A Recurrent Myxoid Liposarcoma with Right Inguinoscrotal Extension – A Case Report*

Jan Michael James Lazaro, MD

ABSTRACT

Liposarcoma is among the most common soft-tissue sarcomas in adults. Retroperitoneal liposarcomas are not usually detected early because of their location hence diagnosed at an advanced stage and when they are, most are usually enormous in size due to the nature of the tumor. They can present in a multitude of ways depending on the size and subtype of liposarcoma. Myxoid retroperitoneal liposarcomas are highly locally recurrent with little to no metastatic potential and such, may present with inguinal herniation. Cases which present in such a way are rare, as such was logged in a study by Rhu et al. Six out of 168 patients with diagnosed retroperitoneal liposarcoma had inguinoscrotal extension. And this scarcity in cases brings out the need for discussion of betterment of future treatment.

Here is a case of a recurrent retroperitoneal myxoid liposarcoma with recurrent right inguinal hernia secondary to mass extension. A 70 year old male was re-admitted due to abdominal enlargement with concurrent right inguinioscrotal mass.The patient was a known case of recurrent retroperitoneal liposarcoma having undergone exploratory laparotomy with retroperitoneal mass excision. Presently, the patient presented with a globular distended abdomen as well as an enlarged scrotal mass. Chest and abdominopelvic CT scan with IV contrast were done. Patient's current case was deemed inoperable as well as high morbidity risk for neoadjuvant treatment due to size and extent hence surgery was put off and the patient was referred to palliative care for management until further notice.

Diagnosis and prognosis of such recurrent retroperitoneal liposarcomas need the use of a combination of histopathology and specific imaging modalities to properly delineate tumor tissue margins since surgical resection is by far the gold standard therapy for most liposarcomas. Due to high recurrence rates, it is highly imperative for aggressive surveillance and complete tumor excision.

Key words: Recurrent myxoid liposarcoma, inguinoscrotal extension

Liposarcoma is by far one of the most common soft-tissue sarcomas in adulthood (approximately 15% of all soft tissue sarcomas).[1,2] It presents more often in the extremities (41%), retroperitoneum (19%). and inguinal region (12%).[5,6] According to the WHO classification of retroperitoneal liposarcomas, there are four subtypes: well-differentiated, dedifferentiated, myxoid, and pleomorphic.[5,12]

Myxoid liposarcomas account for 5% of adult soft tissue sarcomas and 15-20% of all liposarcomas.[1] They (as well as well-differentiated) are minimally metastatic but highly recurrent with around 40% of all surgically-treated patients experiencing relapse.[3] The treatment protocol for soft tissue sarcomas will depend on primary location but more importantly, on the histopathologic pattern of the sarcoma and may range from surgical resection to radiation and/or combination chemotherapy.[2,4] At the latest, complete surgical resection is the most appropriate treatment while adjuvant therapy requires further study to evaluate.[3]

Retroperitoneal liposarcomas should be managed with a balance of aggression and caution given

* From the Department of Radiology

that surgical treatment is the mainstay treatment. Sufficient surgical margins should be achieved by the first operation to make sure of lower recurrence rate which leads us to the importance of proper imaging pre-operatively.

There have been circulating articles of retroperitoneal liposarcomas with presentation of inguinal hernias as well as inguinal liposarcomas mimicking hernias but there is limited specific study on retroperitoneal myxoid liposarcomas with consequential extension into the inguinal canal as hernia.

The objective of this report was to present a rare case of recurrent retroperitoneal myxoid liposarcoma with concurrent right inguinal hernia secondary to mass extension.

THE CASE

A 70 year old male came in for admission with chief complaint of abdominal enlargement with concurrent right inguinoscrotal mass.

The patient was a known case of retroperitoneal liposarcoma having undergone exploratory laparotomy with excision of retroperitoneal mass twice, first in 2016 with appendectomy then in 2018 due to mass recurrence with adhesiolysis as well as right inguinal herniorrhaphy secondary to mass extension into the right inguinal canal. Patient also underwent 1 cycle of chemotherapy (Doxorubicin and isofosfamide) in 2016 but was also lost to follow-up.

Few hours prior to admission, the patient's cumulative experience with difficulty of breathing, intermittent constipation, difficulty in micturition and bilateral edema of both lower extremities prompted consultation and was then subsequently admitted.

Physical examination on admission showed a globular distended abdomen with a circumference of 113cm, visible dilated umbilical vessels, dull on percussion, with firm, non- movable, non-tender mass on palpation as well as an enlarged right scrotal mass with circumference of 75 cm. Chest radiograph (Figure 1) upon admission showed a homogenous opacity occupying most of the right hemithorax obscuring the right hemidiaphragm and cardiac border.

Abdominopelvic CT scan with intravenous contrast (Figures 2A-F) was done which showed multiple varisized complex abdominal masses causing compression and displacement of adjacent structures with right pelvic herniation of the aforementioned abdominal mass towards the right scrotal compartment. Multiple enlarged mesenteric lymph nodes were also seen as well as bilateral moderate pelvocaliureterectasia secondary to compression of the distal ureters. Contrast enhanced chest CT scan was also performed with visualization of bi-apical fibrotic residuals and right lung compression secondary to upward liver displacement compressed by known primary.

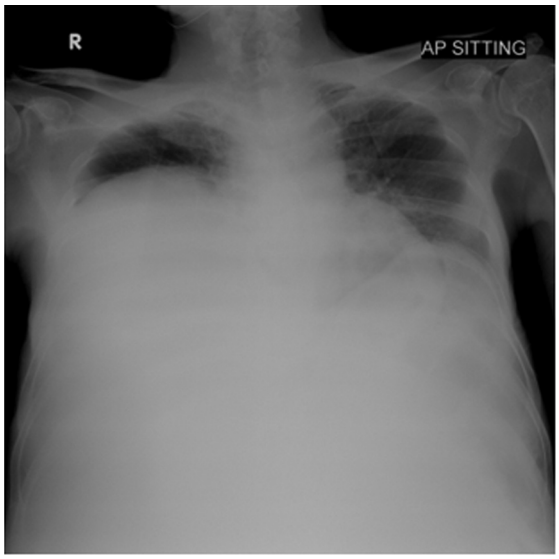

Figure 1. Chest radiograph showed a homogenous opacity occupying most of the right hemithorax obscuring the right hemidiaphragm and cardiac border. Consider very elevated right hemidiaphragm due to eventration. Other differentials are mass lesion and pleural effusion. Suggest CT scan correlation.

Patient was prepared for emergent surgical excision of the retroperitoneal mass and hernia repair. Patient was referred to the oncology, cardiology, pulmonary, anesthesiology and general medicine departments for pre-operative clearance.

After collaborative pre-operative deliberation from several departments, the state of the patient's current case was deemed inoperable as well as unfit for neoadjuvant treatment hence the patient was then referred to palliative care for pain management.

DISCUSSION

Around 40% of all liposarcomas are found in the retroperitoneum and are confined within it. However, there is direct connection of the inguinal canal with the retroperitoneum thus giving rise to the chance

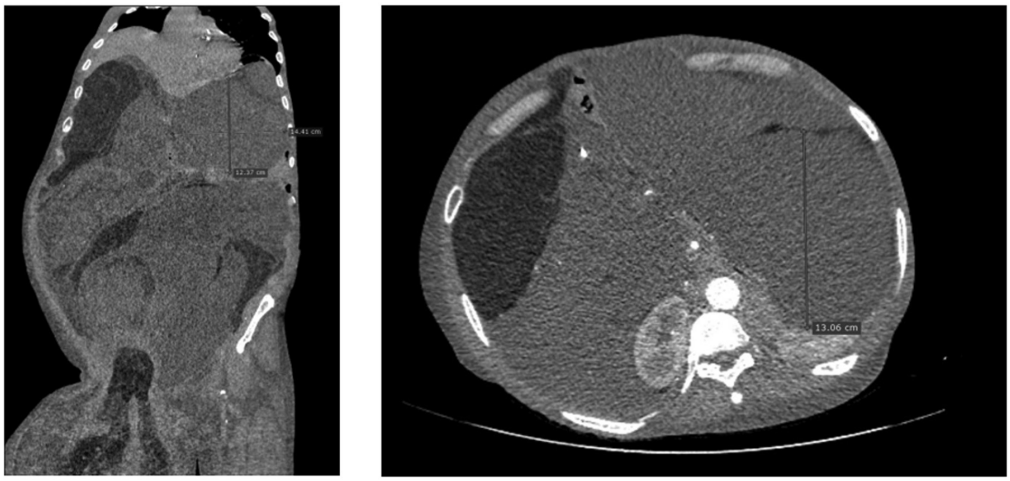

Figure 2-A & B. There are multiple varisized complex enhancing masses seen occupying the entire abdomen with the largest in the left hemiabdomen measuring 12.3 cm x 14.4 cm x 13 cm.

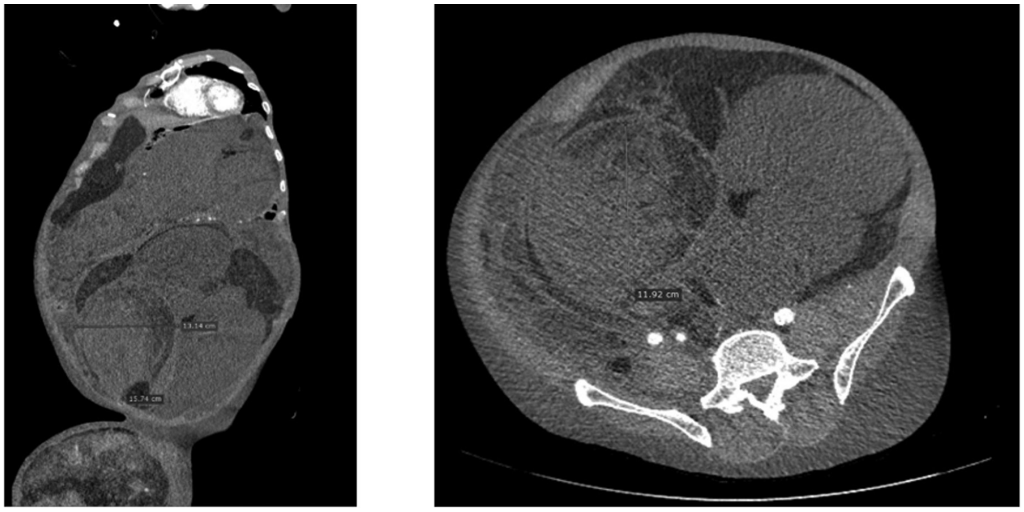

Figure 2-C & D. There are multiple varisized complex enhancing masses seen occupying the entire abdomen with the largest in the right hemiabdomen measuring 15 cm x 13 cm x 11 cm.

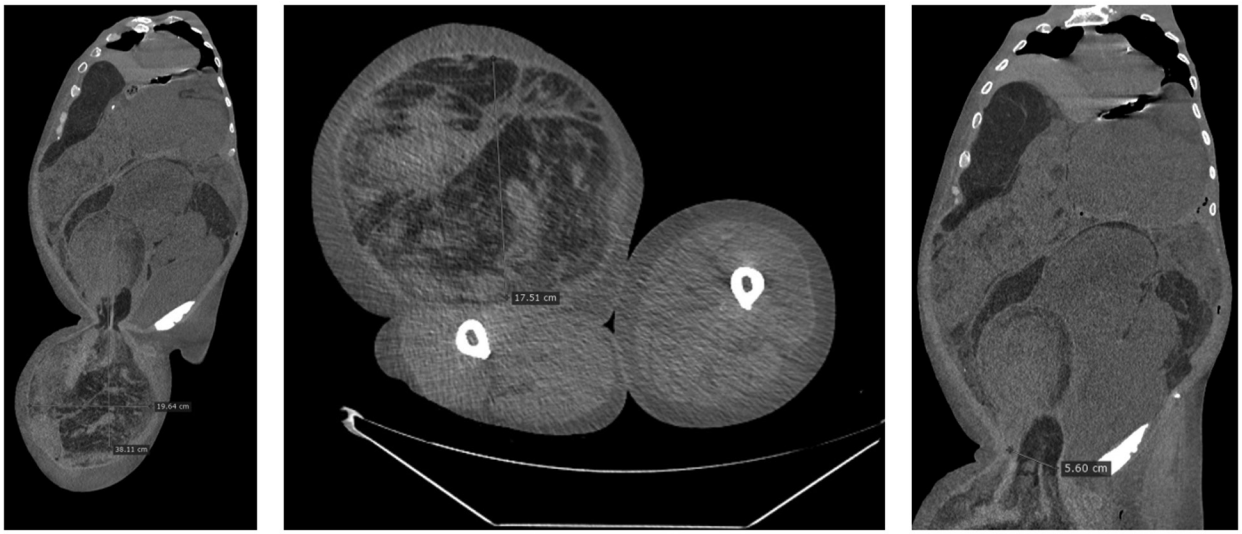

Figure 2-E to G. Likewise, there is evident herniation of the pelvic mass towards the right scrotal compartment approximately measuring 38.9 cm x 19. X 17.5 cm with pelvic floor defect, with the widest diameter of 5cm.

for extension towards the inguinoscrotal region.[11] Malignant tumors found within the inguinal canal are rare at around 30% incidence rate.[7]

Retroperitoneal sarcomas tend to be slow growing and are diagnosed late due to peritoneal accommodation of the tumor. This means it will have to attain massive size in order for it to affect the adjacent structures which is so often the case.[3]

There have been several journal articles which have discussed retroperitoneal sarcomas extending into the inguinal canal but of the well differentiated subtype.[6,7,9,11,14] There is limited study of retroperitoneal liposarcoma extension into the inguinal canal but in a retrospective study of 133 liposarcoma patients by Knebel, et al., 40 were of myxoid subtype with retroperitoneal liposarcomas accounting for 3.76% of the total.[4] Furthermore a study by Rhu, et al. of 168 patients with retroperitoneal liposarcomas who underwent treatment, 3.6% of the total had inguinoscrotal extension.[11]

In this prsent case, there is prior knowledge of the specific malignancy via histopathology so the role of imaging modalities such as contrast enhanced CT scan and contrast enhanced MRI, especially in such recurrent retroperitoneal liposarcoma is of utmost importance due to excellent volumetric evaluation and delineation of tumor margins to assess the prognosis of the patient. CT attenuation may help in the diagnosis of liposarcoma as well as in the prediction of the subtype and prognosis before surgery. Abdominal CT scans also provide an extremely accurate tumor volume measurement.[10]

In this patient, one of the factors which led to unfavorable prognosis may have been imaging findings of size greater than 5 cm, known to be contributory to decreased survival rate for myxoid liposarcomas.[2,4] In addition, it was found out that retroperitoneal liposarcoma with inguinal extension had significant higher postoperative mortality and morbidity rates in comparison with direct inguinoscrotal liposarcoma patients.[11] The capacity of myxoid liposarcomas for tumor bulk of massive proportions more or less contribute to the distention of the adjacent inguinal connection to the retroperitoneum which enables the tumor itself to infiltrate and extend into the scrotal region of patients even with such poorly metastasizing disease.

There are several independent factors which define unfavorable prognosis in most liposarcoma patients namely grading, liposarcoma subtype, surgical margins, metastases and tumour size.[3] Surgical removal is a requirement for disease free future, but complete surgical resection is difficult and recurrence is common.[9]

For most studies, gadolinium enhanced MRI was the most sensitive of imaging modalities for diagnosis of soft tissue and bony involvement due to its ability to demonstrate the solid nature of myxoid liposarcoma metastases.[12,13] such that it must be imperative to include it in protocols which involve lipomatous tumors for better evaluation.

For patients who had previously repaired hernias, re-protrusion of the tumor indicates increased high retroperitoneal pressure equating to possible unmanageable size and spread.[11]

Present case has been lost to follow up beyond the current NCCN Guidelines for retroperitoneal liposarcoma surveillance which is physical examination with appropriate imaging studies every 3-6 months post resection for the following 2 to 3 years which could be one of the possible reasons why case was been deemed unresectable. This essential time period for surveillance has been reiterated in a retrospective study by Kim, et al. in which a 3 month follow up interval is ideal for early detection of recurrent retroperitoneal liposarcomas.[10] Since liposarcomas have such a very aggressive recurrence rate, the earlier the surveillance done improves the survival rate of the patient.

SUMMARY/CONCLUSION

Retroperitoneal myxoid liposarcomas are rare entities, accounting for less than 1% of all malignancies such that there is very limited study about it.[3] It has a high recurrence rate despite low metastatic potential and should be approached with caution. For patients who had previously repaired hernias, re-protrusion of the tumor indicates increased high retroperitoneal pressure equating to possible unmanageable size and spread.[11]

With this in mind, the use of the appropriate imaging tools in present case is vital for the appropriate course of action for myxoid liposarcoma since surgery is the gold standard of treatment, especially in recurrent cases. A combination of contrast enhanced CT and MRI is suggested to be the most sensitive protocol for follow up soft tissue surveillance.[2,3,12]

Protrusion in a previously repaired inguinal canal as was the present case is a definitive sign of late retroperitoneal liposarcoma.[11] Proper surveillance

imaging post-tumor resection should be given more emphasis such as to avoid extensive spread and growth to unresectable proportions such as found in our case. In line with this train of thought, it has been suggested as possible improvement for survival time for recurrent myxoid liposarcoma cases is to have shorter follow-up intervals than the usual, suggested at least 3 months status post surgical resection to improve complete resection rate.[10]

REFERENCES

1. Muratori F, Bettini L, Frenos F, et al. Myxoid liposarcoma: Prognostic factors and metastatic pattern in a series of 148 patients treated at a single institution. Int J Surg Oncol 2018; Article ID 8928706, 9 pages, 2018.
2. Loubignac F, Bourtoul C & Chapel F. Myxoid liposarcoma: a rare soft-tissue tumor with a misleading benign appearance. World J Surg Oncol 2009; 7: 42.
3. Guo S, Xu Y, Qian F, Ma J, Wang S, Chen P, Zong L. A recurrent giant retroperitoneal myxoid liposarcoma: a case report and literature review. Transl Cancer Res 2019; 8(7): 2672-6.
4. Knebel C, Lenze U, Pohlig F, et al. Prognostic factors and outcome of Liposarcoma patients: a retrospective evaluation over 15 years. BMC Cancer 2017; 17: 410.
5. O'Regan KN, Jagannathan J, Krajewski K, Zukotynski K, Souza F, Wagner AJ, Ramaiya N. Imaging of liposarcoma: classification, patterns of tumor recurrence, and response to treatment. Am J Roentgenol 2011; 197(1): W37-43.
6. Baldassarre E, Santacroce C, Barone M, Torino G, Siani A, Valenti G. Retroperitoneal well-differentiated liposarcoma presenting as an incarcerated inguinal hernia. G Chir 2007; 28(8-9): 315-7.
7. Kim JM, Choi MH, Lee SL & Ku YM. Dedifferentiated retroperitoneal liposarcoma presenting as right inguinal hernia: A case report. J Korean Soc Radiol 2016; 74(1): 66.
8. Chowdhry V, Goldberg S, DeLaney TF, et al. Myxoid liposarcoma: Treatment outcomes from chemotherapy and radiation therapy. Sarcoma 2018, Article ID 8029157.
9. Fiaschetti V, Castellani F, Croce G, Mondillo M, Amico A, De Majo A, Floris R. Incidentally discovered well-differentiated retroperitoneal liposarcoma with inguinal canal herniation: report of 2 cases. Radiology Case Reports 2017; 12(2): 308–12.
10. Kim EY, Kim SJ, Choi D, Lee SJ, Kim SH, Lim HK, Song SY. Recurrence of retroperitoneal liposarcoma: imaging findings and growth rates at follow-up CT. Am J Roentgenol 2008; 191(6): 1841-6.
11. Rhu J, Cho CW, Lee KW, Park H, Park JB, Choi YL & Kim SJ. Comparison of retroperitoneal liposarcoma extending into the inguinal canal and inguinoscrotal liposarcoma. Canadian J Surg 2017; 60(6): 399–407.
12. Sheah K, Ouellette HA, Torriani M, Nielsen GP, Kattapuram S, Bredella MA. Metastatic myxoid liposarcomas: imaging and histopathologic findings. Skeletal Radiol 2008; 37(3): 251-8. doi: 10.1007/s00256-007-0424-1. Epub 2007 Dec 21.
13. Sung MS, Kang HS, Suh JS, Lee JH, Park JM, Kim JY, Lee HG. Myxoid liposarcoma: appearance at MR imaging with histologic correlation. Radiographics 2000; 20(4): 1007-19.
14. Noguchi H, Naomoto Y, Haisa M, Yamatsuji T, Shigemitsu K, Uetsuka H, Hamasaki S, Tanaka N. Retroperitoneal liposarcoma presenting as indirect inguinal hernia. Acta Med Okayama 2001; 55(1): 51-4.