Abstract: 32 yrs male with complaints of swelling over right upper back for 8 months with restriction of shoulder movements. X-ray was not confirmatory, MRI came as chondrosarcoma. Scintigraphy showed no evidence of skeletal metastasis. Tru cut biopsy was not confirmatory. Open biopsy report was DD of fibromatosis and neurofibroma. Confirmed as fibromatosis and subtotal scapulectomy was done with preservation of glenoid.

Keyword: Fibromatosis is an aggressive benign soft tissue tumor

Introduction: Fibromatosis refers to a group of benign soft tissue tumor which have certain characteristics in common, including absence of cytologic and clinical malignant features, a histology consistent with proliferation of well-differentiated fibroblasts, an infiltrative growth pattern, and aggressive clinical behavior with frequent local recurrence.

Clinical features: A 32 yrs male with C/O pain in Right shoulder for 8 yrs and Swelling in Rt. Upper back 8 months. Pain in shoulder for 8yrs. Pricking nature, present at rest and aggravated during work. Noticed a swelling in upper back 8 months ago insidious onset, gradually increasing in size and now attained the present size.

H/o restriction of movements around shoulder joint.

No H/o fever, loss of weight, loss of appetite or cough with expectoration

H/o biopsy done twice.

O/E: A 4cm scar seen behind the posterior axillary border. Fullness in axilla present. Diffuse swelling around 8x12 cm extending from inferior, lateral, posterior border of scapula to the axilla. Firm to hard in consistency, no lymphadenopathy.

Both active and passive movements of abduction, flexion, external rotation movements are restricted.

Pre op clinical photo
MRI: multiple exostosis in the right scapula and clavicle with enhancing intramuscular soft tissue lesions in subscapularis and lattissmu dorsi. Suggestive of malignant transformation of osteochondroma (Chondro sarcoma)

Scintigraphy:

- Tru-Cut Biopsy: Section shows tiny fragments consisting of neoplasm of soft tissue and intervening fibrocartilagenous tissue. Suggestive of soft tissue neoplasm. Exact nature of tumor could not be determined. If clinically suggestive of malignancy biopsy has to be repeated.

Intra op photo

Excised Scapula

HPE of specimen: Suggestive of Fibromatosis with inferior, lateral and posterior margins of scapula positive.

Discussion:

Fibromatosis is a non metastasizing spindle cell proliferation with high potential of local infiltration and recurrence. Aggressive fibromatosis is a type of musculoskeletal fibromatosis while it is a non metastasising fibrous lesion. It is thought to be a true neoplasm that arise from fascial and musculo-aponeurotic coverings. It does not have any propensity for metastasis although can aggressively invade local structures. The optimal management for aggressive fibromatosis depends on tumor location and extent. Surgical resection is the main stay of treatment to give a local clearance. Surgery done is radical excision with a wide margin and/or radiation. Moderate-dose radiotherapy alone for gross disease or after a microscopically incomplete resection yields local control rates of approximately 75-80%. Treatment with pharmacologic agents Tamoxifen, COX-2 inhibitor or NSAID results in objective response rates of approximately 40 to 50%. The likelihood of local recurrence after surgery is high, particularly if margins are positive. Despite their local infiltrative and aggressive manner mortality is minimal to non-existent in peripheral tumor. Most unpredictable outcome, usual recurrence in 1 year. Absence of clinical recurrence for 18 to 20 months is a favourable outcome.

Conclusion: Aggressive fibromatosis is a locally aggressive benign tumour arising from the musculo-aponeurotic coverings and invade the bone which need wide local excision to prevent local recurrence.