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Available online at: http://www.iajps.com *A Case Report*

SURGICAL RESECTION OF GIANT LIPOSARCOMA RARELY MIMICKING ANGIOMYOLIPOMA

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Abstract:

Background

Retroperitoneal Liposarcoma can mimic several masses; including benign tumors like lipoma, angiomyolipoma. Medical literature usually describes cases where an angiomyolipoma were masqueraded as Liposarcoma. To the best of our knowledge, this is the second case in medical literature were a Liposarcoma was mimicking an angiomyolipoma.

Case report

We report a rare case of a 55-year-old male presenting to the clinic complaining of abdominal enlargement and signs of renal failure. CT scan showed a huge heterogeneous mass arising from the lower pole of the right kidney. The mass was found to be a retroperitoneal liposarcoma mimicking an angiomyolipoma.

Conclusion

Distinguishing liposarcoma from angiomyolipoma is important as liposarcoma are usually associated with a high reoccurrence rate that needs frequent follow-up. One of the important markers is the MSA/ HMB45/ Melan stain, which, if positive, usually associated with angiomyolipoma.

Keywords: *liposarcoma; retroperitoneal liposarcoma; angiomyolipoma; giant; rare.*

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INTRODUCTION :

Liposarcoma (LPS) is one of the most common types of adult soft-tissue sarcoma. Although fat / lipogenic differentiation is common to all, term liposarcoma includes somewhat diverse neoplasms with different clinic-pathological presentation, prognostic behavior, and genomic alterations. (1) Comparing it to other sarcomas, 10-20% of all primary retroperitoneal sarcomas are liposarcomas. (2) This represents overall incidence of 0.3%–0.4% per 100,000 of the population. (3) The puzzling thing about this specific type of sarcoma is that till this day no aetiological factors have been linked to it. (4) Predilection of Retroperitoneal Liposarcoma (LSRP) is between 40 and 60. (5)

Retroperitoneal LPS can mimic several masses; including benign tumors like lipoma, angiomyolipoma (AML), and other different types of sarcoma. Specifically, AML can be differentiated from well-differentiated liposarcomas (WDLS) by the presence of renal parenchymal defects and large vessels on imaging studies. (6) Unlike WDLS, dedifferentiated liposarcoma have a metastatic potential. It is estimated that 10% of WDLS progress into dedifferentiated liposarcoma over a period of 7 to 8 years. Another complication of WDLS is local reoccurrence, which in some medical literature, was said to effect 100% of patient who have a deep-seated retroperitoneal WDLS. (7)

The diagnosis of WDLS is generally suspected when a patient present with a slow growing painless mass, and because masses located in the retroperitoneum maybe symptomless for a prolonged period, these lesions – when discovered – can grow to a large size. At the time of diagnosis, the patient usually come presenting with an increase in abdominal girth, or any symptom caused by the compression of adjacent organs. (8) The mean diameter for a retroperitoneal WDLS is 20 cm, yet the presence of even larger masses at the time of diagnosis had been reported. (9)(10)(11) Nevertheless, the use of imaging had greatly decreased the size of these lesion nowadays. A Computed tomography (CT) or Magnetic Resonance Imaging (MRI) can help delineate the mass, which will be isointense compared to subcutaneous fat elsewhere in the body. However, the definitive diagnosis can only be made with histology. (7)

Case reports published in medical literature usually describe cases where an AML were masqueraded as WDLS. (12)(13) but instead, in this article, we report a case of a giant WDLS mimicking an angiomyolipoma. To the best of our knowledge, only one case was reported as retroperitoneal liposarcoma mimicking angiomyolipoma in English literature. (14)

CASE PRESENTATION:

A 55-year-old male was presented to the clinic complaining of abdominal enlargement and signs of renal failure. Other parts of the examination were normal. the Ultrasound (US) was performed for the abdominal mass and for the high creatine level and it showed a huge right sided abdominal collection measuring about 14x11 cm suspected to be psoas hematoma/abscess, the mass displaces the lower pole of the right kidney. The liver Portal Vein (PV), Common Bile Duct (CBD), Gallbladder (GB), and spleen were normal with average size and with no focal lesions. Therefore, an un-enhanced axial CT scan (Fig.1 & 2) of the abdomen and pelvis with sagittal and coronal reformat was performed and it showed a huge heterogeneous mass of soft tissue and fatty density arising from the lower pole of the right kidney. The mass was measuring 18x12cm denoting a huge renal angiomyolipoma. There was mild right side hydronephrosis, but the left kidney was of average size. No free fluids or free air. Solid abdominal organs were grossly unremarkable. Bilateral lower lung lobes were patchy with ground glass appearance. Moreover, a huge complex retroperitoneal lesion that was seen abutting the lower pole of the right kidney and compressing its outer cortex with mixed densities inside and prominent fat. The mass also showed ‘Claw’ sign which indicated that it might be a parenchymal tumor.

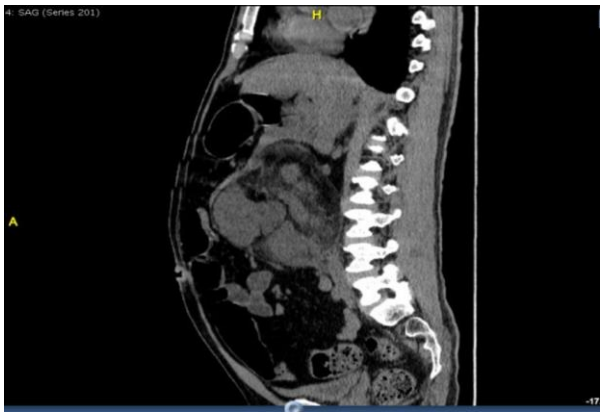
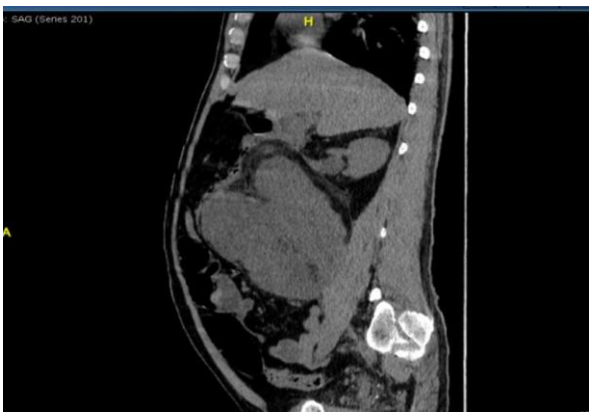
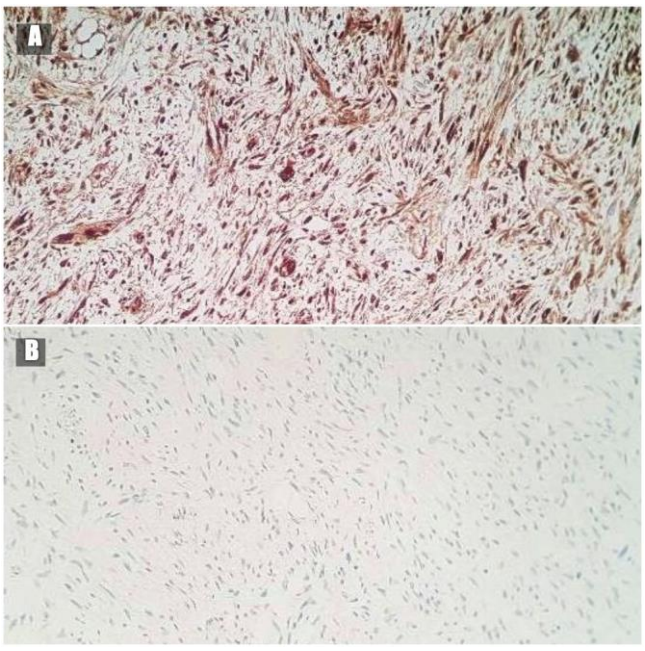
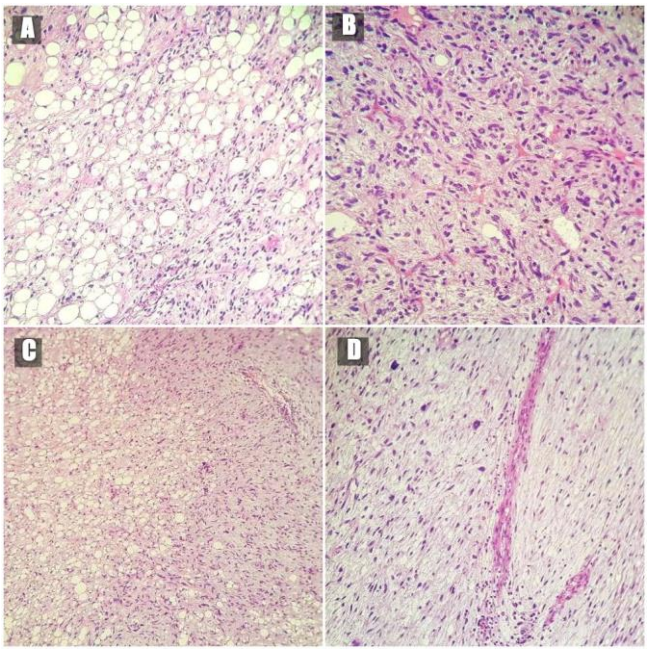



Figure 1 & 2: CT scan of the abdomen and pelvis with sagittal reformat showing a huge heterogeneous mass arising from the lower pole of the right kidney.

At this point, a histopathology biopsy was mandatory to recognize the nature of the mass. A US guided core needle biopsy (CNB) sections (fig. 3 & 4) revealed cores of adipose tissue showing lipocytes mainly with some spindle cells. Few lipoblasts were noted. Some cells have enlarged hyperchromatic nuclei, some are multinucleated. Mitosis was not seen. Foci of ischemic necrosis was noted. The main thing was that it is Muscle-Specific Actin (MSA)/

Human Melanoma Black (HMB45)/ Melan A: Negative

Figure 3 (A : p16- B: Melan-A) & 4 (Hematoxylin and eosin stain A:fatty areas: B: high power C: Medium power D: Gains cells): histopathology sections revealed cores of adipose tissue showing lipocytes mainly with some spindle cells with enlarged hyperchromatic nuclei.

The histopathology results went against the primitive impression of the radiology team that it was AML. Later upon excision (Fig. 5 & 6), we found grossly an oval firm circumscribed mass that was measuring 26 x 16 x 13 cm. The outer surface was mostly smooth at the anterior aspect (peritoneal covering), and it was rough elsewhere and vaguely nodular. Surrounded by little fat, gonadal vessels were seen. Cut section showed a solid grey vaguely nodular with focal myxoid areas and focal necrosis. Sections reveal mesenchymal tumor formed mainly of proliferated spindle cells having mostly bland looking elongated nuclei, granular chromatin, small to inconspicuous nucleoli and ill-defined cytoplasm. Few scattered mitotic figures were seen. These cells were arranged in sheets, fascicles with focally myxoid stroma and scattered large bizarre hyper-chromatic to multinucleate cells/Floret cells were noted. A thin fibrocollagenous layer is noted at the periphery. No lymph node tissue could be identified in the surrounding fat.

Figure 5 & 6: Gross section revealed an oval firm circumscribed mass that was measuring 26 x 16 x 13 cm.

CASE DISCUSSION:

Retroperitoneal tumors represent a diagnostic dilemma while recognizing and managing them very early is censorious. These tumors can be identified in late stages when they show mass effect, renal failure as in our patient or infiltrating the adjutant vital organs. In addition, while some of them are benign it is four times more likely for it to be a malignant tumor rather than a benign lesion. Although the most appropriate treatment for these lesions is complete surgical reaction, the nature of the mass can give you important guideline for exploring other methods of treatment and the follow up plan. (1) After reviewing recent development in imaging and histopathological techniques we offer here some points that might help physicians to establish the diagnosis of LPS when it is bizarrely behaving and mimicking AML. the difficulty of diagnosing this patient was established by some unique features that this mass had and need to be reported. Firstly, as a result of the massive size of the tumor which waited around 2.5kg upon extension, the radiological study reported the major "beard beak" that usually appears in tumors that originate from the renal parenchyma like AML and Willis tumor. This suggested a very important distinguisher point between AML and LPS and show LPS mimicking AML. (15) On the other hand, our histopathological CNB could not evidently make the diagnosis due to the abundant spindle cells that showed on the section. However, it did show MSA/ HMB45/ Melan A: Negative which, if positive, usually associated with AML. At this point, complete surgical resection was the best option to manage this tumor and to diagnose the nature of it. Surgical resection made the final diagnosis of the mass and a flow up plan was mandatory at this point for the fact that LPS has a high recreant rate.

CONCLUSION

Distinguishing liposarcoma from angiomyolipoma is important as liposarcoma are usually associated with a high reoccurrence rate that needs frequent follow-up. One of the important markers is the MSA/ HMB45/ Melan stain, which, if positive,

usually associated with angiomyolipoma.

Ethical approval:
It was not necessary

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Abbreviations: LPS, Liposarcoma; LSRP, Retroperitoneal Liposarcoma; AML, angiomyolipoma; WDLS, well-differentiated liposarcomas; CT, Computed tomography; MRI, Magnetic Resonance Imaging; US, Ultrasound; PV, Portal Vein; CBD, Common Bile Duct; GB, Gallbladder; CNB, core needle biopsy; MSA,

Muscle-Specific Actin; HMB45, Human Melanoma Black.