International Surgery Journal
Choudhary SK et al. Int Surg J. 2019 Feb;6(2):633-635
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20190421

Case Report

Low-grade myofibroblastic sarcoma of retroperitoneum: a rare case

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Received: 14 December 2018
Accepted: 08 January 2019

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ABSTRACT

Low-grade myofibroblastic sarcoma (LGMFS) is an uncommon tumor which develops mainly in the bone or soft tissues of the head and neck region, trunk, or extremities and extremely rarely found in the abdominal cavity. The rarity of the disease and its low-grade features make an accurate diagnosis difficult in most cases. We recently encountered a giant LGMFS which had developed in retroperitoneum and surgically resected with gratifying results.
An 18 years old female presented with complaint of left sided abdominal pain since 8 months and left sided abdominal lump since 5 months. Imaging examinations revealed retroperitoneal soft tissue mass, and surgical treatment was scheduled. During operation, a tumor about 20x18x15cm in diameter with its anterior aspect covered with the pancreas, mesocolon and splenic flexure of colon with densely adhered to splenic vessels and left renal vessels was found. The tumor had firm adhesions to the surrounding tissues, and it was excised with concomitant distal pancreato-splenectomy, left nephrectomy and resection of splenic flexure of colon with colo-colic anastomosis. Histopathologically, fusiform cells were arranged in a complicated or storiform pattern, and immunohistochemical staining revealed that the tumor was positive for vimentin and focally positive for α-smooth muscle actin (SMA), negative for desmin, CD34, CD117, EMA, DOG-1 and S-100. Diagnosis of LGMFS was made. During 6 months follow up patient is asymptomatic and ultrasound abdomen is normal. In case of LGMFS, favorable prognosis can be attained by complete resection of the primary lesion and regular follow up of patient by physical examination and imaging modality.

Keywords: Myofibroblastic sarcoma, Retroperitoneum, Sarcoma

INTRODUCTION

Low-grade myofibroblastic sarcoma (LGMFS) is an uncommon tumor which originates mainly in the bone or soft tissues of the head and neck region, trunk, or extremities.[1,2] The oral cavity is the most frequent site of the tumor development. The tumor is found most commonly in deep soft tissues but may also arise in the subcutis, submucosal tissue, and bone. This tumor is rarely found in the retroperitoneum and only a few cases of LGMFS in retroperitoneum were reported in literature.[1-3] The rarity of the disease and its low-grade features make an accurate diagnosis difficult in most cases.[4,5] Such tumor may be found in people of all age groups but is slightly more frequently observed among males. The natural history of LGMFS indicates very poor survival.[4] Local recurrence is the major cause of mortality for retroperitoneal sarcomas in contrast with extremity sarcomas where the principal cause of death is distant metastases. Any local recurrence should be detected as soon as possible by periodic imaging examinations. This depends on the difference in anatomic location and tumor biology.[6] They should be managed by excision with a wide margin of normal tissue and adjuvant radiation therapy or systemic chemotherapy.[7] Some patients have been treated concomitantly with radiation therapy and chemotherapy, but therapeutic effects in these patients have not been documented. Due to the difference in prognosis, a precise pathologic diagnosis is essential,

which requires a combination of thorough morphologic examination and immunohistochemistry.

Here authors discuss a case of giant retroperitoneum LGMFS, which was surgically resected with gratifying results.

CASE REPORT

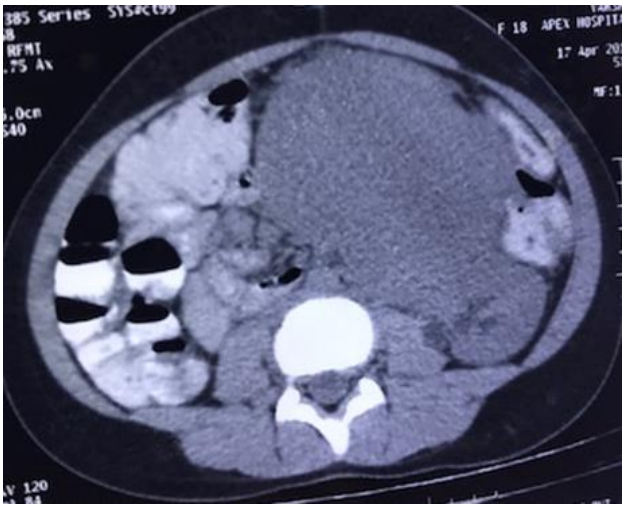
An 18 years old female patient presented with complaint of left sided abdominal pain since 8 months and left sided abdominal lump since 5 months. On physical examination, a non-tender lump was palpable in left hypochondrium and left lumbar region.

Routine hematological and biochemical tests were with in normal limit. Serum tumor markers like, Carcinoembryonic antigen (CEA) and Carbohydrate antigen (CA19-9) was normal.

Contrast enhanced computed tomography (CECT) whole abdomen showed a large approximately 17x14x10cm, well defined soft tissue density lesion in left para-aortic region. It is abutting body and tail of pancreas and displacing it anteriorly.

It is abutting upper and mid pole of left kidney and left renal vessels displacing them posteriorly. It is showing mild heterogeneous enhancement in portal and venous phase. Mass is causing compression of splenic vessels and left renal vessels.

Radiological differential diagnosis was paraganglioma or lymph nodal mass. (Figure 1). USG guided trucut biopsy of lump suggested fibroconnective tissue only. No evidence of malignancy seen. Repeat trucut biopsy suggested possibility of benign mesenchymal lesion.



The major axis of the tumor is about 14 cm.

Figure 1: Computed tomography (CT) showing a tumor in contact with the anterior surface of the pancreas.

A diagnosis of retroperitoneal giant tumor was made, and surgical resection of the tumor was scheduled. Left subcostal incision was made and extended across the midline. On exploration, a mass of approximately 20x18x15cm in size with its anterior aspect covered with the pancreas, mesocolon and splenic flexure of colon with densely adhered to splenic vessels and left renal vessels was found. The tumor had firm adhesions to the surrounding tissues, and it was excised with concomitant distal pancreato-splenectomy, left nephrectomy and resection of splenic flexure of colon with colo-colic anastomosis.

Histopathological examination reveals low grade myofibroblastic sarcoma, which was abutting the pancreas but not infiltrating it. On immunohistochemical staining, tumor was positive for vimentin and focally positive for α-smooth muscle actin (SMA), negative for desmin, CD34, CD117, EMA, DOG-1 and S-100 (Figure 2 and 3). On follow up of 6 months, patient is asymptomatic with normal ultrasound abdomen.

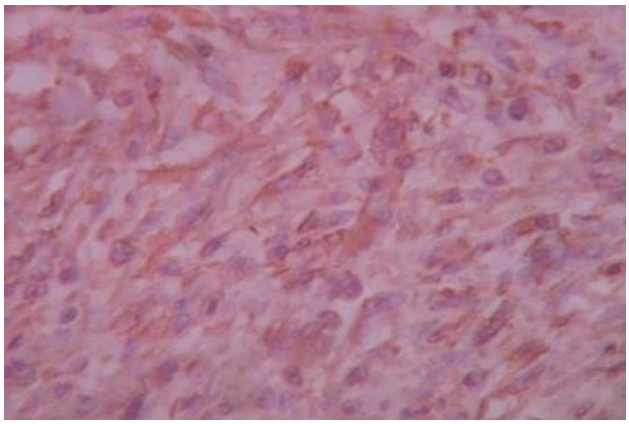


Figure 2: Positive for vimentin.

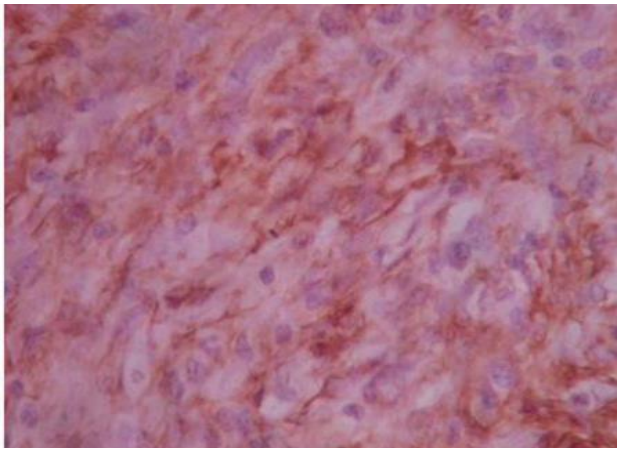


Figure 3: Positive for Smooth muscle actin (SMA).

DISCUSSION

LGMFS is a rare fusiform cell neoplastic disease that arises in the bone or soft tissues.[1,2] The oral cavity is the

most frequent site of the tumor development, followed in order by the extremities, pelvis, lung, mammary gland, salivary gland and perineum.[1,2,8-12] Development of the tumor in the retroperitoneum, as seen in our patient, is extremely rare. In the literature, there is not more than 7 cases reported.[1,2]

Indolent enlargement of the mass is a typical clinical manifestation of LGMFS, but patients may present with pyrexia, chills and leukocytosis.[8,13]

Histopathological, LGMFS is characterized by fusiform tumor cells which are arranged in complicated, sheet-like or storiform patterns and show a diffuse, infiltrative growth.[1,2,9] The cytoplasm is indiscrete and faintly eosinophilic, and the slender, undulate nucleus contains uniformly distributed chromatin and a small nucleolus. The interstitial tissue consists of collagenous fibers and is often hyalinized. Inflammatory cell infiltrates are sparse with no necrosis.[1,5,14]

On the immunohistochemical staining, this tumour is positive for α-SMA, muscle-specific actin (MSA), desmin, calponin and fibronectin, and negativity for laminin, S100β and EMA.[1,2] In the present case, tumour was positive for α-SMA and negative for S100β, H-caldesmon, CD34, β-catenin, c-KIT and EMA, which was in favour of diagnosis of LGMFS. Treatment of this disorder usually consists of surgical resection. Some patients have been treated concomitantly with radiation therapy and chemotherapy, but therapeutic effects in these patients have not been documented.[1,2] Local recurrence is the major cause of mortality for retroperitoneal sarcomas in contrast with extremity sarcomas where the principal cause of death is distant metastases. This depends on the difference in anatomic location and tumor biology.[6]

CONCLUSION

In case of LGMFS, favorable prognosis can be attained by complete resection of the primary lesion and regular follow up of patient by physical examination and imaging modality.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Choudhary SK, Gupta S, Bansal S, Kumar N. Low-grade myofibroblastic sarcoma of retroperitoneum: a rare case. Int Surg J 2019;6:633-5.