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Original Research Paper

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INGUINAL LIPOSARCOMA : A RARE CASE

B. Ananda Rama Rao* Professor of surgery SVS Medical College, Mahabubnagar, Telangana, India. *Corresponding Author

K. Gayathri Resident of surgery SVS Medical College, Mahabubnagar, Telangana, India.

ABSTRACT A liposarcoma is a bulky yellow malignant tumour. This tumour often occurs in thigh, legs, behind the knee, or in the abdomen, but it can be found in other parts of the body, in the retroperitoneum; and less often, in the head and neck area. We report a case of a 50 year old male with swelling over left inguinal region since three years, diagnosed as liposarcoma with the help of imaging studies and histopathological examination. After confirmation of the diagnosis, enucleation was done. The use of chemotherapy in liposarcoma remains experimental. The case is reported because of its rarity.

KEYWORDS : Liposarcoma; Retroperitoneal tumour, Lipoma

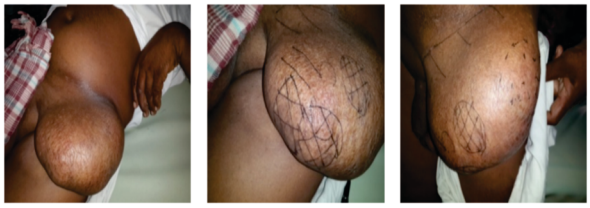
INTRODUCTION:

Liposarcoma is a rare cancer of connective tissues that resemble fat cells under a microscope. It accounts for up to 18% of all soft tissue sarcomas, which is difficult to differentiate from a lipoma. Lipoma is the most common benign tumour of the inguinal region. The lipoma is seen as a hyperechoic mass on sonography, which may be difficult to differentiate from a liposarcoma. [1,2] The World Health Organization classification of soft tissue tumors recognizes 5 types of liposarcomas: 1.Well differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes;2. dedifferentiated; 3.myxoid; 4.round cell; and 5.pleomorphic. A liposarcoma is a bulky yellow tumor similar to a lipoma but generally more complex and contains areas of prominent sclerosis.[4] Commonest sites involved by dedifferentiated liposarcoma are retro-peritoneum, extremities, gluteal region, trunk, scrotum/spermatic cord, and also subcutis. The sonographic findings of a liposarcoma are variable and non-specific. Early diagnosis and complete resection plays key role in the treatment of liposarcoma. There are no metastases and the overall prognosis is good. Different studies showed that the majority of dedifferentiated liposarcomas presented as denovo lesions, whereas the remainder developed as a late complication of a pre-existing well-differentiated liposarcoma.[5,6] Although surgical resection is the mainstay of curative treatment, patients with large high-grade liposarcomas may benefit from multimodality treatment with chemotherapy and radiation.

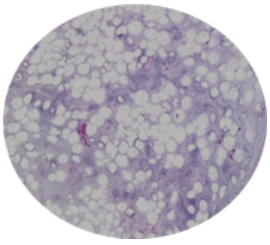
CASE REPORT:

A 50 year old male Anjalaiah reported to the department of surgery with swelling over left inguinal region since 3 years. Initially the swelling was smaller in size, but gradually increased in size over a period of one year and thereafter the size of the swelling (fig.1) remained constant. He denied any recent trauma to that area. There was no history of pain. On local examination, a single swelling of about 20*15cm was noted in left inguinal region. The scrotum and penis were normal. The swelling was soft in consistency in few areas and firm, hard and calcified in few areas. Swelling was freely mobile without any fixation to the underlying structures. The physical examination revealed no discernable loss of motor or sensory lower extremity function. There were no specific abnormalities in the laboratory data, and the tumor markers were within normal limits.

(Fig.1) Swelling over left inguinal region showing variable consistency

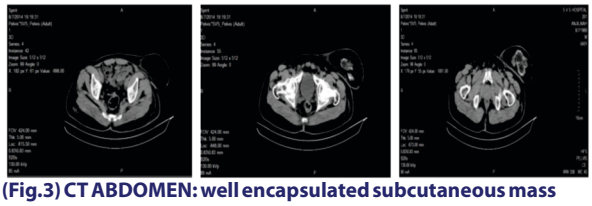


The swelling was diagnosed as Lipoma and histopathological examination was performed of the inguinal mass. FNAC of the mass revealed a tumor mass composed of well-differentiated adipocytes. Myxoid changes were noted. Lacks spindle cell component. Prominent chicken wire vascular pattern was noted. No Lipoblasts were noted. No dense collagen bundles. No areas of haemorrhage or necrosis were seen. FNAC of swelling was diagnosed as benign lipoma (fig.2)



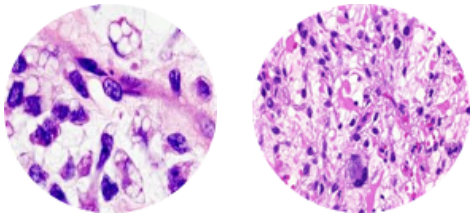
(Fig.2) FNAC: Myxoid changes in lipoma- benign

Ultrasonography of the swelling was done and it showed well defined hypoechoic mass with minimal internal flow. CT Abdomen suggestive of capsulated subcutaneous mass (fig.3).



(Fig.3) CT ABDOMEN: well encapsulated subcutaneous mass

After the cytology reporting as Lipoma from inguinal region, enucleation of tumour was done.

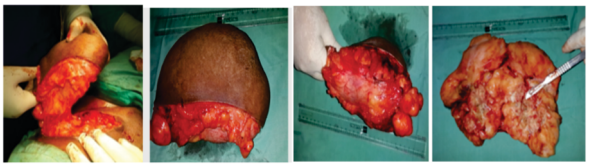


(Fig.4) H&E stained sections showing Myxoid & round cell liposarcoma - low grade (Resection margins free). Occasional large multi-nucleated lipoblasts.

Histopathologic examination of specimen revealed myxoid and round cell well-differentiated low grade liposarcoma.

Resection margins were clear. The post-operative course of the patient was uneventful. A periodical follow-up was performed every 3 months and no evidence of recurrence or metastasis was seen for 6

months after his operation, without any postoperative adjuvant therapy. The case is reported because of its rarity.



(A) (B) (C) (D)

(Fig.5)-B: Enucleated specimen, D: Cross section of mass.

DISCUSSION:

Liposarcoma is a rare mixed histologic subtype defined by the association of well-differentiated liposarcoma and a non-lipogenic sarcoma of variable histological grade usually with histologically abrupt transition.[7] The World Health Organization classification of soft tissue tumors recognizes 5 types of liposarcomas: 1.Well differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes; 2.dedifferentiated; 3.myxoid; 4.round cell; and 5.pleomorphic. Myxoid round cell liposarcoma (MRCL) is the second most common variant at around 20% of all lipogenic sarcomas. These tumors may have a hypercellular round cell component that portends a worse prognosis. It is suggested that round cell components above 25% indicate a high-grade neoplasm; however, there have been reports confirming a lower threshold of 5% as the cut off for high-grade tumors. Additionally, MRCLs have demonstrated a unique metastatic pattern with a propensity for fat-bearing areas such as bone marrow, mediastinum, retroperito neum, etc. Patients with MRCL, 17% developed skeletal metastases with the most common sites being the spine and ribs. Non-skeletal sites included the lungs, abdomen, and retroperitoneum. It is, therefore, important to mention that restaging considerations for these patients include abdominal and pelvic imaging, as well as evaluation of the spine by magnetic resonance imaging for metastatic lesions, in addition to the more common routine local and pulmonary surveillance. The analysis of the excision margins is critical; excision margins that exceed one centimeter might be considered as negative; margins under this value are considered as " suspicious " or positive (intra-tumors).[11]

Other differential diagnosis include epidermoid cyst. An epidermoid cyst develops from a remnant of ectodermal tissues misplaced during embryogenesis and often has a thin wall lined by stratified squamous epithelium surrounding a mixture of desquamated debris, cholesterol, keratin and water. On sonography, it usually appears as a hypoechoic mass with internal echogenicity, which can be explained by keratin materials within the mass.[3] Other benign tumors include leiomyomas, dermoid cysts and lymphangiomas.

The behaviour of dedifferentiated liposarcoma as a whole is that of a high-grade sarcoma.[8] Good prognosis in de novo dedifferentiated liposarcomas seems unrelated to the extent, grade, or morphologic pattern of dedifferentiation. However, high mitotic activity in the dedifferentiated component was associated with more aggressive clinical course.[9] The prognosis of liposarcomas with de-differentiated component of entirely low grade was more similar to traditional liposarcoma than to that of well-differentiated liposarcoma.[10]

CONCLUSION:

The prognosis of MRCLS is good for patients with low grade myxoid liposarcoma (defined as pure myxoid or less than a 5% round cell component), the 5-year survival rate is 92%. A significant (5% or greater) round cell component is associated with a much poorer prognosis, with a 5-year survival rate of 74%. The case is rare and hence reported.

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