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A Case of Large Extra Abdominal Fibromatosis (Desmoid Tumor) Over Back

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I. Introduction

Extra abdominal fibromatosis (EAF) is proliferation of highly differentiated fibrous tissue. It arises principally from the connective tissue of the muscle & the overlying fascia or aponeurosis (Musculoaponeurotic fibromatosis). It chiefly affects the muscles of the shoulder, pelvic girdle & thigh of adolescents & young adults. Histologically it is benign but locally aggressive fibrous neoplasm originating from the musculoaponeurotic structures with nonspecific findings.

EAF constitutes < 0.03% of all the neoplasms. They are rare lesions with an estimated incidence of 2-4 individuals per million. Other terms used are Extra abdominal desmoid, Desmoid tumor, Well differentiated non metastasizing fibrosarcoma, Grade 1 fibrosarcoma AND Aggressive fibromatosis.

II. Etiopathology and epidemiology

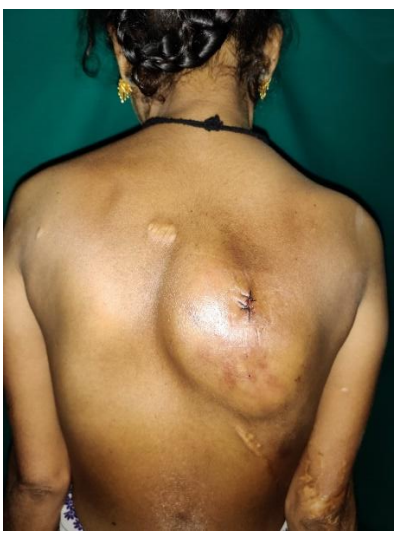
Etiology is probably multifactorial. Genetics, endocrine and physical factors seem to play role in its pathogenesis. Possible risk factors include female sex, previous history of trauma/surgery/pregnancy. Mutations in the APC/Beta catenin pathways were identified in the majority of sporadic EAF.

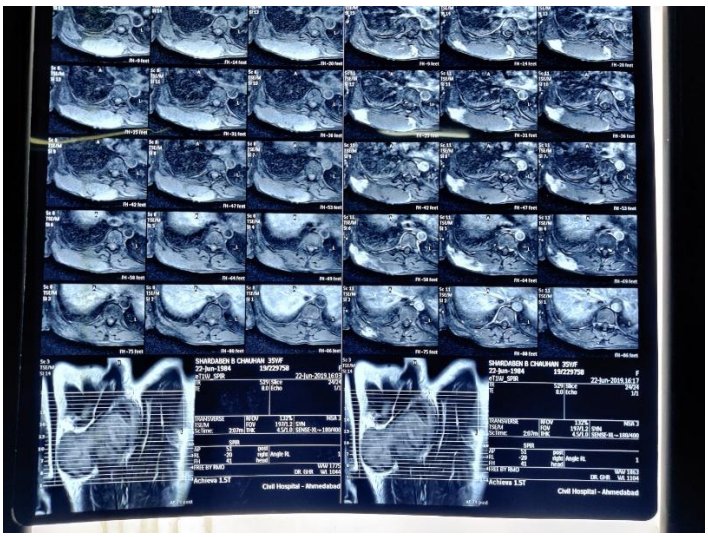
EAF is most common in patient between puberty and 40 years age with peak incidence between the ages of 25-35 years. These tumors are two to three times more common in females than in males. Most patient presents with deeply situated, firm poorly circumscribed mass that has grown insidiously and causes little or no pain. Neurogenic symptoms include numbness, tingling or stabbing/shooting pain or motor weakness may occur when the lesion compresses nearby nerves. Principle sites of EAF is musculature of the shoulder > chest wall > back > thigh > Head & Neck. (chest wall and back – 17.2%). 5% of these EAF are found to be multicentric.

III. Case report

History

A 35-year Hindu married female coming from low socio-economic class presented in OPD with complain of swelling over right upper back since last 6 to 8 month and pain in swelling since 1 to 2 months. There was antecedent history of trauma (laceration) over back due to attack by pig before 4 years, and it was primarily sutured then. Patient was vitally stable. Patient had no other comorbid condition.





Examination

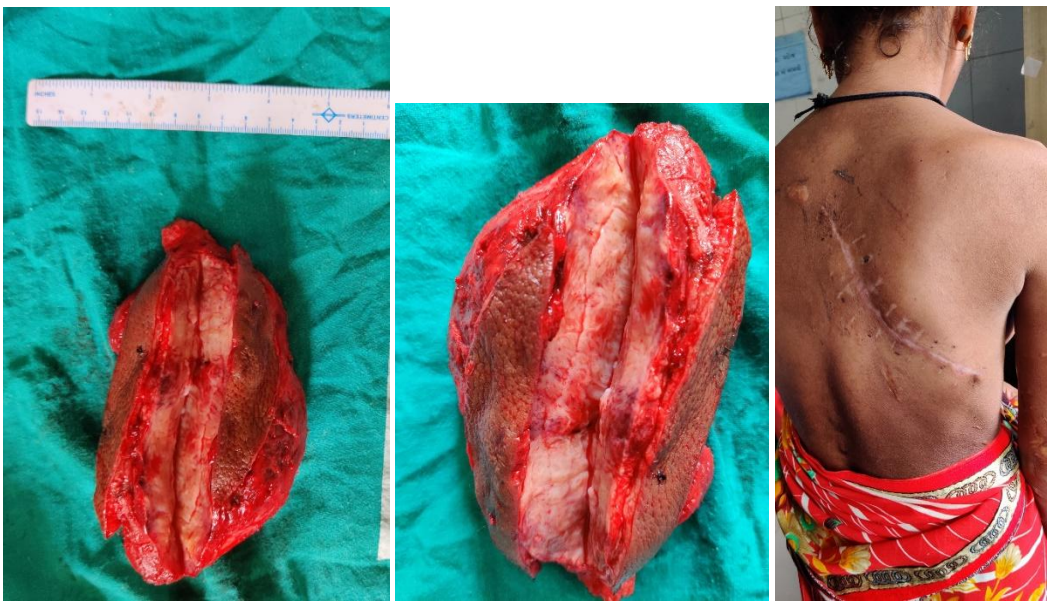
Swelling measuring 16cm*10cm*4cm in size, single, oval shaped, obliquely placed over right upper back, extending from T4 to T12 level vertically and from midline to posterior axillary fold horizontally, crossing the midline for 1cm, firm to hard in consistency with well-defined edge and margin, relatively fixed not showing mobility in any direction, overlying skin is smooth showing sutured wound of incisional biopsy, without impulse on coughing.

Investigation

Patient's routine blood investigation were normal. Chest X-Ray, Abdominal-X Ray and X ray D.L. L.S. spine was normal. MRI study suggested benign mesenchymal neoplasm deep to subcutaneous plane. FNAC was not conclusive, incision biopsy showed vascularized fibrocollagenous tissue.

Management

The tumor was surgically excised and it was in subcutaneous plane without infiltrating skin or muscle. Skin was primarily closed with negative suction drain in subcutaneous plane.



The tumor was firm in consistency which cut with gritty sensation and on cross section it revealed glistening white surface measuring 13.5cm*7cm*5cm in size.

The specimen was sent for HPE and it suggested benign spindle cell tumor with possibility of extra abdominal fibromatosis. On Immunohistochemistry it suggested diffuse beta-catenin nuclear staining. Post-operative recovery was uneventful. There is no recurrence till 1 year follow up.

IV. Discussion

In 85% cases, EAF have mutation in exon 3 of the CTNNB1 gene that activates beta-catenin. Physical factors such as a trigger mechanism, as examples of EAF have been reports in the chest wall following trauma and reconstructive mammoplasty. Large studies of EAF have been reported an antecedent history of trauma in 16-28% of cases. In a small proportion of desmoid, Beta-catenin is activated because of germline mutation in the APC gene; these desmoids occur in parallel with familial adenomatosis polyposis in Gardener's syndrome. Although incapable of metastasizing EAF often recur. They can rarely cause death when they involve or compress a vital structure (such as occurs with tumors in Head & Neck). Because the microscopic picture does not reliably reflect the growth potential of the tumor, therapy should be predicted on its extent and anatomic relations. Surgical resection with wide local excision has been the main stay of the therapy. Wide local excision is recommended as most studies have found a lower rate of recurrence when excision includes a wide margin of uninvolved structures around the grossly visible tumor. Recurrence is variable in literatures ranging from 19% to as high as 77% with average of 40%. Although tumor size was associated with risk of local recurrence, microscopically positive margins did not predict recurrence risk. However, studies have found extent and adequacy of initial excision to be progressively significant. Spontaneous regression of these tumors has been observed in sporadic cases, particularly around menarche and menopause. In situations where wide local excision cannot be performed, postoperative radiation therapy seems to be indicated for tumors that are incompletely

excised. Post-operative radiation therapy is reported to be helpful in local control. Only few studies have been done for evaluation of neoadjuvant chemoradiotherapy.

V. Conclusion

EAF is rare soft tissue tumor. It lacks metastatic potential but it can grow aggressively in a locally infiltrating pattern. EAF frequently recur after surgical excision, which remains the treatment of choice. Tumors should be removed as soon as possible after identification in order to achieve the most optimal resection possible. MRI is the imaging modality of choice but diagnosis is confirmed histologically. This tumor continues to present a problem in recognition and management, especially because of the striking discrepancy between its deceptively bland microscopic appearance and its propensity to recur locally and infiltrate neighboring soft tissues. Fibrosarcomatous transformation of fibromatosis is exceedingly rare. Most tumors measure 5-10 cm in greatest dimension, although lesion as large as 20 cm have been reported. We reported this case as it is the largest size of EAF over back in female patient to the the best of our knowledge

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