CASE REPORT

A young woman with multiple soft tissue tumors in various parts of the body, (Extra Abdominal Fibromatosis and Lymphangioma in both Axillas)

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Abstract:

The term fibromatosis refers to a group of benign soft tissue tumors (fibromas), 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. Lymphangiomas are malformations of the lymphatic system, which is the network of vessels responsible for returning to the venous system of excess fluid from tissues. Here we report a case of 30 years old lady with history of multiple soft tissue tumours in thigh, axilla and abdomen. On physical examination the swelling of the thigh and abdomen were firm and those of axilla were soft. All the tumours were excised surgically and diagnosis of thigh swelling was fibromatosis and that of axilla was lymphangioma, on histopathology.

Introduction :

Fibromatosis is a broad group of benign fibrous tissue proliferations of similar microscopic appearance that are intermediate in their biological behaviour between benign fibrous lesions & fibrosarcoma.[1] Like fibrosarcoma they are characterized by infiltrative growth & tendency towards recurrence, but unlike fibrosarcoma they never metastasize. Subtypes of fibromatosis include 1. Superficial (fascial) – Palmar fibromatoses, Plantar fibromatoses, Penile fibromatoses, and Knuckle pads. 2. Deep (musculoaponeurotic) -Abdominal & intra-abdominal fibromatoses, Extra-abdominal fibromatoses. The exact histogenesis remains unexplained. Trauma, endocrine & genetic factors have been implicated, but there is still much uncertainty whether any of them play a major role in the development of the disease.[1, 2]

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Aggressive fibromatosis is a rare condition marked by the presence of desmoid tumors, which are benign, slow-growing tumors without any metastatic potential.[3] Despite their benign nature, they can damage nearby structures causing organ dysfunction. Most cases are sporadic, but some are associated with familial adenomatous polyposis (FAP). Approximately 10% of individuals with Gardner's syndrome, a type of FAP with extracolonic features, have desmoid tumors.[5] Treatment includes prompt radical excision with a wide margin & or radiation. Despite the local infiltrative and aggressive behavior of these tumors, mortality secondary to these tumors is minimal to nonexistent.[3]

Lymphangiomas are malformations of the lymphatic system, which is the network of vessels responsible for returning to the venous system of excess fluid from tissues.[2] These malformations can occur at any age and may involve any part of the body, but 90% occur in children less than 2 years of age and involve the head and neck. Since they have no chance of becoming malignant, lymphangiomas are usually treated for cosmetic reasons only. Treatment includes aspiration, surgical excision, laser and radiofrequency ablation, and sclerotherapy.[5]

Case History:

A 24-years old lady was admitted in the surgery unit of Shaheed Shurawardy Medical College Hospital, Dhaka with the complaints of multiple swellings in various parts of the body for 3 years. The swelling over the lower part of back of right thigh appeared first and subsequently the swelling over left thigh, both axillae and abdomen appeared, all were initially painless and gradually increasing in size but later became painful and there was sudden increase of size

causing difficulty in walking. She gave no history of fever, anorexia, cough, bony pain, significant weight loss, and bowel or bladder abnormalities. She has no history of diabetes mellitus, hypertension, jaundice or bronchial asthma.

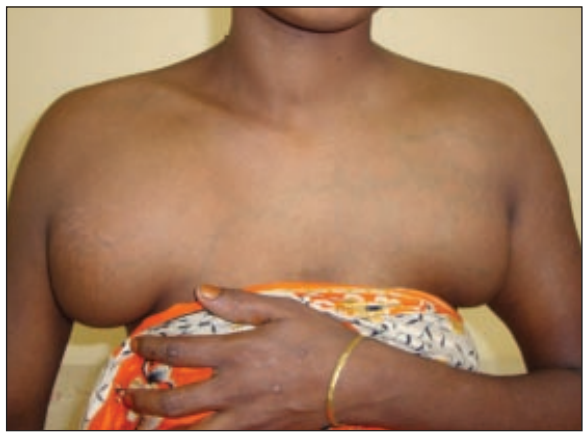


fig1: swelling of both axillae

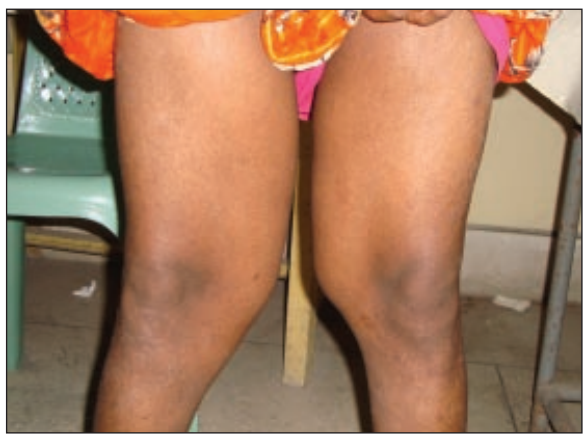


fig2: swelling of thigh

On general examination the patient was conscious, co-operative, her pulse, blood pressure and respiratory rate were normal; there was no accessible palpable lymph node, no engorged neck gland or neck vein. On local examination, the swellings of both thigh and abdomen were non tender, firm, measuring about 25x 8 cm and 4x2cm in size respectively, with smooth surface and ill defined margin; The swellings were free from overlying skin but fixed with the underlying structures and were mobile from side to side. The local temperature was normal. Both the axillary swellings were soft, non tender, non compressible, irregular in shape, measuring about 8x6cm in size, occupying the anterior portion of the axilla and extended into the upper part of front of chest, free from overlying skin and underlying structures with normal local temperature and skin condition. The locomotor system showed mechanical difficulty in walking but other than that all other systemic examination no abnormality was detected

Initially an incisional biopsy was done from the left thigh swelling and histogolical report revealed fibrolipoma. All other investigations done for general anaesthesia revealed no abnormality. X-ray of thigh showed soft tissue tumors without any bony deformity. CT- scans and MRI were not done.

The tumors of both thigh and axilla were removed by three stages of surgery. The thigh swellings were approached through a curved incision over the posterior aspect of the left thigh. The underlying hamstrings were involved along with deep fascia.

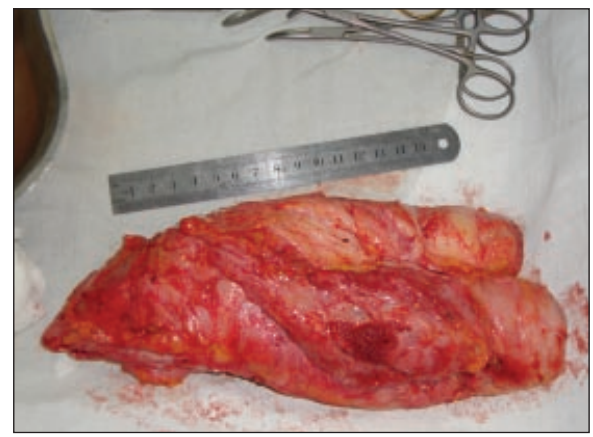


fig3: excised thigh lump

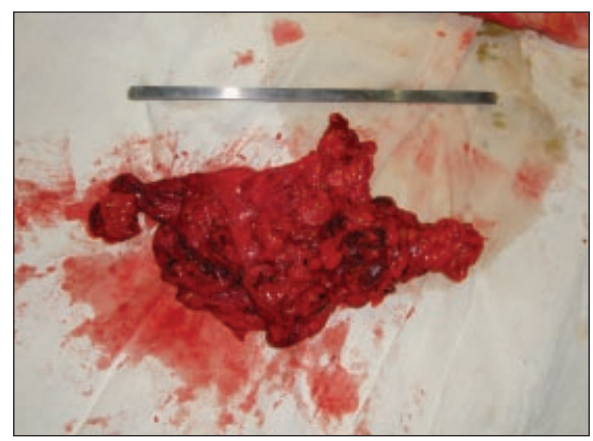


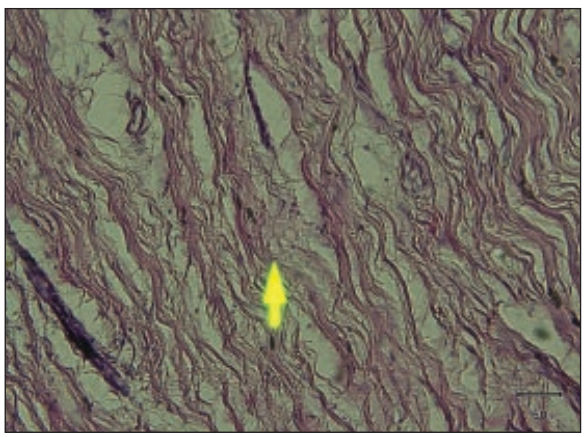
fig4: excised axillary lump

The lump was totally excised with the involved muscle and fascia, the axillary swelling was exposed by a transverse curved incision; adhesions with the surrounding axillary sheath and fat was released and lump was completely excised along with some lymph

nodes. The anaesthetic recovery and post –operative period were uneventful. All the specimens were sent for histopathological examination. Reports were: tissue from both thighs: extra-abdominal fibromatosis, and tissue from axilla: lymphangioma.

Discussion:

Extra-abdominal fibromatosis is a relatively common occurrence, in which there is striking discrepancy between its deceptively harmless microscopic appearance & its potential to attain a large size, to recur, & to infiltrate neighboring tissues in the manner of a fibrosarcoma[1]. Despite its bland microscopic appearance, the tumor frequently behaves in an aggressive manner & recurs in a high percent of cases following excision.[6] Based on the anatomical distribution, the principal location of the tumor is the musculature of shoulder, followed by that of the chest wall & back, the thigh & the mesentery[1,2,4]. Nearly always the tumor is confined to the musculature & the overlying aponeurosis or fascia.[1] Occasionally it extends along the fascial plane or infiltrates the overlying subcutaneous tissue, especially in those cases that reach a large size.[1] The average size of the tumor mass varies considerably & ranges between 5 & 10 cm in diameter, rarely 20 cm.[1] Two lesions must be primarily considered in differential diagnosis: (1) Fibrosarcoma ,(2) Reactive fibrosis.[1]



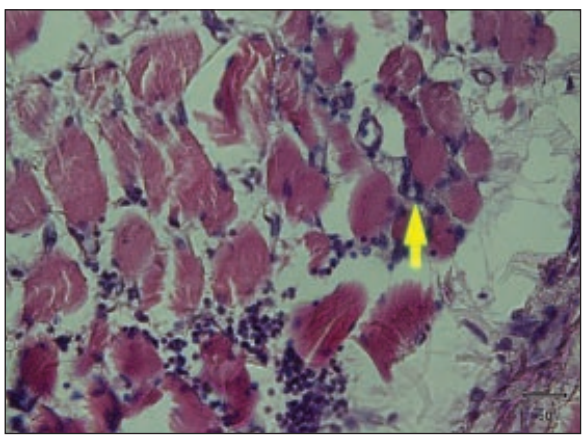


fig5: histological picture of fibromatosis

Because the microscopic picture does not reflect reliably the growth potential of the tumor, therapy is predicated upon its extent and anatomical relationship.[1] In view of the high recurrence rate, it seems mandatory to treat all tumors showing gross involvement of muscle by prompt radical excision, including excision of a wide margin of uninvolved structures around the grossly visible tumor[1,11]. In view of the excellent prognosis in regard to patient survival, amputation or other mutilating procedures should be done only for palliative reasons or if the extent of the tumor or the threat of complications leave no other choice[1]. More recent treatment includes watching and waiting, prompt radical excision with a wide margin and/ or radiation (median dose, 5000 rad), antiestrogens and NSAIDs, or chemotherapy.[6,7] Despite adequate margins, local recurrence is a significant problem that has been reported to range from 25% to 77% at ten years.[11] There has been considerable controversy on the role of post surgical irradiation in preventing relapse. Nevertheless, there is accumulative evidence on the potential benefit of radiotherapy as an adjunct to surgery both in primary and recurrent disease.[11] Wide resections with negative margins have generally been correlated with lower recurrences.[11] A reasonable explanation for this indiscrepancy is that these tumors may extend through fascial planes among muscle bundles, limiting a reliable estimate of the disease extent during surgery. A second reason is the reluctance to perform a mutilating surgery in a benign setting that prefers to encircle major neurovascular structures[11]. Aggressive fibromatosis is a very rare neoplasm arising from the musculoaponeurotic structures. It is characterized by locally aggressive growth, and a tendency to relapse but not to metastasize. Long-term immuno intervention with pegylated interferon alfa-2b, however, led to marked clinical improvement of the patient's condition and a radiologicaly proven stabilization of the disease.[13] Despite the local infiltrative and aggressive behavior of these tumors, mortality secondary to these tumors is minimal to nonexistent.[6,7] Contamination due to inadequate surgery influences success rates of the secondary operation and primary surgical operation is an important factor for prognosis.[1,11]

Most lymphangiomas are benign resulting from

malformations of the lymphatic system lesions that result only in a soft, slow-growing, "doughy" mass. Lymphangiomas are rare, accounting for 4% of all vascular tumors in children.[9] Although lymphangioma can become evident at any age, 50% are seen at birth,[8] and 90% of lymphangiomas are evident by 2 years of age.[8] Lymphangiomas have traditionally been classified into three subtypes: capillary and cavernous lymphangiomas and cystic hygromas. This classification is based on their microscopic characteristics. A fourth subtype, the hemangiolymphangioma is also recognized.[8] Lymphangiomas may also be classified into microcystic, macrocystic, and mixed subtypes, according to the size of their cysts.[8] The lesions will grow and increase to a larger size if they are not completely removed in surgery.[9] Since they have no chance of becoming malignant, lymphangiomas are usually treated for cosmetic reasons only. Treatment includes aspiration, surgical excision, laser and radiofrequency ablation, and sclerotherapy.Radiotherapy and chemical cauteries are not as effective with the lymphangioma as they are with the hemangioma.[10]

Conclusion:

Previous reports even large studies discussing the prognosis of aggressive fibromatosis have included tumors from intra- and extra-abdominal sites as well as incomplete resection[14]. Regardless of primary or recurrent disease, microscopically negative margins should always be the goal for extra-abdominal fibromatosis surgery, if no cosmetic defects or function demolition is encountered. Extra-abdominal fibromatosis deserve more attention and should be treated more aggressively, especially when leaving positive margins.[14] The role of adjuvant radiotherapy has been a matter of ongoing debate in studies addressing this issue. There are data suggesting that postoperative irradiation is beneficial in disease control, whereas some have not been able to show any benefit. A comprehensive evaluation on radiotherapy by Nuyttens et al have shown significantly increased local control with adjuvant irradiation after surgery.[12] Postoperative irradiation may provide benefit in patients with microscopic or macroscopic residual tumors, as well as relapse disease.

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