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Retroperitoneal Liposarcoma: A Concern in Inguinal Hernia Repair

M. Lechner, MD, K. Borhanian, MD, S. Mitterwallner, MD, R. Bittner, MD, E. Klieser, MD, G. Köhler, MD, K. Emmanuel, MD, F. Mayer, MD

ABSTRACT

Background and Objectives: The goal of the study was to evaluate retroperitoneal sarcomas with continuous growth into the scrotum through the inguinal canal with regard to diagnostic approach, surgical treatment, and outcome. The analysis is based on a comprehensively documented case and a complete systematic review of published literature. Potential pitfalls are highlighted.

Methods: We describe the case of a 57-year-old male Caucasian who presented with a swelling in the right groin. Suspecting a scrotal hernia, transabdominal preperitoneal plasty surgery was planned but intraoperatively a large retroperitoneal mass was revealed. After computed tomography scan and magnetic resonance imaging, a complete resection of the tumor was performed. Ten previously published cases describing the same pathology were retrieved from the PubMed database and analyzed systematically in a complete literature review.

Results: Histology showed a well-differentiated liposarcoma with tumor-free resection margins. Twenty-two months postoperatively, the patient is in complete clinical remission.

Conclusion: Preoperative clinical suspicion of retroperitoneal involvement is paramount for developing of a surgical strategy and in unclear cases demands extended preoperative diagnostic workup. Following the appropriate patient management is crucial to prognosis.

Department of Surgery, Paracelsus Medical University (Drs Lechner, Borhanian, Mitterwallner, Bittner, Köhler, Emmanuel, and Mayer), Salzburg, Austria; Department of Pathology, Paracelsus Medical University (Dr. Klieser), Salzburg, Austria; Sisters of Charity Hospital (Dr. Köhler), Linz, Austria.

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Address correspondence to: Dr. Michael Lechner, Department of Surgery, Paracelsus Medical University, Müllner Hauptstraße 48, 5020 Salzburg, Salzburg, Austria. Telephone: +43 57 2550 57832, Fax: +43 57 2550 51008, E-mail: m.lechner@salk.at

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INTRODUCTION

Spermatic-cord lipomas are a common intraoperative finding during inguinal hernia surgery. Depending on their size and shape they are often resected, sometimes reduced to the preperitoneal space, or left alone when small.

Opposed to these benign lipomas, sarcomas of the spermatic chord are rare findings with approximately 200 reported cases. Retroperitoneal sarcomas occur in 0.4/100,000/year.[1] Retroperitoneal sarcomas protruding though the groin, however, are extremely rare, with only 10 published cases over the last 30 years. Since the retroperitoneal space communicates with the inguinal canal, large lipomatous tumors, including liposarcomas, may protrude through this natural weak spot.

Typically, liposarcoma is a rare malignant entity that affects the thigh or the retroperitoneum. Histologically it can be subdivided into four subgroups in order of increasing malignancy: 1) well differentiated, 2) myxoid, 3) dedifferentiated, and 4) pleomorphic liposarcoma.[2] Liposarcoma is typically diagnosed in a late stage due to its lack of general symptoms, like fever, night sweats, and weight loss. Patients usually have normal lab results and become symptomatic by means of extrusive tumor growth and subsequent mass effects.

Inguinal liposarcoma can originate from different sites: Fatty tissue of the groin, paratesticular fatty tissue, omental tissue within a hernia sac,[3] the spermatic cord and retroperitoneal tissue that protrudes though the inguinal canal; the latter constellation is exceedingly rare. However, because of the potentially fatal consequences of a missed correct diagnosis, it is important to adhere to a different surgical approach, which is discussed in this study.

Database

We report a fully documented, extremely rare case of a low-grade retroperitoneal liposarcoma that protruded

through the inguinal canal and mimicked an irreducible scrotal hernia along with its clinical course and operative management. Published literature was systematically reviewed by using "liposarcoma and hernia" as search items in the PubMed database. Eighty publications were identified and the search extended through their citations. Only publications describing retroperitoneal sarcoma protruding through the inguinal canal, mimicking groin hernias were included. After exclusion of all publications but the ones that describe a primary retroperitoneal sarcoma protruding though the inguinal canal we thus delineated 10 cases from 1987 to 2017, analyzed them in detail, and compared the findings to the presented case.

We outline the case of a 57-year-old male Caucasian who works as a caretaker of several apartment blocks, thus carrying out hard physical work on a regular basis. On clinical examination, he was an American Society of Anesthesiologists (ASA) II patient with a body mass index of 31.5 kg/m^2, weighing 89 kg at a height of 168 cm. He reported complaints of an irreducible swelling in the right groin with discomfort over the course of 1 year. On physical examination, the swelling was confirmed and its contents protruded into the right scrotum; in addition a small umbilical hernia was detected. The ultrasound report stated a pathological finding, consistent with a large "lipomatous" hernia, descending into the scrotum with a defect size of 2 cm. The hernia sac was "too large to be displayed on one screen." The content was "not clearly distinguishable but likely intraperitoneal fatty tissue." Only because of the patient's demand of swift postoperative resilience and due to the combined hernia findings, an endoscopic mesh-augmented hernia repair by transabdominal preperitoneal plasty was scheduled, as opposed to standard Lichtenstein procedure that might have been favored under either technologically and economically less favorable circumstances or in outpatient settings.

Intraoperatively, a large lipomatous extraperitoneal mass without any hernia defect was visible **(Figure 1)**. As recommended by the Trans-Atlantic Retro Peritoneal Sarcoma (RPS) Working Group[4] when the tumorous mass was found, no further steps were taken for assessment or exploration at the time of initial surgery. However, it was noted that no apparent infiltration of neighboring organs was present. The procedure was aborted and a computed tomography scan **(Figure 2)**, as well as, as well as magnetic resonance imaging **(Figure 3)** were carried out, which revealed a large extraperitoneal, lipomatous tumor extending through the inguinal canal. After interdisciplinary discussion, a primary complete curative (R0) resection was aimed for. Intra-operative radiotherapy was discussed

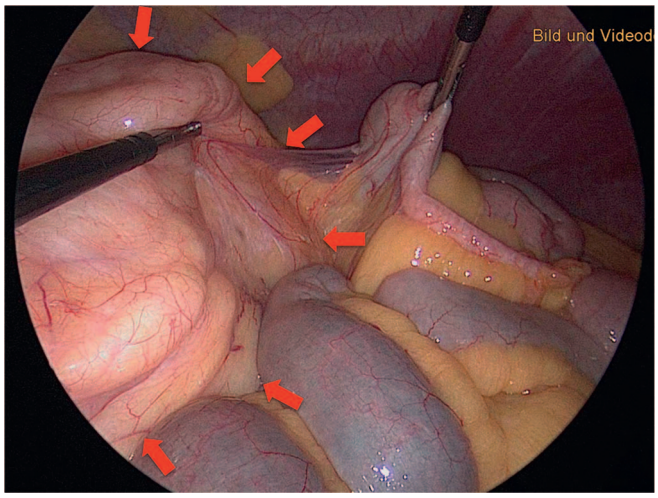

Figure 1. Intraoperative laparoscopic view of retroperitoneal tumor, tumor margins outlined by arrows.

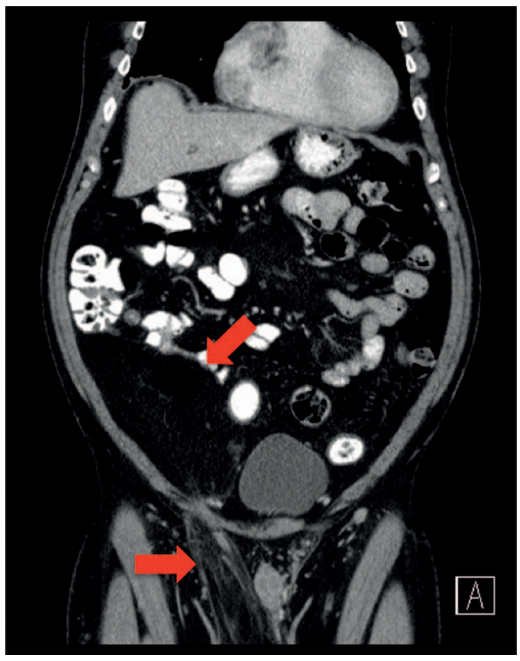

Figure 2. Preoperative computed tomography scan showing retroperitoneal and scrotal mass (arrows).

but ruled out due to vast tumor size. The treatment plan was early elective laparotomy with complete tumor resection, followed by Lichtenstein repair of the resulting defect since even after extensive information about the suspected condition the patient did not consent to any mutilating procedures, including orchiectomy. Therefore, the patient was not referred to a sarcoma center and surgery was carried out as planned.

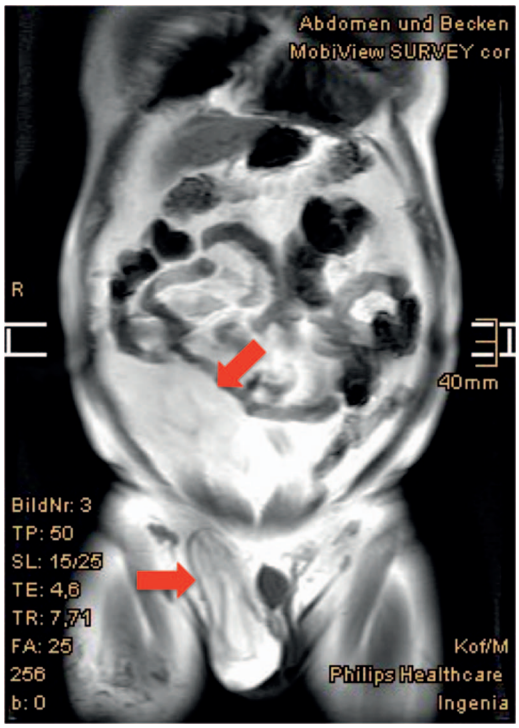

Figure 3. Preoperative magnetic resonance imaging scan showing retroperitoneal and scrotal mass (arrows).

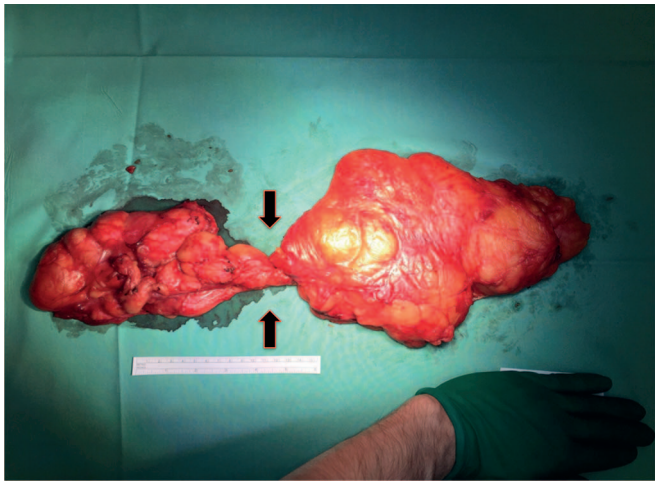

Figure 4. Specimen with retroperitoneal and scrotal part after resection (arrows show narrowing of tumor in the inguinal canal).

RESULTS

After carrying out the surgery a 1.261-g specimen of 46 × 18× 6 cm in size with a marked isthmus, separating the retroperitoneal from the inguinal-scrotal part of the tumor **(Figure 4)** was retrieved. The initial pathology-report

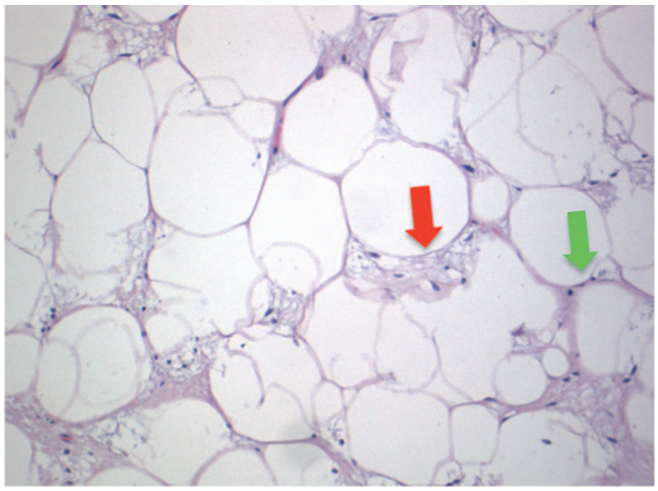

Figure 5. Microscopic view (magnification 400x): most adipocytes have one vacuole and an eccentric nucleolus (shown by green arrow). However, some appear multivacuolated and small with a concentric nucleolus (shown by red arrow)..

described a lipomatous tumor, namely a lipoma. Further histopathological and immunohistochemical workup of the specimen was performed only "due to tumor size and location." The definitive pathological diagnosis was significantly delayed due to complex workup. Morphology and immunohistochemistry did not lead to a result. Eventually fluorescent in-situ hybridization with Mouse Double Minute 2 (MDM-2) and Cyclin dependent kinase-4 (CDK-4) sensor showed an amplification and only after consultation with a sarcoma expert of another university hospital, revealed a well-differentiated (G1) retroperitoneal liposarcoma, R0 in multiple examined margins **(Figure 5)**. At a clinical and radiological followup through computed tomography scan 22 months postoperatively, the patient was free of recurrence. Indefinite followup is scheduled.

The structured literature review, according to the search criteria described in the methods section, yielded 10 cases, which were reviewed in detail and comprise all publications over 30 years from 1987[5] to 2017. Mean patient age was 58.5 years (53[6] to 86[7] years). All documented patients were male. Mean onset of symptoms was 1 years (7 months[8] to 5 years[9]) before presentation. Side of the body was almost evenly distributed with 5 right-sided[6–8,10] and 4 left-sided[9–13] manifestations. Three cases appeared as an incarcerated hernia,[11,14] of note one tumor appeared as a reducible hernia[8]; the others were not documented. In 5 cases a computed tomography scan was performed preoperatively[6,9–11,13]; notably none of them was conducted

due to the combined suspicion of an inguinal hernia and a retroperitoneal tumor.[13]

On histological examination, most specimen was diagnosed as well-differentiated liposarcomas, comprised of 7 cases.[5,7,8,10–12] The large tumors' weight ranged widely from 1.26 kg in the presented case up to 42 kg.[9] R0 resection margins are documented in only 2 cases after primary surgery.[9] Adjuvant therapy was administered in only 3 cases[6,9,13]; one patient received radiotherapy after resection of a 42-kg tumor.[9] Two patients underwent chemotherapy, one due to a synchronous existing lymphoma,[6] the other one for positive resection margins in a dedifferentiated pleomorphic sarcoma.[13] In two cases liposarcoma was resected with positive margins and postoperatively a second surgery was planned to achieve R0 resection.[7,8]

Recurrence rates for retroperitoneal tumorous lesions of up to 91% are published.[14] In 2 of the analyzed cases, recurrence occurred after 12[7] and 18 months,[15] respectively. The longest documented followup at the time of publication was 57 months.[8]

DISCUSSION

Inguinal hernia surgery is performed in large numbers around the world, often considered a minor procedure and therefore commonly left to surgeons still in their learning curve. In addition to variable levels of surgical experience, laparoscopic techniques are not universally available. The combination of these facts poses a significant risk for missing the correct diagnosis for the rare constellation of a retroperitoneal tumor protruding through the inguinal canal and thus mimicking a hernia.

Even though the case presented appears typical of the condition described when compared to the literature, the connection between the clinical appearance of a hernia and a large retroperitoneal tumor was not initially made. As in all other published cases the diagnosis was not attained directly. Together with a high level of clinical suspicion, a structured approach to the condition in accordance with the Trans-Atlantic Retro Peritoneal Sarkoma (RPS) Working Group guidelines[4] was established in our department:

1. Intraoperative incidental diagnosis of retroperitoneal tumors has to be avoided through thorough preoperative workup.
2. If in any hernia, regardless of reducibility, its sac, or content cannot clearly be delineated, preoperative routine sonography of the groin must be extended to the retroperitoneum.
3. In case of unusual retroperitoneal sonographic findings, computed tomography scans or magnetic resonance imaging should be performed to clarify the findings.
4. Upon discovery of a retroperitoneal mass, the further investigation of the tumor must be prioritized over the initially suspected hernia, regardless of other factors, namely local discomfort, urging early surgery.
5. Since malignancy is common in retroperitoneal masses, interdisciplinary discussion, preferably in a sarcoma board, must be sought. Further management may well include core needle biopsy and planning of wide tumor excision, preferably at a dedicated sarcoma center.
6. When, however, a large, retroperitoneal tumor mass and no visible hernia is found intraoperatively during an attempt of laparoscopic hernia repair, the procedure is to be aborted without further assessment, exploration, or biopsy. The further management of the case can then safely be organized in accordance with published guidelines for workup of retroperitoneal tumors.[4]
7. If, on the other hand, operating surgeons are surprised by these findings during attempted open inguinal hernia surgery, it must be assumed that the tumor's integrity has been compromised. A strategic withdrawal in this situation with a patient usually unconsented for the necessary further extension of the surgical procedure is far more challenging. Even more serious, the inguinal-scrotal part of the tumor could be mistaken for a common lipoma and simply be resected without further exploration or thought of an underlying malignancy.
8. Therefore, if after sectional imaging any doubt remains regarding the inguinal pathology; for example, in the presence of an only small retroperitoneal tumor component, a laparoscopic exploration, and if indicated, transabdominal preperitoneal plasty repair should be favored over an open access. Total extraperitoneal hernia repair surgery does not allow for intraabdominal exploration and bares a high risk of disrupting tumor integrity and should therefore be avoided in this specific scenario.
9. When dissection of an unusual lipomatous mass from the spermatic chord has already occurred, the pathologist has to be informed specifically about the clinical suspicion to initiate appropriate workup. High variability on immunochemistry, nonspecific markers, and very subtle histological differences between lipomas and liposarcomas make extensive analysis necessary. Ade-

quate histology has been described as demanding to obtain.[9] This is also reflected in the case presented. In light of this case, the value of previously promoted intraoperative frozen sections[16] remains controversial.
10. Followup for these patients should be life long.[4]

Contrary to published recurrence rates for retroperitoneal tumorous lesions of up to 91%,[5] the literature review formally revealed only 2 cases of recurrence. This discrepancy is to be interpreted in view of a potential bias trough the small number of cases owed to the scarcity of the condition. In addition, the high proportion of initial R1 resections and the short or undocumented follow-up periods may well contribute to the mismatch.

Limitations of the study comprise the small number of traceable cases, the often incomplete documentation and short published followup. Strongpoints are the completeness of documentation of the case presented. Furthermore, the analysis represents the most detailed and structured review of the largest case series to date.

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