



### **Case Report:**

## **Aggressive Fibromatosis in Neck.**

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**Abstract:** Aggressive fibromatosis (AF) is a locally aggressive infiltrative low-grade benign tumor that accounts for approximately less than 3% of all soft tissue tumors. In the head and neck region this tumor tends to be more aggressive and associated with significant morbidity. Aggressive surgery is a viable management option and may be successfully used as a single modality treatment, or in combination with radiotherapy. We report a rare case of AF in a 38 year old female, who presented with a painless mass over the left supraclavicular fossa, extending inferiorly into the thoracic inlet, which was excised successfully in toto with the help of cardiothoracic vascular surgeon (CTVS).

**Key Words:** Aggressive fibromatosis; Supraclavicular fossa; Benign tumor.

### **Introduction**

Aggressive fibromatosis (AF), also known as desmoid tumor, is a rare benign connective tissue disorder originating from the fascia and musculoaponeurotic tissues with local aggressive behaviour.(1) AF is responsible for less than 3% of all soft tissue tumors. The incidence of AF is 2 to 4 per 1 million individuals a year with a female to male ratio 3 to 1.(2,3) AF when occurs in the head and neck region tends to be more aggressive and associated with significant morbidity, which may be attributed to the vital vascular, neurological or anatomical structures lying in its close proximity. In AF, the treatment of choice is surgery with broad resection of margins. If tumor free margins cannot be obtained for functional or cosmetic reasons, postoperative radiotherapy improves local control.(3) We report a rare case of AF of neck in a 38 year old female, who presented with a large painless mass in the left supraclavicular fossa extending inferiorly below the clavicle and going towards the thoracic inlet. Patient was managed by wide local excision. As the mass was firmly attached inferiorly with the periosteum of

clavicle it was excised in toto with the help of CTVS surgeon. Patient has been on regular follow-up for six months without any local recurrence.

### **Case Report:**

A 38 years old female presented to ENT OPD with a seven-month history of a swelling over the left side of neck. The swelling was gradual in onset, progressive in nature and painless. There was no history of trauma, use of oral contraceptive pills, and weakness of upper extremity. On physical examination a fixed mass approximately 8x6 cm in size was present in the left supraclavicular region, not associated with any overlying skin changes. The mass was found extending superiorly 4 cm below the mastoid tip, medially involving the lateral half of left clavicle and laterally extending to anterior border of trapezius. On palpation surface of the swelling was smooth, firm in consistency, with ill defined margins, non tender, non mobile, non compressible, non fluctuant (Figure 1).

A contrast enhanced computed tomography (CECT) of neck revealed a 6.5x7.4x6.5cm large well defined heterogeneous mass lesion in left supraclavicular region extending inferiorly below the clavicle, going into the thoracic inlet (Figure 2). Fine-needle aspiration cytology (FNAC) of the swelling showed few benign spindle shaped cells with elongated vesicular nuclei and pink cytoplasm admixed with pink fibrocollagenous material suggestive of fibromatosis.

The patient underwent wide local excision of the tumor. A horizontal incision was made over the tumor, trapezius was found stretched out and overlying muscles were separated from the tumor. The tumour was freed from all the surfaces but was found attached firmly to periosteum of clavicle and going towards the thoracic inlet. Tumour was removed in toto with the help of CTVS surgeon. Tumour was sent for histopathological examination (HPE).

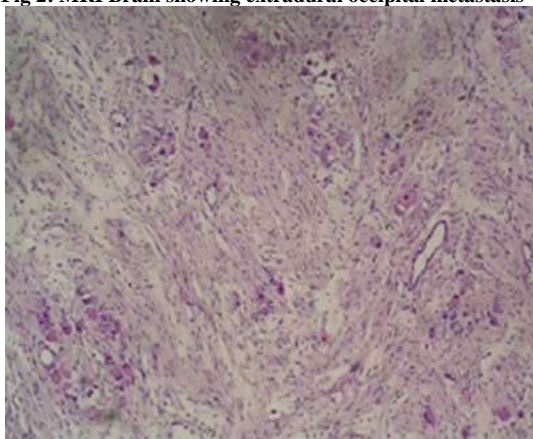
HPE of the excised tissue showed spindle cells arranged in fascicles and sheets with dense collagen tissue seen in between the spindle cells at few places, correlating with the findings of AF (Figure 3).



**Fig 1: Mass over the Left supraclavicular fossa**



**Fig 2: MRI Brain showing extradural occipital metastasis**



**Fig 3: Microphotograph (H/E staining 40 x) showing benign spindle cell tumor, favouring fibromatosis.**

**Discussion:**

AF is a locally aggressive infiltrative low grade tumor.(4,5) Fibromatosis has been described under a variety of synonyms e.g. desmoids tumour, desmoid fibromatosis, non-metastasizing fibrosarcoma etc. (6) The patients age range from newborn to 70 years, with most of cases appearing in the third and fourth decades.(7) AF is found to be more common in the female, with female/male ratio between 3:2.(8) The supraclavicular fossa is the most common site for head and neck fibromatosis (40-85 per cent) followed by the face (about 25 per cent).(7)



**Fig 4: Photograph showing excised tumour of fibromatosis.**

The most common presenting symptoms are an asymptomatic / painless mass or neurologic symptoms, including pain, radiculopathy, or weakness in upper extremity (9). In our case although the mass was large in size, but no neurological symptoms like radiculopathy or weakness of the upper extremity was present. MRI is the investigation of choice for assessing extension of the disease and follow-up because of better soft tissue delineation compared to CT.(10)

Treatment of AF also consists of wide surgical resection, a combination of surgery with radiotherapy, or radiotherapy alone.(11) Complete three dimension excision of the tumour is considered to be the only effective method of cure by most authors.(7) However surgical excision of the tumor is challenging for ENT surgeon in the head and neck region, as preservation of vital structures and their function may impede this objective of complete excision.(12) In our case as the tumor was firmly adhered to the clavicle, the help of CTVS surgeon was sought for its complete removal.

Long term follow- up is necessary because regrowth or recurrence could occur after a long period of stabilization.(13)

**Conclusion:**

Fibromatosis is one of the rare tumors with aggressive behaviour. Anatomical locations, especially in the head and neck regions, with wide extensions may be challenging for its complete removal. Due to its aggressive nature to invade the surrounding structures, surgery is a viable management option for AF. Long term follow-up are required as recurrence is reported even after a long period.

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