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Case Report OMICS International

A Case Demonstrating the Diagnostic and Surgical Difficulties of a Primary Dedifferentiated Retroperitoneal Liposarcoma

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Abstract

This case report describes the presentation and management of an extremely large, rare, dedifferentiated primary retroperitoneal liposarcoma. We discuss the diagnostic and surgical challenges due to massive tumor size and dedifferentiated tissue characteristics. Radiologic and pathologic features are reviewed to promote awareness and understanding of Dedifferentiated Liposarcoma (DL).

Keywords: Dedifferentiated; Liposarcoma; Malignancy; Retroperitoneal; Sarcoma

Abbreviations DL: Dedifferentiated Liposarcoma; STS: Soft Tissue Sarcomas; RPS: Retroperitoneal Sarcomas; ALN/WDL: Atypical Lipomatous Neoplasm/well-differentiated Liposarcoma; MDM2: Murine Double Minute type 2; CDK4: Cyclin-dependent Kinase 4; RUQ: Right Upper Quadrant; CTA: CT Angiography; IVC: Inferior Vena Cava; ICU: Intensive Care Unit.

Introduction

Soft tissue sarcomas (STS) are rare, comprising <1% of all adult malignancies, but represent over 50 types and subtypes of mesenchymal tumors [1]. Liposarcoma is the most common variant, accounting for 20% of all STS and 50% of all retroperitoneal sarcomas (RPS) and is classified by four histologic subtypes: Atypical lipomatous neoplasm/well-differentiated liposarcoma (ALN/WDL), myxoid liposarcoma, pleomorphic liposarcoma, and dedifferentiated liposarcoma (DL) [1].

DL is a non-lipogenic, high grade, malignant sarcoma, 90% arising de novo with a 15-20% metastatic rate, usually in the 6th-7th decad [1]. Due to the expandable deep space and complex anatomy of the retroperitoneal cavity, DL usually presents late, often greater than 10-20 cm, making surgical resection difficult [1]. Prognosis is variable, but often poor. Even with complete gross resection, local recurrence is common and usually the cause of death [1]. Murine double minute type 2 (MDM2) and cyclin-dependent kinase 4 (CDK4) amplification are characteristic on histopathology [1].

Case Report

A 74-year-old man with a history of atrial fibrillation (on warfarin) and bradycardia status post pacemaker placement, presented to the ED with the acute onset of right flank pain. The episode awoke him from his sleep, was described as sharp, pleuritic, and radiated to the chest and right upper quadrant. A right upper quadrant (RUQ) ultrasound and CT angiography (CTA) of the thorax revealed a large, partially visualized, complex cystic mass inseparable from the right kidney, new compared to a CTA thorax less than 1.5 years prior. A CT abdomen/pelvis was then performed, with residual IV contrast from the previous CTA then in the late excretory phase. The mass measured 17 × 14 × 15 cm, ranging from 10-50 Hounsfield units (without gross fat), causing mass effect on the IVC, right adrenal gland, and liver (Figure 1). Although the dominant cystic portion did not contain contrast, a duplicated collecting system was identified, and a markedly dilated, nonfunctioning, right upper pole moiety was postulated. The primary diagnostic considerations were an obstructing urothelial neoplasm versus a large centrally necrotic renal cell carcinoma. The patient was diagnosed with suspected malignancy and discharged with follow-up in outpatient urology.

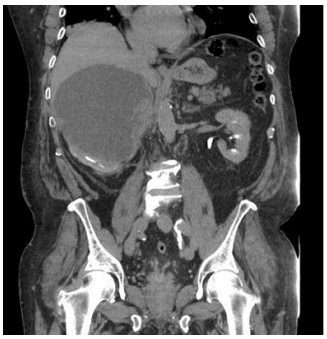

Figure 1: Initial CT of the abdomen and pelvis (IV contrast from preceding CTA of the thorax in the late excretory phase) shows a large mass in the upper pole of the right kidney, with solid and cystic components but no visible gross fat.

Over the next three weeks, the patient's symptoms progressively worsened. Follow up outpatient CT imaging revealed an enlarging heterogenous RUQ mass, now measuring 19 × 15 × 18 cm. The patient was admitted with an INR of 10.8, worsening right flank pain,

intractable nausea, vomiting, and low urine output. His hemoglobin trended down, and a repeat CT confirmed acute hemorrhage within the mass (Figure 2). A blood transfusion was given and the patient was transferred to a tertiary center for further evaluation and management.

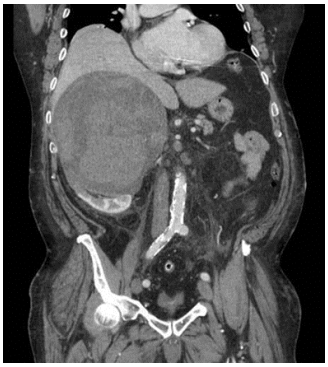

Figure 2: Follow up CT 3 weeks later revealed an enlarging mass with heterogeneously increased density consistent with acute internal hemorrhage.

The patient was stabilized, his anti-coagulation reversed, and interventional radiology was consulted to perform an embolization. On angiography, there was no visible arterial blood supply to the tumor from the aorta, right renal artery, or right hepatic artery, and no extravasation of contrast to localize the hemorrhage; therefore, no embolization was performed (Figure 3).

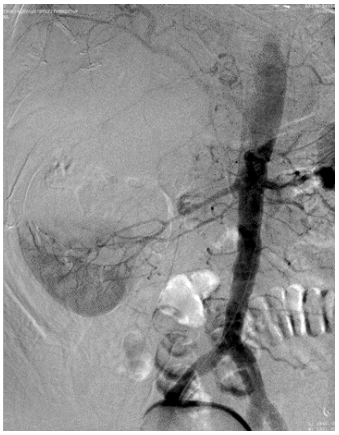

Figure 3: Catheter directed angiography did not show any arterial blood supply to the tumor from the aorta, right renal or right hepatic arteries, and no extravasation of contrast was seen to localize the source of bleeding. The mass is represented by a large filling defect, with only a small portion of the relatively preserved inferior pole of the right kidney visible.

The patient was urgently taken to the operating room due to respiratory compromise and uncontrolled pain. The large mass obstructed the surgical field and was densely adherent to the retroperitoneum, so the tumor/hematoma was entered and evacuated allowing better exposure.

There were no identifiable planes to distinguish the tumor from the right kidney, IVC, diaphragm or psoas muscle, and no normal right adrenal gland was identified. After dissection, the tumor and kidney were removed en bloc. A 4 cm retro-hepatic IVC laceration was repaired, and minor bleeding from the liver and psoas was controlled. The patient was transferred to the surgical ICU in stable condition, and discharged on post-operative day 15. Unfortunately, he suffered from multiple post-operative complications due to ongoing retroperitoneal hemorrhage, and after two readmissions with declining clinical status and no further treatment options, the family elected to pursue comfort care. The patient passed away later that night, three months after his initial presentation.

Pathology revealed a dedifferentiated liposarcoma (DL), stage pT2b, with a mitotic rate >20/10 high-powered field, 50% necrosis and a histologic grade of 3. Immunohistochemistry revealed diffuse positivity for CDK4 and scattered positivity for MDM2. Cytogenetic analysis was positive for MDM2 amplification. Tumor cells were negative for all other markers.

Discussion

DL accounts for 18% of all liposarcomas, occurring at a 3:1 ratio within the retroperitoneum vs. the extremities. Grossly, DL present as yellow-tan multinodular masses with tan-gray areas of dedifferentiated tissue (Figure 4) [2,3]. Histologic diagnosis can be difficult; however, Binh et al. [4] has demonstrated that immunohistochemical stains for MDM2 and CDK4 help separate DL from various poorly differentiated sarcomas. Specifically, in their study, DL stained positive for MDM2 in 97% of cases and for CDK4 in 92% of cases (Figure 5) [4]. In our case, the tumor stained positive for both MDM2 and CDK4, assisting in the pathological identification.

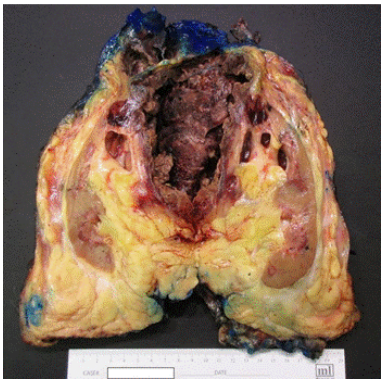

Figure 4: Gross pathologic specimen demonstrating a centrally hemorrhagic/necrotic, yellow and tan tumor completely engulfing the right kidney. All margins were positive for sarcoma.

The clinical behaviors of the subtypes of liposarcomas differ. On CT and MR imaging, WDL appears similar to adipose tissue and tends to be well encapsulated. In contrast, DL typically demonstrates a non-lipomatous heterogenous mass [5]. There is a local recurrence rate of about 40% for DL, with 28% passing away because of the malignancy [2].

Surgical resection of DL may be difficult, and incomplete resection is common. The masses tend to be large at identification, distorting surrounding anatomy and making the procedure more complex and

hazardous. In one study, Lewis identified that initial stage, high histological grade, positive margins, and inability to completely resect the primary tumor were associated with a higher mortality rate [6].

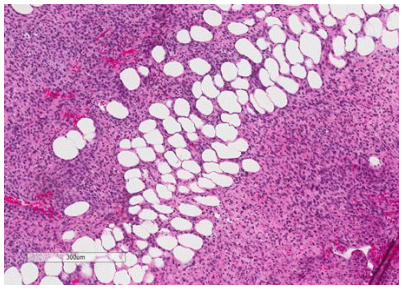

Figure 5: Adipocytes with interspersed with pleomorphic spindle cells. Immunohistochemistry revealed diffuse positivity for CDK4 (Cyclin-dependent Kinase 4) and cytogenetic analysis was positive for MDM2 (Mouse double minute 2 homolog) gene amplification, consistent with dedifferentiated liposarcoma.

Our Case

In our case, radiographic identification of the DL was challenging due to lack of gross fat and extensive involvement with the right kidney. Management was complicated by the patient's coagulopathy and intratumoral hemorrhage. Lack of arterial blood supply on angiography precluded pre-operative embolization. Intra-operative hematoma evacuation and IVC laceration likely contributed to post-operative complications, including multiple expanding retroperitoneal hematomas. An earlier, accurate diagnosis may have prompted an earlier intervention, but the outcome would likely have remained unchanged.

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