Giant Inflammatory Variant of Well-Differentiated Liposarcoma of Retroperitoneum: A Rare Case Report

Shubha P. Bhat • Kishan H. L. Prasad • Rohan Shetty •
Rajesh Ballal • Sunil Y. Kumar • Panna Hegde

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Introduction

Well differentiated liposarcoma(WDLPS), is a low grade malignant mesenchymal tumor. The inflammatory variant is a relatively rare subtype of WDLPS, accounting for 2 % of all liposarcomas[1]. Majority of these, have occurred in the retroperitoneum[2]. Cases have also been described in the pleura, scrotum, and the spermatic cord[3]. Its peak incidence is between 50 to 70 years[4]. These patients usually have no symptoms till tumor reaches large size and causes disturbances in adjacent structures, but at times they may present with pyrexia of unknown origin[5, 6]. Computed Tomography(CT) and Magnetic resonance imaging(MRI) are the most effective radiological procedures determining the extent of local and distal tumor invasion[5]. Surgical excision is the treatment of choice. Role of chemotherapy and radiotherapy is disputable[5]. Factors influencing survival rates are site, tumor size, depth, invasion of surrounding structures and completeness of surgical excision[6]. There is only one case report of giant inflammatory variant of well differentiated liposarcoma in world literature and this is second of its kind. We present a case of Giant inflammatory WDLPS in a 52-year-old male who came with left lower abdominal pain. The patient is doing well since 8 months and is on regular follow up.

Case History

A 52 year male came with complains of left lower abdominal pain of 1 month duration. The pain was insidious in onset, gradually progressive and dull aching type. There was no history of weight loss or loss of appetite. Bowel and bladder habits were normal. General physical examination was normal. Per abdomen on right lateral position, showed a vague mass in left lumbar region. Complete hemogram was normal except for erythrocyte sedimentation rate, which was 65 mm at the end of 1 hour. CT abdomen and pelvis, showed a large retroperitoneal tumor encasing and displacing the left kidney anteriorly, extending superiorly upto spleen and diaphragm, and inferiorly upto pelvis with mixed density containing fat, solid and calcified areas.(Fig. 1) CT guided fine needle aspiration cytology(FNAC) was inconclusive. Laparotomy and surgical excision of the mass lesion was done. Grossly the soft tissue mass was large, partially capsulated, bosselated and yellowish measuring 28×15×7 cm, weighing 8 kg. Cut surface was lobulated with yellowish areas and focal grey white areas.(Fig. 2) Histopathology showed scattered atypical, mononucleated and multinucleated lipoblasts. (Fig. 3) Also noted extensive lymphoplasmacytic aggregates with plasma cell rich zones, with many russel bodies.(Fig. 4) Adjacent focus showed lipoma-like areas.(Fig. 5) Occasional atypical mitosis was seen. Hence, final diagnosis of giant inflammatory variant of well-differentiated liposarcoma was made. He is on regular follow up since 8 months without any recurrence.

R. Shetty • R. Ballal
Department of General Surgery, K S Hegde Medical Academy
of Nitte University, Deralakatte,
Mangalore, Karnataka, India 575018

S. P. Bhat • K. H. L. Prasad (✉) • S. Y. Kumar • P. Hegde
Department of Pathology, K S Hegde Medical Academy of Nitte
University, Mangalore, Karnataka, India 575018
e-mail: dr_kishanpath@rediff.com

Discussion

Retroperitoneal tumors are an extremely heterogenous group of neoplasms, 85 % of which are malignant[4].

Fig. 1 CT abdomen showing large retroperitoneal tumor with mixed density containing fat, solid, and calcified areas

Liposarcomas are neoplasms derived from adipose tissue and correspond to 10–14 % of all soft tissue sarcomas[7]. WDLPS is the commonest subtype of liposarcoma, accounting for 40–45 % of all cases[3]. In 1997, a unique subtype of WDLPS, in which there was extensive lymphoplasmacytic infiltrate, was described, which was later termed as, well differentiated inflammatory liposarcoma[1, 2]. Majority of these, have been described in the retro peritoneum, with only 2 reported cases occurring in the thigh[1]. Few cases have also been described in the pleura, scrotum, and the spermatic cord[3]. Clinically these tumors present with diffuse abdominal pain, anorexia, weight loss, and increase in abdominal girth. Abdominal symptomatology is due to compression of the organs[7]. Our patient was 52-year-old male who came with left lower abdominal pain of 1-month duration.

Fig. 2 Gross showing large, partially capsulated, bosselated and yellowish soft tissue mass with lobulated cut surface

Fig. 3 Histopathology showing scattered atypical multinucleated lipoblasts amidst the inflammatory cells. [H&E, 400]

Basic abdominal imaging studies, as well as FNAC are auxiliary diagnostic tools of great utility for diagnosis and staging of these tumors. Ultrasonography is more precise for localization of the tumor. CT scanning is highly sensitive in determining the size, extension, as well as relation to neighbouring organs, in order to permit an adequate preoperative evaluation. MRI is superior to CT scan due to its capacity to differentiate the tissue types[4]. In our case, CT abdomen and pelvis, showed a large retroperitoneal tumor with mixed density containing fat, solid and calcified areas.

The WDLPS are further classified as adipocytic, sclerosing, inflammatory and spindle-cell subtypes. Inflammatory liposarcoma is an unusual variant of WDLPS. The mean size of retroperitoneal liposarcomas reported is 15.7 cm[6]. The tumor in our case was much larger (28×15×7 cm) and weighed 8 kg. There is only one case report of the Giant inflammatory liposarcoma till date. Ours is second of its

Fig. 4 Histopathology showing extensive lymphoplasmacytic aggregates with many russel bodies. [H&E, 400]

Fig. 5 Histopathology showing lipoma like areas. [H&E, 400]

kind. Key histological features includes: 1. Nodular lymphoplasmacytic aggregates, 2. Intervening paucicellular stroma containing fibroblastic elements, 3. Merging of atypical adipocytic and inflammatory elements, 4. Adjacent clearly defined zones of lipoma-like, or more rarely, sclerosing-type liposarcoma, all of which were seen in our case[8].

Wide surgical excision is the most effective procedure preventing the recurrence and dedifferentiation[5]. Laparotomy and complete excision of the mass was done in our case. Chemotherapy and radiotherapy have shown little efficacy in the treatment of retroperitoneal liposarcomas and locoregional recurrence remains the main cause of death in these patients[9]. Other factors influencing survival rates are the tumor size, depth, invasion of the surrounding structures and the completeness of surgical excision[1]. In our case, the patient did not get any adjuvant therapy because it was totally excised with free margins. The patient is doing well since 8 months without any recurrence.

Conclusion

Inflammatory liposarcoma is an unusual variant of well-differentiated liposarcoma. Awareness of this rare variant

of well-differentiated liposarcoma is necessary. Wide surgical excision is the treatment. Knowledge of the pathological features and clinicopathological correlation will enable accurate diagnosis. These patients require regular postoperative radiological follow-up to identify recurrences and increase the survival.

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Conflict of interest Nil

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