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Giant Fibromatosis of the Chest Wall: A Rare Entity

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Abstract: Fibromatosis also termed as desmoids is a rare benign but locally aggressive neoplasm characterized by mass like or infiltrative growth of fibrous tissue. It usually arises from the abdominal wall or the extremities, rarely affects chest wall representing only 8-10% of all deep fibromatosis. Fibromatosis is locally aggressive but does not metastasize. Here, we present a rare case of fibromatosis of scapular region, treated successfully by wide local resection.
Keywords: Fibromatosis, Scapula, Desmoid

INTRODUCTION

The term "fibromatosis" refers to a broad spectrum of fibrous tissue proliferations characterized by infiltrative growth into the adjacent tissues, accounting for only 0.03% of all the neoplasms [1]. They are locally aggressive, but rarely metastasize [2]. They arise from the connective tissue of the muscle and the fascia or from the aponeurosis. Fibromatosis may occur in a variety of anatomical locations such as abdomen wall, extremities, girdle, chest wall, head and neck and rarely involves peritoneum [3]. Fibromatosis of Chest wall is a rare entity and very few cases have been described in the literature Fibromatosis display local aggressiveness, with complete resection being the treatment of choice.

CASE REPORT

A 32 year old gentleman presented to us with gradually increasing swelling over right scapular region, with duration of 10 months and recent onset of pain in the swelling since 1 month. There were no other relevant symptoms. Physical examination showed a large lobulated mass with firm to hard consistency palpable in right scapular region, extending from lateral border of scapula towards midline with restricted mobility.

CECT scan revealed a well circumscribed non enhancing soft tissue lesion measuring 16 x10 x8cm seen at the inner aspect of left scapula likely between and underlying the subscapularis muscle on lateral aspect , erector spinae & serratus anterior on medial aspect and trapezius on lateral aspect. No evidence of bony invasion and any intrathoracic extension. Metastatic workup was negative. Trucut biopsy of the swelling confirmed the diagnosis of fibromatosis.

In view of no major vascular involvement / chest wall infiltration and preoperative diagnosis of fibromatosis, the patient was planned for surgery. Intraoperatively , there was large multilobulated mass measuring 18 x 10 cm in the left scapular region, She underwent enbloc resection of the tumor with sleeve resection of underlying and surrounding muscles namely latissimus dorsi , subscapularis and serratus and anterior in order to achieve negative margin. Patient underwent extensive physiotherapy in the postoperative period and entire course was uneventful.

Gross examination showed globular soft tissue mass measuring 16 x 10 x 6 cm, covered by fascia and partly by muscle. Cut surface of the specimen was fasiculated and pinkish gray in color. Microscopic examination revealed elongated and spindle shaped cells with minimal nuclear pleomorphism; collagenisation and perivascular inflammation. Immunohistochemistry study showed tumor cells positive for Vimentin and negative for Desmin, CD 117, smooth muscle actin [SMA], S-100, CD-34, Pan CK and low ki-67 proliferative index. Final diagnosis of fibromatosis was done. No recurrent lesion was found using a local imaging at 8 months after completion of treatment.

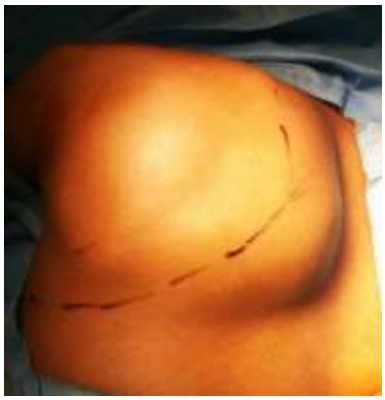

Fig. 1: Physical examination revealed large lobulated mass in left scapular region

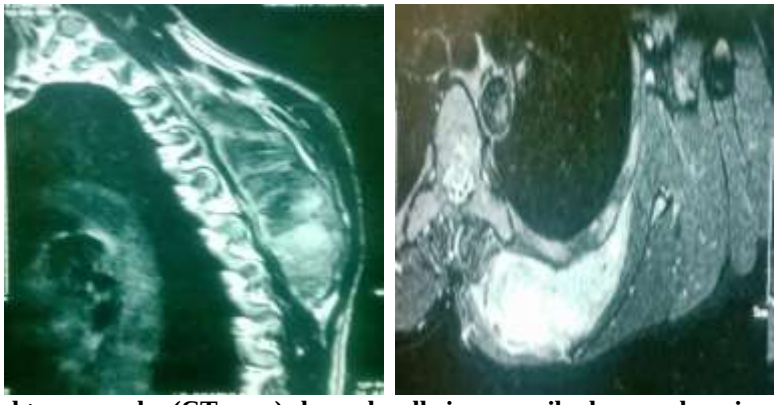

Fig. 2a & 2b: Computed tomography (CT scan) showed well circumscribed non enhancing soft tissue lesion at the inner aspect of left scapula

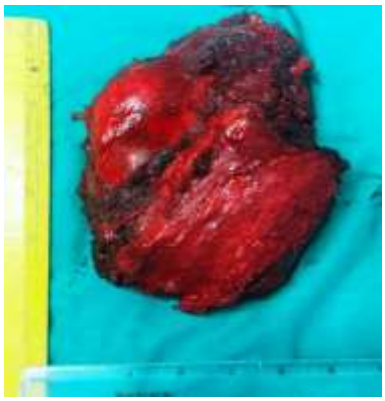

Fig. 3: Specimen photograph revealed large encapsulated mass covered with fibromuscular tissue

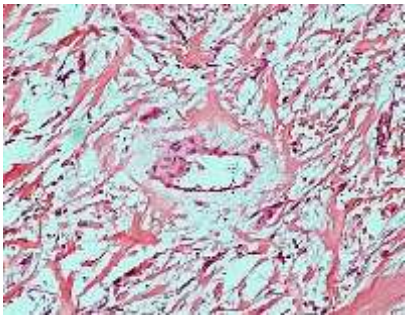

Fig. 4: Microscopy revealed scanty spindle shaped cells with minimal nuclear pleomorphism among bundles of dense collagen [H.E.X20]

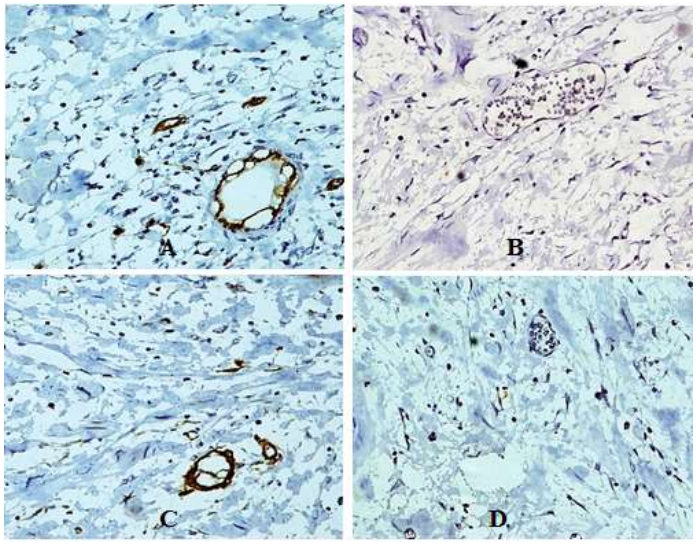

Fig. 5: (A) Immunohistochemical examination (20x), Tumor cells showed negativity for CD117. (B) Tumor cells stained negative for SMA. (C) Tumor cells negative for DOG-1. (D) Tumor cells stained negative for CD-34.

DISCUSSION

Fibromatosis refers to a group of fibrous tissue proliferations of benign nature [4]. It may arise from connective tissue of muscles, fascia or from the aponeurosis [5]. They are characterized by an infiltrative growth with a tendency towards recurrence and they never metastasize [6]. Fibromatosis have distinct biological behavior, characterized by initial rapid growth, followed by stability or even regression [7]. Mueller described these tumors as "Desmoid tumor" in 1838, referring to their tendon like consistency [8].

Majority of fibromatosis occur sporadically, or in association with Gardener syndrome, familial adenomatosis polyposis coli (FAP) and bilateral ovarian fibromatosis [7, 9]. Clonal chromosomal aberrations-trisomies of chromosome 8 and 20 has been seen [10]. Risk factors include pregnancy, previous abdominal surgery, trauma and estrogen therapy. Exact etiology and natural course is not well understood [7]. Majority of fibromatosis occur in women of child bearing age but there is no gender or racial predominance [7, 11].

Fibromatosis are broadly classified into two groups: superficial (fascial) and deep (musculoaponeurotic) [12]. Desmoid tumors also termed as deep fibromatosis were first described in the abdomen wall by Mc Farlane in 1832 [13]. Extra-abdominal fibromatosis occur in extremities, shoulder girdle [14], chest, neck wall [15]. Fibromatosis of chest wall represents 8-10% of all cases. Extra-abdominal desmoids, local recurrence rates is reported to be ranged from 24% to 77% [16]

Based on study by Abbas et al. the mean age at diagnosis was 39 years with a range between 10 and 78 years [17]. Fibromatosis of chest wall usually in the form of swelling of various sizes [18]. Histological pattern does not reflect the growth potential of the tumor, hence recurrence cannot be predicted [12]. Ultrasonography (USG) or computed tomography (CT SCAN) may be helpful in diagnosis but ultimately, final diagnosis is made by excision and histological examination and immunohistochemistry (IHC) studies [7].

Treatment is usually based on the extent and the anatomical relationship of the tumor [12]. Fibromatosis should be treated mainly with surgery. Chemotherapy, targeted agents, NSAIDs and antiestrogen therapy carries minimal benefit. Surgical excision is the only curative method of treatment with ill-defined role of medical or radiation therapy [7].

Surgical approach includes wide local excision of these tumors as they have tendency toward local recurrence [7, 19]. Despite curative surgery, fibromatosis are prone for local recurrence [7]. Recurrent tumors can be treated by local excision and adjuvant radiotherapy [12]. Postoperative radiotherapy is indicated in close or positive margin. Role of chemotherapy is not well established. Certain drugs like vincristine, actinomycin, cyclophosphamide, interferon alpha and Dacarbazine have limited role with minimal response.

Patients with fibromatosis have a prolonged survival even in advanced cases. Some authors recommended a trial of observation with antiestrogens, nonsteroidal anti-inflammatory drugs such as sulindac with limited role of targeted agents such as Imatinib [7]. Progression of desmoids is highly unpredictable [20]. Spontaneous regression has been reported in the literature, in the absence of any treatment or partial excision [18, 21]. Fibromatosis have better survival rate

(93% at 5 years), the probability of recurrence after 5 years is an estimated 29% [22, 23].

CONCLUSION

Primary tumors of the chest wall are rare and there is little information available in the literature. Chest wall fibromatosis is usually benign tumor but locally aggressive. Fibromatosis should be considered in differential diagnosis of chest wall neoplasm's. Majority are asymptomatic, and difficult to diagnose based on clinical presentation and radiological investigation. Final diagnosis is usually made on histopathology and immunohistochemistry (IHC) studies. Surgical excision is the only curative method of treatment with ill-defined role of medical or radiation therapy. Despite curative surgery, fibromatosis are prone for local recurrence.

Abbreviations

FNAC- Fine needle aspiration cytology, IHC- Immunohistochemistry, CK- Cytokeratin, SMA- Smooth muscle antigen, CD- Cluster of differentiation

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