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MANAGEMENT OF A RECURRENT GIANT LIPOSARCOMA IN THE UPPER BACK : A CASE REPORT

General Surgery

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ABSTRACT

Myxoid liposarcoma is the second most common soft tissue sarcoma in adults, which usually involves lower extremities with a predilection for men. Till date, only a few cases of large myxoid liposarcoma have been reported. We report a case of huge recurrent mass of size 50cmx 30cmx 8 cm over the right scapular region with extension to the neck, shoulder and infra clavicular area on the same side in a 70 years male; which developed from a preexisting lipoma operated twice and was gradually increased in size and local extension to adjacent areas. He was treated with wide local excision under general anesthesia and after follow up for one year he is free of recurrence.

KEYWORDS

Large recurrent liposarcoma, Scapular region ulceration, Wide local excision.

INTRODUCTION

Of all malignant neoplasms, soft tissue sarcoma accounts for about 1% of the mesenchymal tissue origin. Liposarcoma is the most common soft tissue sarcoma accounting for about 20% and roughly 30% of liposarcomas are myxoid variety. It is more common in men and peaks in the fourth and fifth decades of life[1,2]. These originate from the precursor lipoblasts of adipocytes, or fat cells, found in adipose tissues [3]. Mostly they occur de novo and develop frequently from deep seated stroma rather than the submucosal or subcutaneous fat. The most common site of occurance of liposarcoma is the lower limbs, trunk, retroperitoneum and the abdominal cavity. The World Health Organisation in 2020 reclassified liposarcomas into five major subtypes as:

1. Well-differentiated liposarcoma (WDL)/atypical lipomatous tumor (ALT)
2. Dedifferentiated liposarcoma (DDL)
3. Myxoid liposarcoma
4. Pleomorphic liposarcoma
5. Myxoid pleomorphic liposarcoma.[4]

While liposarcoma forms are classified as being aggressive and malignant, or, in the case of the atypical lipomatous tumor/well-differentiated liposarcoma, as relatively non-aggressive and benign , All five liposarcoma forms have the ability to invade nearby tissues and organs, occur in surgically inaccessible sites next to vital organs (such as the retroperitoneum), recur after surgical removal, and progress to life-threatening diseases [5].

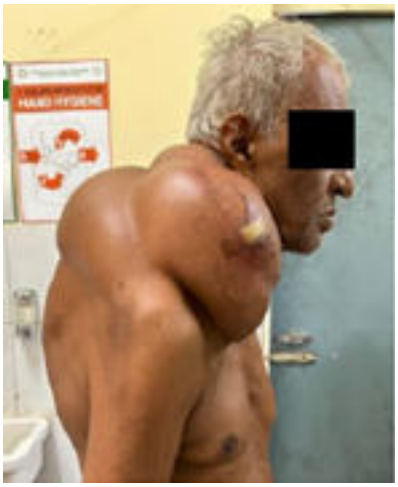

Fig 1 : Recurrent giant liposarcoma in the upper back

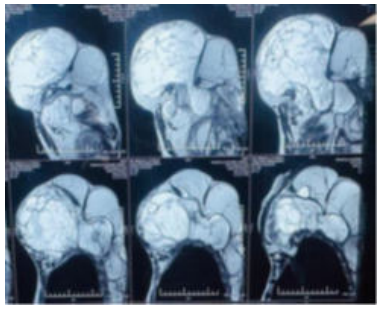

Fig 2 : MRI showing large heterogenous mass in right side of chest wall and neck showing T1 isointense and T2 hyperintense with central multiseptate component with peripheral hyperintensity.

Fig 3 : Completely excised tissue

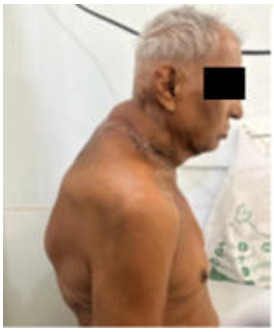

Fig 4 : Postoperative Day 8 picture.

On examination, a large multiple swelling approximately 50cm x 30cm x 8 cm extending over right anterior chest , shoulder, neck and upper back with smooth bossylated surface, well defined edges, non pulsatile, no wasting of distal muscles extending. The swelling extended anteriorly upto 4cm below the clavicle, posteriorly occupying shoulder region and scapula till 10 cm below the tip of right scapula, medially adjoining with neck till margin of right mandible, laterally till right shoulder joint without axillary involvement. Skin over the swelling was stretched with dilated veins. Mass effect causing tilting of neck towards left side and trachea deviated to left side.

The swelling is of variable consistency, no fluctuation, non-translucent, non-pulsatile, non-compressible, immobile, no fixity to overlying skin, restricted neck movements, restricted abduction of right upper limb. His routine biochemical and serological investigations were within normal limits. Recent FNAC suggested features of myxoid liposarcoma, USG Neck showed heterogenous hypoechoic lesion with multiple septations , without any vascularity and calcification. X-ray chest and CECT whole abdomen were normal. CECT neck and thorax revealed a large ill defined mass in right side of thorax and neck with mass effect and bony erosion of right clavicle. No evidence of pulmonary nodules or enlarged lymph node in the chest. MRI Neck revealing showing large heterogenous mass in right side of chest wall and neck showing T1 isointense and T2 hyperintense with central multiseptate component with peripheral hyperintensity.

Subclavian vessels coursing through without any obvious encasement. Patient underwent wide local excision of the entire lesion under G.A. with prior consent for incomplete removal, removal of supraclavicular nerves, spinal accessory nerve, possibility of postoperative scapular instability, shoulder dysfunction in form of restricted abduction, IR and ER and the need for localised radiotherapy and chemotherapy.

Intraoperative findings include a large heterogenous, multiloculated lipomatous soft tissue mass adherent to scapula, neck, upper back and thorax muscles without involvement of major vessels. As the tumor encased spinal accessory nerve it was sacrificed. A negative suction drain was kept. Dressing and drain were removed and patient was discharged on POD 5. He was advised absolute bed rest for 3 weeks which meant no scapular movement. He then got reviewed in OPD after 3 weeks and found to develop a seroma which was aspirated out and yielded about 600 ml of clear fluid. He had no reaccumulation and the suture healed normally. The histopathology revealed as myxoid liposarcoma, FNCLCC Grade : 2 (Differentiation -2 , Mitosis -1 , Necrosis – 1), no lymphovascular Invasion and a pathological staging of pT4 pNx pMx.

DISCUSSION

Myxoid liposarcoma is the second most common subtype of intermediate form of liposarcoma accounting for upto 20% . Frequently present as slowly progressive, painless swelling in the lower limbs (75-80%), especially involving thigh in 2/3rd of cases. These lesions are usually located in the intramuscular fascia, under or on the muscle [1,5]. Less frequent sites include deep-seated soft tissues of the arms (5% of cases), lower legs (10–15% of cases), retroperitoneum (8%). They rarely occur in the back [6]. This subtype on microscope shows of round cells which when accounts for >5% , is considered high grade and runs a poorer prognosis [7]. Further it contains a TLS-CHOP fusion gene on cytogenetic analysis in upto 95% of cases as a result of t(12;16)(q13;p11) translocation [8]. These Myxoid or round cell liposarcoma can metastatize the lung, skeletal bone, or other extremity and/or other soft tissue including mediastinum and retroperitoneum [9]. Thus in the diagnosis and extent of the lesion including the metastatic work up in such cases patient undergoes imaging tests, like tissue biopsy, X-ray, CT scan, and/or MRI, to check for bone metastases at the time of presentation. Wide local excision with negative margins is the standard of therapy for patients with local disease. Myxoid liposarcoma is relatively more radiosensitive subtype [10]. Radiotherapy and chemotherapy are added to patients with metastases or that are inoperable. Neoadjuvant standard regimen with anthracycline + ifosfamide (AI) or equally potent histology-tailored Trabectedin regimen can be given in selected localized high-risk myxoid liposarcomas of the extremities or trunk [11]. 5 year follow up studies have shown a local disease recurrence rate of approximately 8% [7].

CONCLUSIONS

Recurrent large swelling with previous histopathology report of lipoma, is a challenging issue in properly diagnosing any missing link, in order to institute the best possible treatment plan and avoid further recurrence. Myxoid liposarcoma should be kept as a possible differential diagnosis in large lesions over the back and thoroughly worked up preoperatively. Wide local excision is accepted as the standard of care for local disease with negative margins. However neo-adjuvant radiotherapy and adjuvant radiotherapy and chemotherapy is offered based on the case scenario as discussed to optimise adequate surgical excision, to prevent recurrence and in metastatic disease.

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