Case Report

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A rare well-differentiated renal retroperitoneal liposarcoma: a case report

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Introduction and importance: Retroperitoneal liposarcomas (RPLPSs) are rare tumors that arise from mesenchymal cells in the peritoneum cavity. The sites of PRLPSs vary a lot, but renal PRLPSs are extremely rare (there are only 45 cases of Renal retroperitoneal liposarcomas on PubMed). In this case, the authors present a rare renal retroperitoneal liposarcoma case, describe the major concepts, and raise awareness about this rare tumor.

Case presentation: A 44-year-old woman presented to the clinic with hirsutism and irregular menstruation; upon physical examination, a large abdominal mass was accidentally identified; the patient had Doppler ultrasound (Doppler US) and MRI, which both showed a mass arising from the right upper pole of the kidney, fine needle aspiration (FNA) confirmed the diagnosis of retroperitoneal liposarcoma, patient underwent surgical removal and her symptoms disappeared.

Clinical discussion: The retroperitoneum is a cavity behind the abdominal wall containing organs like the pancreas and kidneys. Retroperitoneal tumors (RPTs) are rare neoplasms, primarily of mesenchymal origin. Retroperitoneal liposarcomas (RPLPS) are the most common RPT, often asymptomatic until large, and rarely metastasize but frequently recur. Liposarcomas are classified into five subtypes, with well-differentiated liposarcoma being the most common and characterized by high local recurrence. The presence of specific oncologic mutations affects the prognosis and the response to treatment.

Conclusion: In rare cases, retroperitoneal liposarcomas can arise from sites near the kidney and compress the adjoining adrenal gland.

Keywords: case reports, liposarcoma, renal, retroperitoneal

Introduction and importance

Retroperitoneal liposarcomas (RLS) are classified as rare malignant tumors arising from the retroperitoneum cavity's mesenchymal layer, accounting for (12–40)% of all liposarcomas[1]. RLS are often asymptomatic due to their slow-growing and deep location. Still, late signs and symptoms can be presented as an increased abdominal girth, palpable lump, abdominal pain, and compression of the surrounding structures.

They mainly present with non-specific symptoms related to the mass local compression; the patient can describe vague abdominal pain, renal insufficiency, adrenal dysfunction, and neurological symptoms.

HIGHLIGHTS

- Renal retroperitoneal liposarcoma is a rare aggressive tumor manifesting in adults and may compress the surrounding organs.
- Well-differentiated liposacroma has a good prognosis with 5-year survival of 83%; however, there are high rates of local recurrence.
- Treatment options depend on the subtype of the tumor, as not all subtypes respond well to chemotherapy or radiotherapy.

Well-differentiated liposarcoma (WDLPS) caused by an amplification in the 12q13–15 region is the most common type of renal retroperitoneal liposarcoma. Due to its high recurrence rate, surgical removal is the first regimen in the treatment approach of WSLPS.

Renal retroperitoneal liposarcomas are extremely rare tumors with little information in the medical literature (there are only 45 cases of Renal retroperitoneal liposarcomas on PubMed).

Due to its rarity, Renal RLS can be easily misdiagnosed when evaluating renal masses. Thus, in this paper, we are going to present a rare case of renal retroperitoneal liposarcomas, describe the presentation, discuss the potential management techniques, and raise awareness of its rarity.

This work has been reported in line with the SCARE 2023 criteria.

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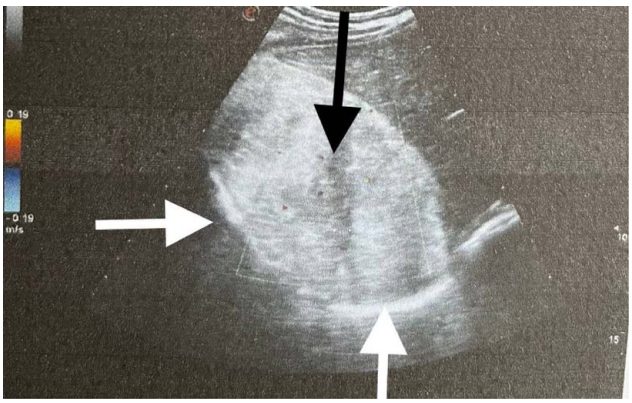

Figure 1. Doppler ultrasound. Revealing a capsulated (white arrows) hyperacoustoic mass (black arrow).

Case presentation

We present a case of a 44-year-old woman who presented to the clinic with hirsutism and irregular menstruation. Upon physical examination, an enlarged abdomen was noticed. The patient underwent an abdominal ultrasound, which revealed a large asymptomatic mass involving the upper pole of the right kidney. General Lab tests were done, and several abnormalities were observed, including elevated serum morning cortisol after a low-dose dexamethasone suppression test (4.2 μg/dl, normal 0.6–1.8 μg/dl), decreased ACTCH (7.82 pg/ml, normal 10–60 pg/ml), and high levels of vanillylmandelic acid in urine (15.5 mg/24 h, normal 1–6 mg/24 h). Doppler ultrasound showed a huge capsulated mass located below the liver and above the kidney and extending to the adrenal chamber and the lateral edges of the right kidney (Fig. 1). however, the gallbladder was normal, as were the spleen and pancreas. No lymph node enlargement was seen, and no free fluid was in the abdomen. MRI revealed a mass with well-defined edges and a high signal. The mass attaches to the adjacent right kidney and pushes it inferiorly and medially (Fig. 2). no metastases were detected. A fine needle aspiration (FNA) with ultrasound guidance confirmed the diagnosis of Retroperitoneal well-differentiated liposarcoma and revealed a multiple infiltration of atypical fatty tissue that resembles lipomas into the renal parenchyma. Closer inspection showed scattered tumor cells with large atypical hyperchromatic nuclei. (differentiation score 1, mitosis score 1, necrotic score 0, total score 2) /Histologic grade 1. (Fig. 3). The patient underwent a complete mass resection with a right nephrectomy (due to the tumor cells infiltration) (Fig. 4), mass measured $16 \times 10.5 \times 7.5$ cm macroscopically, the capsule of the

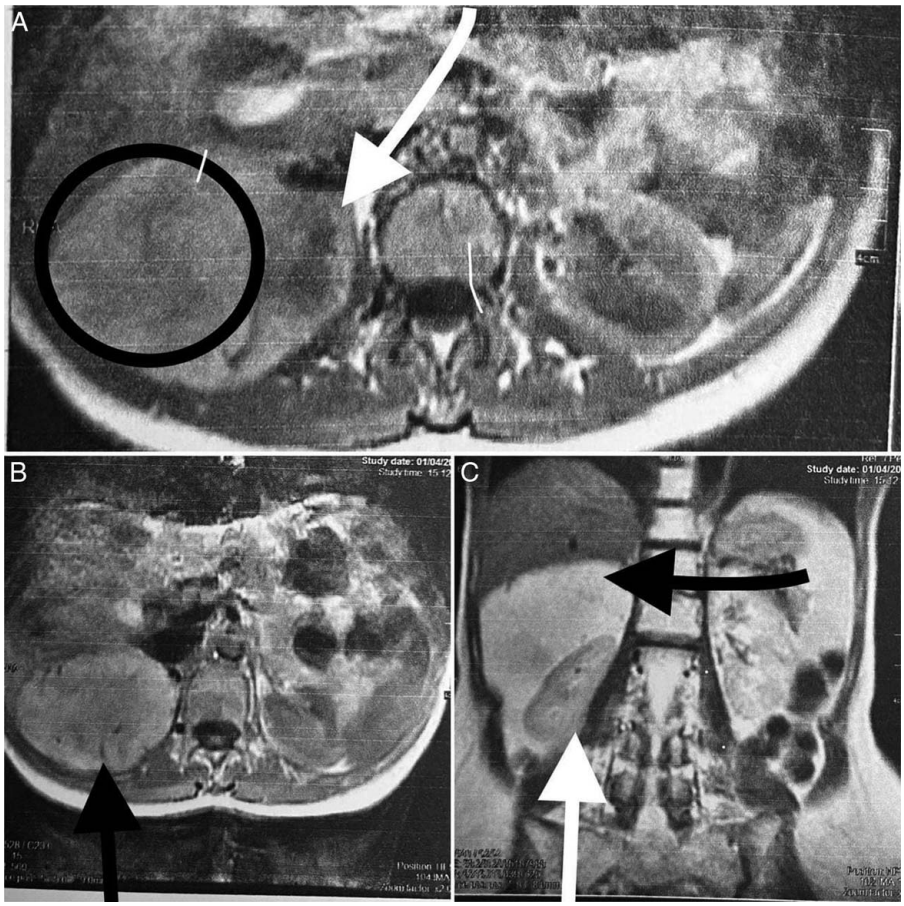

Figure 2. MRI images. (A) (T1 + C): axial MRI showing the large mass (black circle) attach into adjacent right kidney (white arrow) and pushes it inferiorly and medially. (B) (T1 + C): axial MRI showing the large mass (black arrow) at the upper pole of right kidney (located inferiorly to the mass, thus doesn't appear in the image). (C) (T2WI): sagittal MRI showing the mass (black arrow) surrounding the right kidney (white arrow).

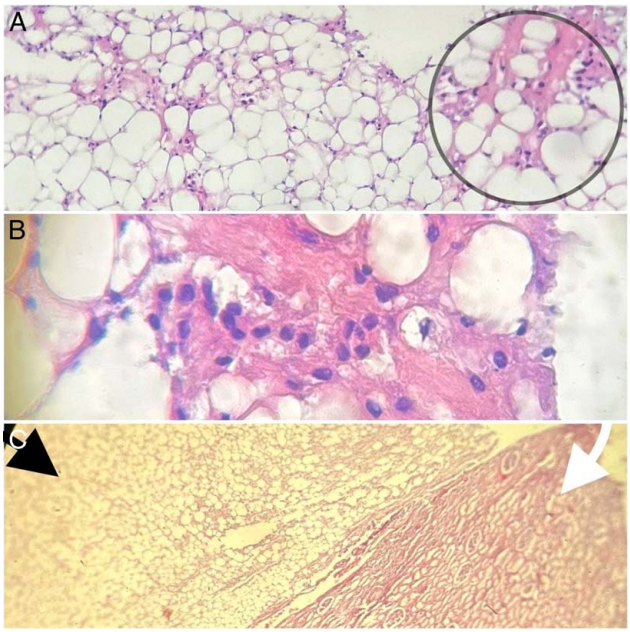

Figure 3. histopathological study of FNA. (A) (20×): showing the lipomalike appearance with a suspicious finding (scattered tumor cells with large atypical hyperchromatic nuclei and atypical fatty tissue that resembles lipomas) (zoom in circle). (B) (40×): studying of the zoom in circle, revealing of a hyperchromatic Polymorphic nucleus (confirm the diagnosis). (C) (10×): normal kidney tissue (white arrow) adjoining the liposarcoma mass (black arrow).

mass was vascular and partially macerated, the mass's cut surface was homogenous, yellow, soft, and resembling cerebral convolutions. Renal parenchyma showed remarkable tumor infiltrations. Surgical margins were free.

After surgery, the patient was given instruction to see an endocrinologist in order to investigate the abnormal test findings, which may suggest Cushing syndrome, but the patient did not. One month after surgery, the patient came for a follow-up, and she revealed that she had no symptoms and was feeling just fine. Low-dose dexamethasone suppression test, ACTH, and vanillylmandelic acid (urine) were done, and they were all in the normal ranges. No other investigations were done, and the adrenal function test abnormalities were assumed to result from compression of the adjoining mass. As for the follow-up, the patient was asked to undergo a repeated abdominal ultrasound every six months (1 year of follow-up until now). Since it's a well-differentiated liposarcoma, no adjuvant radiotherapy or chemotherapy has been given (Not Responding, see discussion).

Clinical discussion

Retroperitoneum (RP) is an anatomical term that defines the cavity located behind the abdominal wall organs located between the posterior abdominal wall and the peritoneum are referred to as the retroperitoneal organs (pancreas, kidneys, adrenals, and part of the duodenum, ascending and descending colon)[1]. Retroperitoneal tumors RPTs are rare neoplasms that arise in the retroperitoneal space; the terminology of RPT was first described by Giovanni Battista Morgagni (1682–1771), reporting a 60-year-old woman with a retroperitoneal lipomatous tumor. later on, RPT was mentioned again by Jean Fréderic Lobstein (1777–1835) in his book Traité d'anatomie pathologique (1829)[1,2]. Most RPTs are known by their mesenchymal origin, a particular type of cells driven from the mesodermal layer and programmed to give several connective tissues[3–5].

Retroperitoneum liposarcoma (RPLPS) is the most common type of tumor arising in the retroperitoneal space. Most RPLPS cases in PubMed are in contact with the liver, spleen, duodenum, pancreas, iliac vessels, iliopsoas muscle, and femoral nerve. Yet, only a few cases are in contact with the kidney[6,7]. RPLPSs are described as silent tumors due to their deep positions, which do not tend to give symptoms until the tumor is already large. In fact, 90% of RPLPSs are greater than 10 cm in diameter at presentation. Common symptoms are all related to the localized compression of the enlarged mass (palpable mass in the abdomen, increased abdominal girth, adrenal dysfunction, renal insufficiency, urological or neurological symptoms)[6]. Despite their large size, RPLPSs rarely metastasize, and according to JS Liles, only 10% of cases are found to have metastatic disease at diagnosis. However, the percentage of local recurrence can be as high as 50% within 5 years after surgery[7]. According to the Memorial Sloan-Kettering Cancer Center (New York, NY), 60% of patients who previously had their tumor resected are at risk of recurrence disease[8]. Based on the WHO 2020 calcifications, liposarcomas are subdivided into five subtypes: well-differentiated, dedifferentiated, myxoid, pleomorphic, and myxoid pleomorphic[9]. Well-differentiated liposarcoma (WDLPS) is by far the most common type of all liposarcomas, commonly in the peritoneum, mediastinum, or para-testicular region and rarely in extremities. Major Histopathological features are a mature-appearing adipose tissue that can look identical to lipoma but has scattered lipoblasts or cells with enlarged hyperchromatic irregular nuclei. WDLPS is caused by an amplification in the 12q13–15 region, which includes a lot of proto-oncogenes MDM2, CDK4, HMGA2, and TSPAN31. WDLPS never metastasizes but carries a high risk of local recurrence. Thus, surgical removal with marginal free is the first regimen in the treatment approach[10] (Table 1). Dedifferentiated liposarcoma (DDLPS) is an aggressive, high-grade disease, arising most commonly within the retroperitoneum and associated with high rates of metastatic disease. DDLPS shares many common features with WDLPS; in fact, DDLPS is histopathologically characterized by more highly cellular areas of high-grade undifferentiated cells within a background of WDLPS[11] (Table 1). Myxoid liposarcomas (MLPS), occurring predominantly in the lower limbs of young adults, are extremely rare in the retroperitoneum.

Histopathologically, it consists of a myxoid stroma with more aggressive round/oval mesenchymal cells and immature lipoblasts. Translocation of t(12;16)(q13;p11), which leads to the production of FUS-DDIT3 fusion protein, is the primary pathological cause. Compared with WDLPS, MLPS has more risk for local recurrence and distal metastasis. Yet, it is more sensitive to chemotherapy and radiotherapy[12] (Table 1). Pleomorphic liposarcoma (PLPS) is a high-grade pleomorphic sarcoma that typically arises in the limbs or, less commonly, the trunk or retroperitoneum. Histological appearance contains high-grade undifferentiated cells with several pleomorphic lipoblasts. Several pathological causes have been described, such as Deletion of 13q14.2-5 (containing RB1), Mutation or loss of TP53, and Loss of the tumor suppressor gene NF1. As MLPS, PLPS has more risk for local recurrence and distal metastasis, yet is more sensitive to chemotherapy and radiotherapy[13] (Table 1). Myxoid

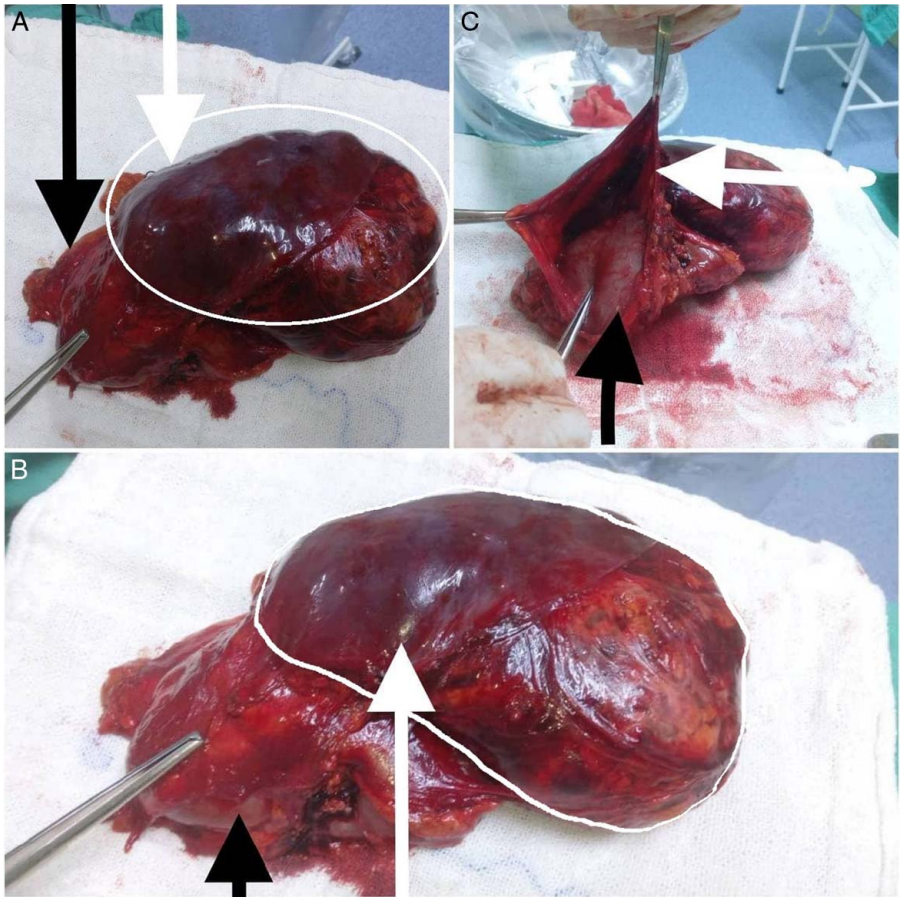

Figure 4. Mass resection with a right nephrectomy. (A, B) Showing the mass (black arrows) attached to the kidney (white arrows + white circles). (C) The mass capsule (white arrow) and the adipose tissue inside (black arrow).

Table 1

Comparison between types of liposarcoma

Prevalence (%)	WDLPS Most common[10].	DDLPS Less common[11].	MLPS Less common[12].	PLPS Rare[13].	MPLPS Extremely rare[9].
Common site/location	Peritoneum, mediastinum, and para-testicular[10].	Retroperitoneum[11].	Lower limbs[12].	Limbs[13].	Mediastinum followed by the limbs and the head and neck region[9].
Histopathology	Adipose tissue that can look identical to lipoma but has scattered lipoblasts or cells with enlarged hyperchromatic irregular nuclei[10].	More highly cellular areas of high-grade undifferentiated cells within a background of WDLPS[11].	Myxoid stroma with more aggressive round/oval mesenchymal cells and immature lipoblasts[12].	High-grade undifferentiated cells with many pleomorphic lipoblasts[13].	Shows features of both myxoid (presence of a rich capillary size vascular network set in myxoid background) and pleomorphic liposarcoma (presence of pleomorphic lipoblasts[9].
Pathological abnormalities	Amplification in the 12q13–15[10].	Amplification in the 12q13–15[11].	Translocation of t(12;16)(q13;p11)[12].	Deletion of 13q14.2-5, loss of TP53, loss of NF1[13].	Have not been detected yet[9].
Metastasis	Negative (never)[10]	Positive[11].	Positive[12].	Positive[13].	Positive[9].
Local recurrence	Positive[10].	Positive[11].	Positive[12].	Positive[13].	Positive[9].
Chemotherapy sensitivity	Negative[10].	Negative[11].	Positive[12].	Positive[13].	Positive[9].
Radiotherapy sensitivity	Negative[10].	Negative[11].	Positive[12].	Positive[13].	Positive[9].

DDLPs, dedifferentiated liposarcoma; MLPS, myxoid pleomorphic liposarcoma; MPLPS, myxoid pleomorphic liposarcoma; PLPS, pleomorphic liposarcomal; WDLPS, well-differentiated liposarcoma.

pleomorphic liposarcoma (MPLPS) is a new calcification that has been added to the 2020 WHO Classification, most commonly arising in the mediastinum, followed by the limbs and the head and neck region. Histologically, myxoid pleomorphic liposarcoma shows myxoid features (presence of a rich capillary size vascular network set in myxoid background) and pleomorphic liposarcoma (presence of pleomorphic lipoblasts). Those tumors are clinically aggressive, with a high rate of local recurrence and early metastatic spread. Genetic abnormalities have not yet been detected (Table 1).

There are few well-documented reports in the literature, and many of them are associated with tuberous sclerosis and probably correspond to angiomyolipomas. Mostly presenting with pain, hematuria, and abdominal mass, but no adrenal dysfunction was observed[14–16].

Conclusion

Retroperitoneal liposarcomas can arise by attaching to the renal tissue and can cause symptoms related to adrenal dysfunction.

Ethical approval

Not applicable.

Consent

Patient's consent was obtained for the publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

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Not applicable.

Author contribution

M.W.S.: writing the manuscript draft, collecting data. R.E.: writing the manuscript draft, collecting data. S.H.: reviewing manuscript draft. B.S.: reviewing manuscript draft. S.N.: did the procedure, reviewed manuscript draft. A.S.: did the procedure, reviewed manuscript draft.

Conflicts of interest disclosure

The authors declare no conflicts of interests.

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4. Our manuscript is a case report and no human participations were done.

Guarantor

Besher Shami.

Data availability statement

All data underlying the results are available in the article; no additional source data are required.

Provenance and peer review

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