

Giant Dumbbell Liposarcoma Thigh Extending to the Retroperitoneum - The Ideal Surgical Approach and Reconstruction - A Case Report and Review of Literature

Dr. Bharath V M¹, Dr. Syam Vikram², Dr. Subin Sugath³, Dr. Nanda Kachare⁴

Department of Surgical Oncology, Centre of Excellence - 1, Aster Medcity, South Chittoor PO, Cheranelloor, Kochi 682027

Abstract: *Dumb bell shaped tumours have been reported infrequently in literature - of which the usual ones being retroperitoneal tumours extending to the scrotum via the inguinal canal or extending through the femoral canal and presenting in the thigh. Cases of a thigh liposarcoma extending to the retroperitoneum are very rare. We could come across only publications in this regard⁵. We report a patient with a large liposarcoma of the thigh extending to the retroperitoneum via the femoral canal. Oncologically sound resection continues to be a challenge in large retroperitoneal tumours with dumb bell extension as in other retroperitoneal tumours due to the large size at presentation. The thoroughness and integrity of the first excision is vital to determine its curative effect². The approach for such tumours and reconstruction post resection should be an essential part of planning. We present our experience of the combined en bloc excision of the dumb bell tumour and reconstruction along with the review of the available literature.*

Keywords: Dumb bell tumour, abdomino- inguinal approach, mesh repair of inguinal ligament, atypical lipomatous tumour

1. Case Report

Our patient is a 47 yr old Lady, a breast cancer survivor since 2000. She had noticed a small swelling in the left thigh mid aspect then and was seen by a surgeon and was clinically suspected to be a lipoma. As she was asymptomatic she was not evaluated further and was kept under follow up. There was a gradual increase in size of the swelling over the time. She had rapid increase in size of the for the past one and half years for which she was evaluated with an MRI thigh in April 2016. It showed a 18.8 x 13.6 x 19 cm swelling in the intermuscular plane with cranial extension suggestive of a liposarcoma. Biopsy showed features of atypical lipomatous tumour. A CT scan of the abdomen showed a large retroperitoneal extension of the soft tissue tumour in the thigh through the femoral canal along the vessels. There were no metastasis.

On clinical evaluation. Her gait is normal. Left thigh shows a 20 x 15 cm large soft to firm mass arising from the anterior compartment of left thigh completely filling the upper half of the anterior compartment. Upper limit of the swelling was felt at the inguinal ligament, lower limit is up to the lower mid thigh. There are dilated veins. There is no distal neurovascular deficit. There was no limb edema. Per abdomen examination showed a well defined mass lesion in the left iliac fossa, lower limit is up to the inguinal ligament. There is no cross fluctuation.

2. Operation Notes

Patient was positioned supine and the incision was planned as an abdomino inguinal approach - Large dumb bell shaped tumour - 15 x 10 cm in the left side of retroperitoneum pushing the colon, left ureter and the iliac vessels medially and posteriorly the mass was densely adherent to the ilio psoas muscle, it was coursing along the femoral vessels

through the femoral canal and entering the thigh. A 25 x 20 cm tumour in the left thigh extending to the mid thigh adherent to the femoral vessels with no obvious infiltration. It was dipping on to the level of lesser trochanter at a single point. It was a well encapsulated tumour with dilated vessels. The femoral nerve was found coursing through the tumour. Femoral vein and artery found to be closely related to the tumour with no features of encasement or infiltration.

Laparotomy was done - colon, ureter and iliac vessels dissected free off the tumour. Posteriorly the tumour was dissected along with the cuff of muscle. Inguinal ligament resected in toto with the tumour in view of dense adhesion and the thigh component of the tumour dissected free. Femoral nerve was encased by the tumour and hence it was sacrificed. Femoral vessels were carefully dissected free off the tumour. Inguinal ligament reconstructed with a prolene mesh. Rectus abdominis sutured back in place and an onlay mesh was kept.



Figure 1: Pre op picture of tumour



Figure 2: Pre op incision marking



Figure 5: Reconstruction with prolene mesh



Figure 3: Retro peritoneal tumour : Dissecting the tumour off the ureter



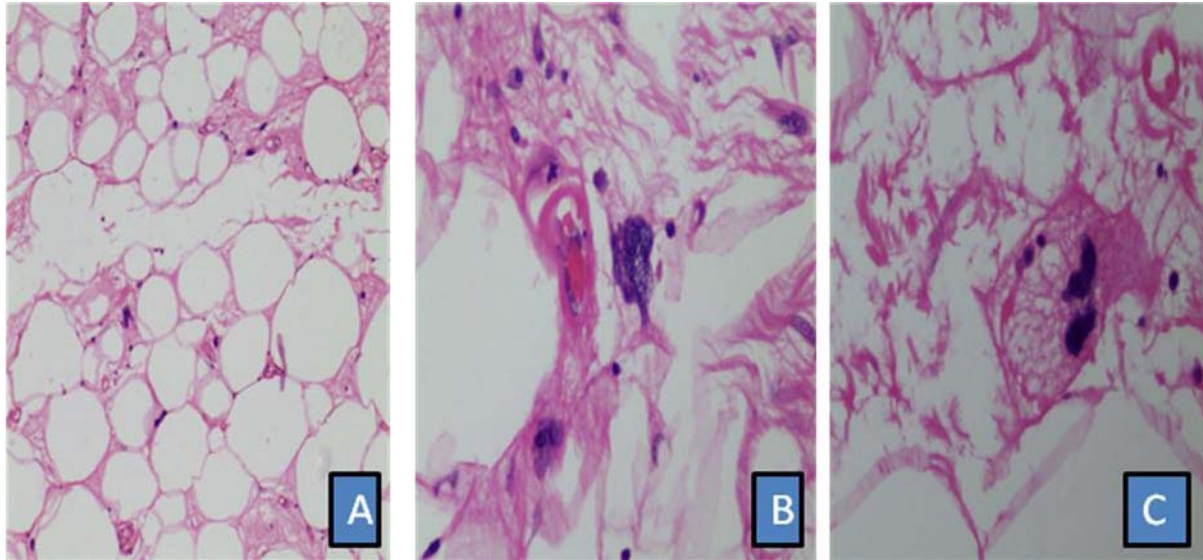
Figure 6: Pathological specimen



Figure 4: Thigh part of tumour through inguinal ligament



Figure 7: Histopathological slide



Histopathology showed predominantly lipoma-like areas with lobules of mature adipocytes with eccentrically placed nuclei and vacuolated cytoplasm. There were no dedifferentiated areas identified in multiple sections examined. Based on the features of lipoma like areas with lipoblasts and atypical stromal cells, the diagnosis of lipoma-like liposarcoma/well differentiated liposarcoma was established.

Post surgery she has weakness on knee extension. She was ambulated early with knee brace which she tolerated well. She has been placed on physical rehabilitation. She has been doing well, considering the well differentiated tumour and a complete excision, it was decided in the multi disciplinary tumour board to keep her under follow up.

3. Review of Literature

Liposarcoma of the thigh extending to the retroperitoneum is quite rare. To the best of our knowledge there has been one published case series from Malaysia.⁵ The approach to these tumours is also not straight forward as many surgeons may not be familiar with the exposure and the reconstruction technique involved. Hence we think that a description of our case along with the review on surgical approach would bring out more clarity on how to deal with such tumours.

There have been a few important changes in the soft tissue tumour classification. The WHO classification of soft tissue tumors 4th edition (2013) divide adipocytic tumours broadly in to benign, intermediate and malignant. Atypical lipomatous tumour / well differentiated liposarcoma is classified under intermediate (locally aggressive) category. Malignant tumours are De differentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma and liposarcoma not otherwise specified. The current classification no longer includes mixed type liposarcoma. Atypical lipomatous tumour (ALT) is divided in to 3 main sub groups(it was 4 previously) 1. Adipocytic (lipoma-like) 2. Sclerosing 3. Inflammatory¹. Well-differentiated (low-grade) liposarcomas are the most common, followed by dedifferentiated liposarcomas. These tumors have no potential to metastasize; as such, they are referred to as atypical lipomatous tumors when they arise in the extremity

or body wall/trunk. However, given their propensity for local recurrence in the retroperitoneum/mediastinum and spermatic cord, these same tumors are referred to as well-differentiated liposarcomas in these locations.^{2,4}

The abdomino inguinal incision allows a vast improvement in the exposure and resectability of tumors in the lower abdomen with fixation to the pelvic side wall or tumours with extension to the thigh. A midline, paramedian, or oblique abdominal incision often does not provide adequate exposure for these tumors. A lower midline incision provides good exposure of the intrapelvic structures. An inguinal incision exposes the femoral vessels. A transverse incision connecting the two extending up the mid inguinal point followed by the vertical incision, by dividing the origin of the rectus abdominis from the pubic crest and the insertion of the inguinal ligament to the pubic tubercle, provides the necessary link that allows a single incision field and optimizes exposure.¹

The difficulty arises like in our case especially with tumors in the lower parts of the pelvis where the anterior abdominal wall converges with the retroperitoneal structures (e.g. iliopsoas muscle, iliac vessels). The reconstruction following the resection is fairly straight forward. Lateral to the vessels the inguinal ligament is approximated to the iliac fascia and medial to the vessels to Cooper's ligament. The rectus sheath and muscle are approximated to their remnants on the pubic crest¹. When a defect in the fascia has been created, it may be covered with a plastic mesh, which also replaces the inguinal ligament. The mesh should not be in direct contact with the vessels.

In our case the tumour was densely adherent to the inguinal ligament and had to be removed in toto with the specimen. We had re created the inguinal ligament by folding it and suturing the ends to the pubic tubercle and to the anterior superior iliac spine and had done an onlay mesh repair over the sutured rectus abdominis. We had carefully sutured the mesh around and re created the femoral canal.

The indications for the abdominoinguinal incision are: (1) abdominal or pelvic tumors extending over the iliac vessels, (2) tumors in the iliac fossa, (3) primary tumors, possibly

involving the iliac vessels or large iliac lymph node metastases, (4) tumors with fixation to the wall of the true pelvis or large obturator nodes, (5) tumors involving the pubic bone with or without extension to the pelvis or adductor group of muscles, and (6) tumors of the groin when they involve the vessels of the lower abdominal wall or extend in the retroperitoneal area.¹

To date, no randomized trial have been completed comparing surgery with or without RT. Postoperative RT is mainly performed in high grade tumors and close or positive resection margins. Histologic differentiation is one of the most important determinant of clinical outcome in liposarcoma patients. De-differentiated subtype has poor outcome compared to well-differentiated but still preferentially recurs in a local pattern, it can be inferred that de-differentiated RPLS might be the subtype that most benefits from adjuvant RT.⁶ Chemotherapy has not consistently shown to have any disease-free survival benefit. However certain histological subtypes have shown response to different chemotherapy agents.⁷

4. Conclusion

Adequate Surgical resection remains key to achieving long term cure. Dedicated Multi disciplinary tumour board discussions to optimise patient care is imperative to achieve optimal results. Surgical resection involves proper planning of the incision for adequate exposure and also meticulous reconstruction. Post operative rehabilitation should be done rigorously to achieve optimal functional status.

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