Case Report

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Anterior abdominal wall myxoid liposarcoma: a rare presentation

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ABSTRACT

Liposarcoma is the most common type of soft tissue sarcoma, a mesenchymal malignancy. The extremities are the most common site followed by the retroperitoneum for liposarcomas. There are 5 histologic types of liposarcoma, as follows: well differentiated; myxoid; round cell; pleomorphic; and dedifferentiated. As very few cases of MLS have been reported, we report a rare case of an MLS of a 72-year-old male who presented with spherical mass in the abdomen wall. Computed tomography (CT) found a tumor in abdominal wall extending with both intra and extra abdominal components. There were no other abnormal symptoms and the laboratory testing was also unusual. The tumor was successfully excised, which was diagnosed MLSs in pathology. Following standard principles, after complete excision, the patient was referred to a higher center for radiotherapy. The patient was followed up for 6 month and no disease recurrence was identified. MLSs are rarely seen in health centers, irrespective of the presenting signs or histologic features.

Keywords: Liposarcoma, Myxoid liposarcoma, Soft tissue sarcomas

INTRODUCTION

Soft tissue sarcomas (STSs) are rare mesenchymal tumours, which originate in non-epithelial connective tissue sources and among them liposarcomas are the most common STS. The incidence of liposarcoma peaks between 40 and 60 years of age. Soft tissue sarcomas correspond to 1% of malignant tumours in adults with occurrence on the abdominal wall considered rare as they correspond to 0.1% of malignant tumours.² This group of neoplasms originates in mesenchymal cells of various sites: adipocytes, muscles, blood vessels, cartilage, nerves and bones.

The potential for destruction and recurrence of the tumour depends on the histological type and degree of mitoses, and immunohistochemical analysis is important particularly to define the source of the tumour and prognostic for the patients.³ More than 95%

myxoid/round cell liposarcomas have either the characteristic t(12;16) (q13; p11) or t(12;22) (q13;q12) chromosomal translocation that results in the fusion of TLS-CHOP or EWS-CHOP.4 Diagnosis remains challenging due to the rarity of this lesion and the nonspecific nature of its symptomatology.

This is a report containing a description of a case of abdominal wall sarcoma.

CASE REPORT

A male aged 72 years, presented with a painless large palpable mass on the abdomen and having first appeared 4 years previously, without other associated symptoms and underwent surgery 8 months back for the same. Spherical mass of size 11x12cm with well-defined edges with a hypertrophied scar measuring 12.5x4.5cm present at the summit and tense glossy pigmented skin extending from the epigastrium to the right hypochondrium. The patient was a smoker and did not have a family history of any type of malignancy.

Ultrasonography (USG) revealed well-defined heteroechoic lesion measuring 9.12x 12.1x12cm seen in epigastrium and right hypochondrium seen separately from liver and pancreas most likely arising from anterior abdominal wall with minimal vascularity (Figure 1).

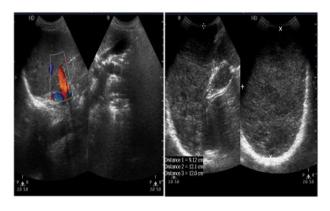


Figure 1: Well-defined heteroechoic lesion measuring 9.12x 12.1x12cm with both intra and extra abdominal components seen on ultrasonography.

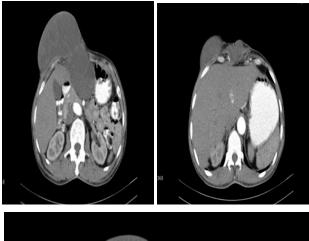




Figure 2: Computed tomography scan of abdomen showing tumor in the epigastric region with both intra and extra abdominal components.

The abdominal CT scan revealed a well-defined multi lobulated cystic lesion seen in epigastric region with both intra and extra abdominal components measuring 15x13.1x11.2 suggestive of primary abdominal wall tumour (Figure 1). The CT scan showed no evidence of metastases (Figure 2).



Figure 3: The en bloc resection with elliptical incision of all the layers of the abdominal wall over the mass and a midline laparotomy incision extending below.

The needle biopsy of the centre revealed a high-grade myxoid and lipoid material admixed with inflammatory cells. The patient underwent en bloc resection with elliptical incision of all the layers of the abdominal wall including the skin over the mass and a midline laparotomy incision extending below (Figure 3). The abdominal wall reconstruction was performed using 15 x 15 cm polypropylene mesh after suturing the entire edge of the abdominal wall defect in order to prevent the intestinal loops from adhering to the prosthesis. The mesh was fixed in place with 2.0 nonabsorbable polypropylene sutures at separate points (Figure 4). Primary closure of the skin was performed using Ethilon 2-0 along with placement of a vacuum drain. The patient progressed satisfactorily and was discharged on the 7th postoperative day.



Figure 4: Abdominal wall reconstruction performed using polypropylene mesh and the final outcome is seen.

Pathological findings

The gross section showed a yellowish tumour well circumscribed capsulated with cut surfaces which show

gelatinous myxoid, yellowish pale areas with secondary degeneration. The margins were clear of the tumour cells. The tumour was histologically entirely composed of infiltrating mesenchymal neoplasm of undifferentiated round cells, atypical spindle cells and lipoblasts scattered throughout the tumour, set in a chicken mesh matrix which is predominantly myxoid and show thin walled blood vessels arranged with focal areas of fibrillary collagen infiltrate. Immunohistochemical staining was performed and revealed the following: CK (-); vimentin (-); S-100 (-); CD34 (-); and SMA (+).

DISCUSSION

Liposarcomas are rare most common soft tissue malignant tumours of mesenchymal origin that are deep seated and accounting for 16-18% and the annual incidence is estimated at 2.5 per million and the average age at presentation is in the 50s.⁵

The patient usually presents with tumor mass of small to big size with well-defined or irregular border, pedunculated, with discoloration of skin. Myxoid liposarcoma (MLSs) are the second most common subtype of liposarcoma, accounting for 40% and (more than two thirds) occur in the lower limbs such as the thighs and popliteal area, and rarely occur in the retro peritoneum. The percentage of round cells is the strongest prognostic factor, as it negatively impacted local recurrence, metastasis and survival.

Grading of the round-cell content of tumors is based on a 5% threshold. The risk of local recurrence was 3.86 times greater if the tumor contained more than 5% round cells. They are very sensitive to radiation therapy and chemotherapy and have an unusual ability to metastasize in locations other than the lungs.⁶ Well-known negative prognostic factors are patient age (>45years), tumor size (>10cm), sub-aponeurotic location, high histological grade, large percentage of round cells and positive resection margins. Surgery is obligatory for treatment of abdominal liposarcoma. Extensive surgical removal is the utmost effective treatment and has a significant effect on the survival rate.

The crucial aim of the surgery is complete resection with negative margins. Cytogenetically, almost all myxoid liposarcoma have a specific reciprocal translocation of chromosomes 12 and 16, t(12;16) (q13;p11), resulting in an oncogenic hybrid protein, FUS-CHOP. The chimeric FUS-CHOP gene gives rise to at least three fusion transcripts, one of which (type II) has been identified in most myxoid/round cell liposarcomas. Patients with localized myxoid liposarcoma are treated with conservation surgery and radiotherapy.⁶

Spillane and associates reported an 84% 5-year local control rate for their series of 50 patients treated with surgery alone, RT alone, or a combination of surgery and RT.⁷ The two most common myxoid sarcomas of adults

that are confused with myxoid liposarcoma are myxoid malignant fibrous histiocytoma and myxoid chondrosarcoma. Overall, 5-year survival for well-differentiated subtypes is 90%, while 5-year survival for pleomorphic subtypes is only 30% to 50%. Dedifferentiated and myxoid/round cell subtypes have 5-year survival rates of 75% and 60% to 90%, respectively.⁸

CONCLUSION

Present case supports the view that myxoid liposarcoma is a puzzling tumor for diagnosis and treatment which are extremely rare in the abdominal wall. Radiological imaging preoperatively is essential for assessing the origin of the tumor, its spread, and planning surgery but final diagnosis is based on histological findings which is the gold standard. The overall 5-year survival rate for these neoplasms remains low and surgery appears to be the only possibility for effective treatment along with chemo- and radiation therapy which may improve local control and survival of the patients.

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