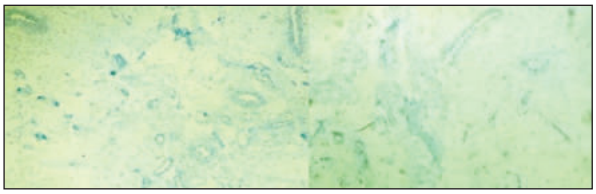

Fig 6: Estrogen receptor-negative

Fig7: Progesteron receptor-negative

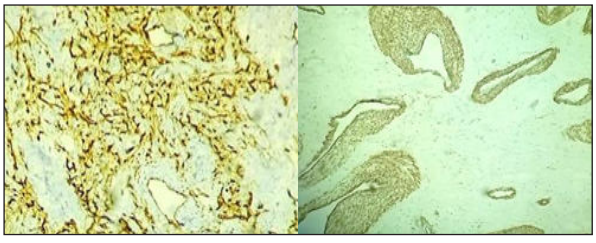

Fig 8: Cd34- positive

Fig 9: SMA- positive

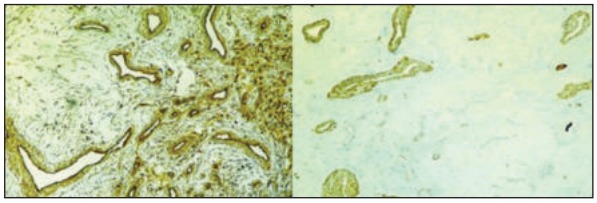

Fig 10: Vimentin- positive **Fig 11:** Desmin- negative

DISCUSSION:

Steeper and Rosai first described nine cases of a distinctive, infiltrative, locally aggressive but non-metastasizing fibro-myxoid soft tissue tumor arising in the pelvis and perineal area of young female patients, which they termed aggressive angiomyxoma (14). Angiomyxomas are commonly seen in female as pelviperineal pathology [1]. A male to female ratio is 1:6, having predilection for females [2]. Scrotum, spermatic cord, inguinal region, prostate and epididymis are the sites involved in males [3–7]. Extragenital AA can be possible [15]. Histopathological examination is of much significance as most cases are diagnosed by histopathological examination after surgical excision [8,9]. The proposed origin of this rare tumor is myofibroblastic. Important differentials of AA include various benign and malignant soft-tissue lesions chiefly including fibroepithelial polyp, angiomyofibroblastoma, superficial angiomyxoma, and myxoid lipomatous tumors [10]. Desmin staining can be absent in few cases as noted in earlier reports [11]. Hypocellularity of the tumor along with the immunohistochemical staining for CD34, SMA and vimentin favoured the diagnosis of angiomyxoma in our case [11].ER and PR negative AA have also been reported in literature, particularly in male patients. [15]

CONCLUSION:

Angiomyxoma is rare entity which has potential to recur. Therefore, it should be distinguished from other myxoid lesions. Appropriate diagnosis, complete resection and close follow up is recommended.

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