Retroperitoneal sarcoma: Case report and review of the literature

PhD.Henri Kolani[1], Earta Gega[4], Dr.Ejona Celiku[3], Prof. Etmont Celiku[1,2]
1-Surgeon at First Clinic of Surgery- University Hospital Center "Mother Teresa" Tiranë, Tirana
2-Chief of Surgery Departement and Chief Surgeon at First Clinic of Surgery- University Hospital Center "Mother Teresa" Tiranë, Tirana
3- AnatomoPathologist Doctor at University Hospital Center "Mother Teresa" Tiranë, Tirana
4- Student of General Medicine at University of Medicine Of Tirana

Abstract
Background: The evaluation and treatment of retroperitoneal sarcomas are challenging because the tumors are relatively rare and frequently present with advanced disease in an anatomically complex location.
Method: We present the case of a 34 year old male diagnosed with retroperitoneal sarcoma. The bibliography used was taken from PubMed.
Summary: Herein, we report retroperitoneal sarcoma measuring 20X15X16 cm encasing the inferior pole of the left kidney and adherent to adjacent structures. We performed an organ-preserving surgical removal The patient was discharged after an uneventful postoperative hospital stay.A postoperative 16 month, follow-up CT was recommended to the patient.
Conclusion: The evaluation and treatment of retroperitoneal soft tissue sarcomas remain challenging. The most important factor in the long-term success in the treatment of primary tumors is complete surgical resection. It is important that patients with these tumors be evaluated and treated at centers with multidisciplinary treatment planning and expertise in treating these rare tumors. Local recurrence remains a difficult problem, with increased associated morbidity and psychological stress for affected patients. We hope that with improved education, early referral of patients with retroperitoneal soft tissue tumors will become the norm, and patients will derive the benefits of multidisciplinary evaluation and treatment of their disease.

1.Background

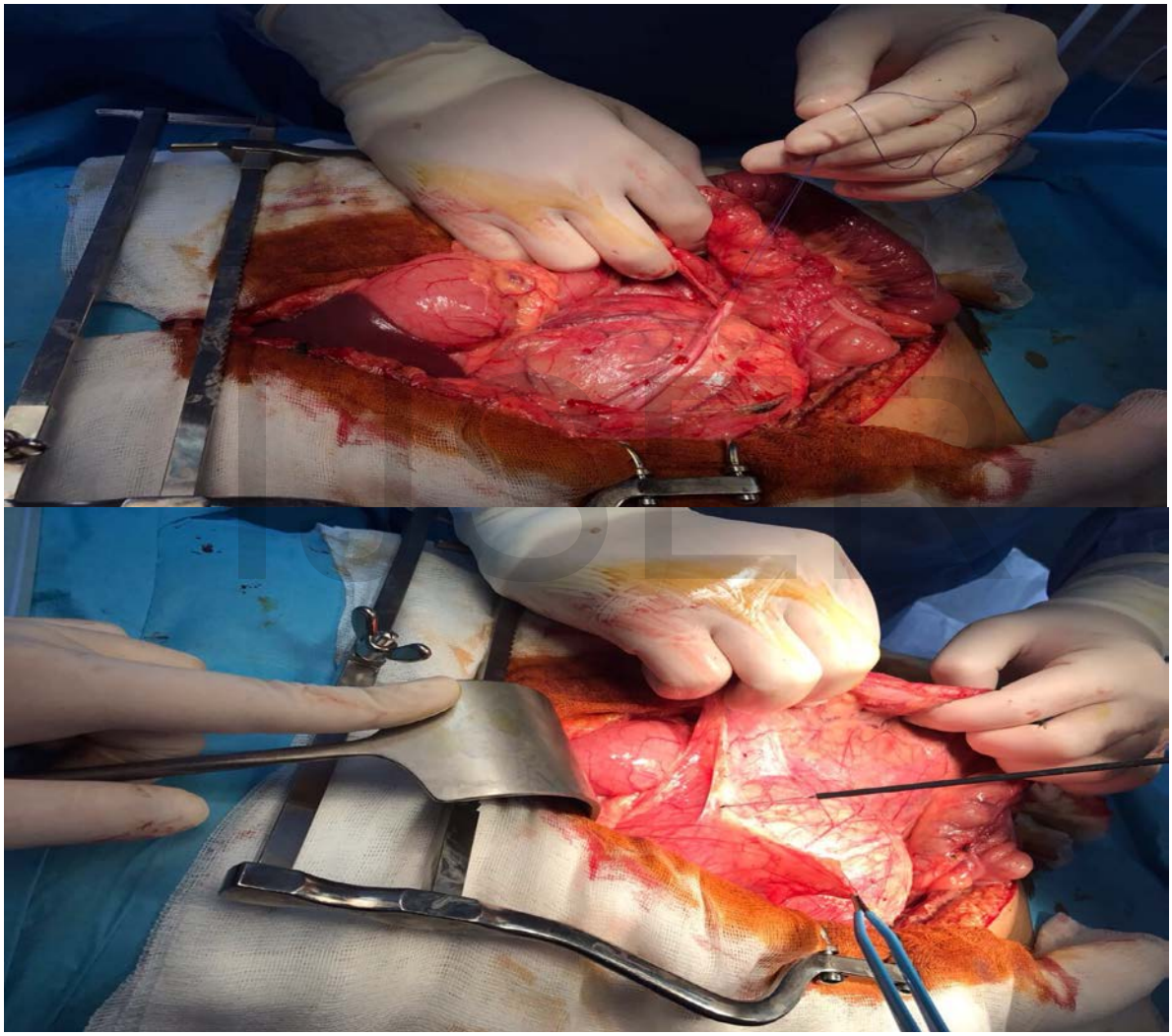
Retroperitoneal sarcoma includes a large group of tumors of mesenchymal origin arising from the retroperitoneum, with the most common histological type of retroperitoneal sarcoma being liposarcoma. Surgical resection remains the main curative treatment for retroperitoneal sarcoma, although local recurrence is the leading cause of mortality in these cases. Histopathological evaluation of the surgical margins is a major predictor of local recurrence and extended *en bloc* resection of the tumor and adjacent organs has been recommended to achieve negative surgical margins and minimize the risk of local recurrence. Although multiple centers perform *en bloc* surgical resection for retroperitoneal sarcoma, the varied anatomical locations of the primary tumor within the retroperitoneum result in local invasion to different adjacent organs, and there is no standard surgical approach or procedure for treating retroperitoneal sarcoma. However, in cases of primary retroperitoneal sarcoma at specific anatomical locations, certain surgical approaches may help shorten the operation time, reduce postoperative morbidity, achieve negative surgical margins, and prevent postoperative recurrence. These approaches could help make extended *en bloc* resection a viable treatment option for retroperitoneal sarcoma, especially at less experienced surgical centers.

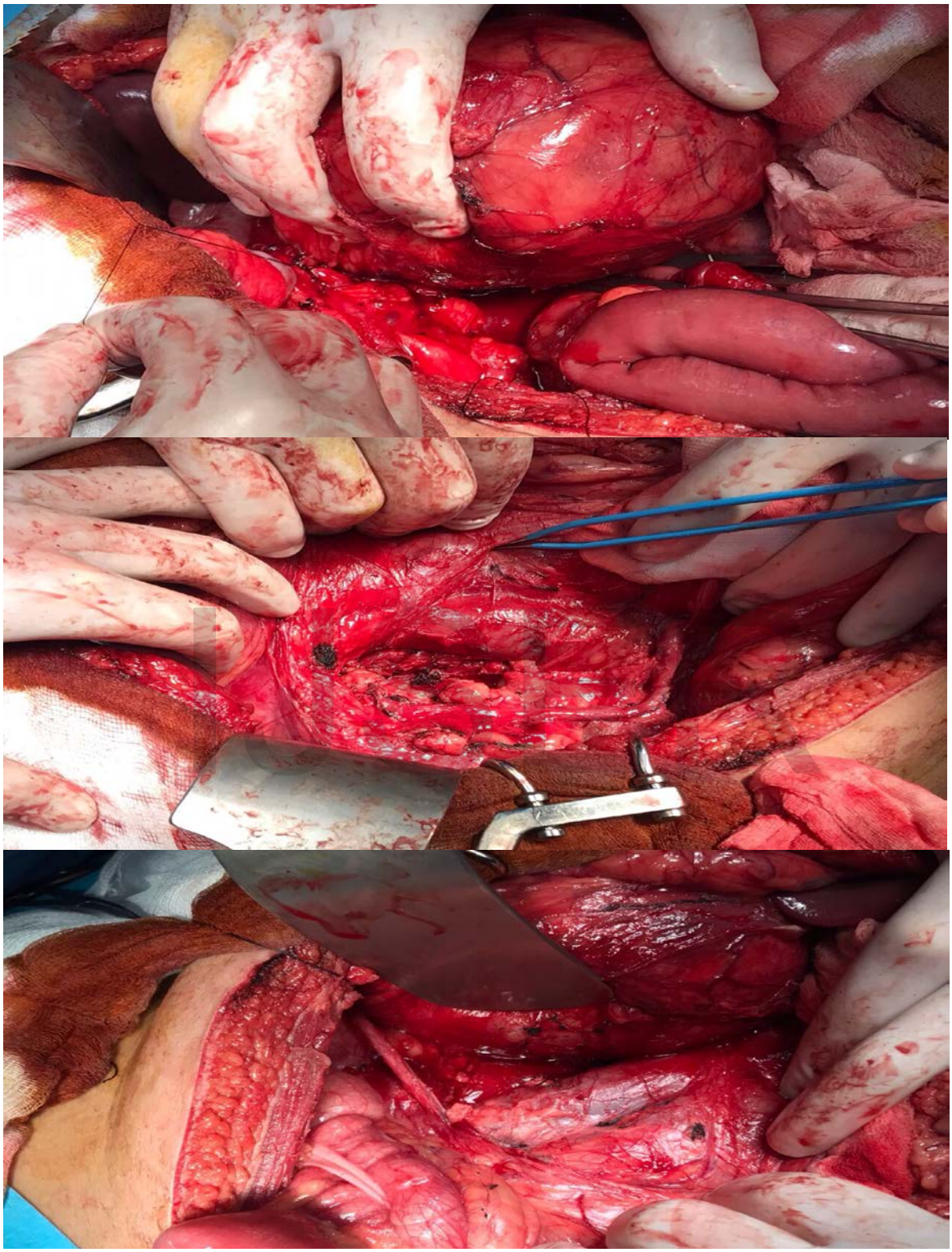
2.Case Report

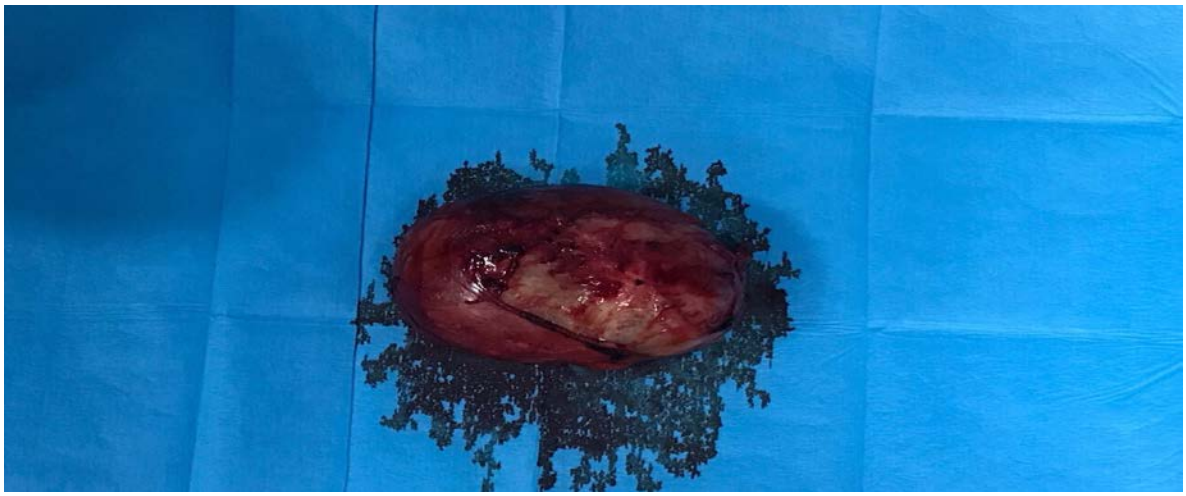
A 34-year-old man presented with complaint of progressive abdominal distension in the left iliac side. The ultrasonography it's not determinat and the radiologist suggested for a CT. The patient underwent contrast-enhanced computed tomography (CT) of the abdomen. The scan revealed a huge fatty mass originating from the retroperitoneum probably indicative of retroperitoneal liposarcoma. The CT scan also showed tumor encasement of the inferior pole left kidney. Septations and solid portions were observed within the mass. Because the patient was young, we decided to attempt organ-preserving surgery for the removal of the tumor to minimize the morbidity.

The mass was approached by making a midline incision. Adherence of the mass to the diaphragm, stomach, spleen, pancreas, and aorta could be observed. The greatest difficulty was that the tumor was encasing the inferior pole of the left kidney and adherent to the aorta. Although the left kidney was encased with the huge tumor, neither

the renal parenchyma nor the ureter was invaded. We successfully performed a salvage of the left kidney by wide excision and separated the tumor from the aorta by shaving it away, thus preserving both kidney and the aorta. The specimen measured 20 × 15 × 16 cm. The patient was discharged after an uneventful postoperative hospital stay. He was told to underwent regular follow-up examinations for 16 months after the operation.







Intraoperative photos of our case.

3.Discussion

Multiple factors, such as site and depth of origin, margin involvement after resection, and histologic grade affect survival rates for patients with sarcoma. Distant metastasis relates to the tumor size. If the size of the tumor is less than 2.5 cm, the rate of metastasis at 5 years is approximately 3%. On the other hand, the rate of metastasis at 5 years is between 55 and 60% in cases of tumors larger than 20 cm. Retroperitoneal origin is a poor prognostic factor. This is because the retroperitoneal space allows the tumor to grow to a large size before the appearance of clinical signs and symptoms. Therefore, the tumor is often diagnosed at the advanced stage. Resection margin involvement also affects prognosis. Cases with involved resection margins are characterized by a tendency towards local recurrence. The histologic subtype was significantly associated with recurrence. The five recognized histologic types are the well-differentiated, myxoid, round cell, pleomorphic, and dedifferentiated type. The well-differentiated type has good prognosis, with 5-year survival rates of approximately 90%. The myxoid type has a less satisfactory progression, and often displays early recurrence. The 5-year survival rates of the remainder of the histo-logic types are poor. High-grade sarcoma shows sensitivity to radiation therapy. However, the toxic effects of radiation therapy limit this option by primary treatment modality. The use of chemotherapy is also controversial. Research has documented little benefit from adjuvant chemotherapy in well-differentiated low-grade tumors, and partial responses in high-grade diseases in up to 50% of patients, with increased overall survival. As a result, complete surgical resection is the gold standard treatment, which might be curative. In many cases, combined resection of involved organs and vasculatures is required to achieve complete resection. Therefore, the most commonly sacrificed organ is the kidney, followed by the colon, pancreas, major vasculature, and spleen. In this study, we reported on a retroperitoneal sarcoma encasing the inferior pole of the left left kidney and adherent to adjacent structures. Furthermore, we described successful organ-preserving surgical removal and discussed the prognosis. Although there has been no evidence of recurrence to date, we will continue to observe our patient closely for recurrence, as in other previously published reports.

4.Conclusion

The evaluation and treatment of retroperitoneal soft tissue sarcomas remain challenging. The most important factor in the long-term success in the treatment of primary tumors is complete surgical resection. It is important that patients with these tumors be evaluated and treated at centers with multidisciplinary treatment planning and expertise in treating these rare tumors. Local recurrence remains a difficult problem, with increased associated morbidity and psychological stress for affected patients. We hope that with improved education, early referral of patients with retroperitoneal soft tissue tumors will become the norm, and patients will derive the benefits of

multidisciplinary evaluation and treatment of their disease.

References

1 Mack TM: Sarcomas and other malignancies of soft tissue, retroperitoneum, peritoneum, pleura, heart, mediastinum, and spleen. Cancer 1995;75:211–244.
2 Shahaji C, Amit P, Prashant P, Sachin T: Giant retroperitoneal liposarcoma: a case report. Case Rep Oncol Med 2012;4:1–2.
3 Caizzone A, Saladino E, Fleres F, et al: Giant retroperitoneal liposarcoma: case report and review of the literature. Int J Surg Case Rep 2015;9:23–26.
4 Leao P, Vilaca S, Oliveira M, Falcao J: Giant recurrent retroperitoneal liposarcoma initially presenting as inguinal hernia: review of literature. Int J Surg Case Rep 2012;3:103–106.
5 Ki EY, Park ST, Park JS, Hur SY: A huge retroperitoneal liposarcoma: case report. Eur J Gynaecol Oncol 2012;33:318–320.
6 McCallum OJ, Bruke JJ II, Childs AJ, Ferro A, Gallup DG: Retroperitoneal liposarcoma weighing over one hundred pounds with review of the literature. Gynecol Oncol 2006;103:1152–1154.
7 Kirane A, Crago AM: The importance of surgical margins in retroperitoneal sarcoma. J Surg Oncol 2016;113:270–276.
8 Nijhuis PH, Sars PR, Plaat BE, Molenaar WM, Sluiter WJ, Hoekstra HJ: Clinico-pathological data and prognostic factors in completely resected AJCC stage I–III liposarcomas. Ann Surg Oncol 2000;7:535–543.
9 Singer S, Antonescu CR, Riedel E, Brennan MF: Histologic subtype and margin of resection predict pattern of recurrence and survival for retroperitoneal liposarcoma. Ann Surg 2003;238:52–65.
10 Jaques DP, Coit DG, Hajdu SI, Brennan MF: Management of primary and recurrent soft-tissue sarcoma of the retroperitoneum. Ann Surg 1990;212:51–59..

Je m'appelle Henri Kolani. Depuis vingt ans je travaille comme chirugien général actuellement dans le Premier Clinic du Centre Hospitalier "Mère Tereza". En 2016 j'ai
finit mon dissertation du docteur. En photo avec moi est le Chef du Premier Clinique de Chirurgie et en même temps Chef du Département de Chirurgie, une figure bien connue éminence dans l'histoire de la chirurgie albanaise.