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Case report

Idiopathic giant unilocular retroperitoneal cyst in a 38-years-old female: Diagnostic and management challenge in resource-limited health facility—A rare case report

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ABSTRACT

Background: Retroperitoneal cysts are rare, accounting for less than 1% of retroperitoneal tumors. Idiopathic giant unilocular retroperitoneal cysts are even more uncommon, with very few reported cases. Their diagnosis and management are particularly challenging in resource-limited settings due to restricted access to advanced imaging and surgical techniques.

Case presentation: A 38-year-old female presented with progressive abdominal distension. Imaging revealed a giant retroperitoneal cyst. Due to limited resources, an open laparotomy was performed, and a 33 cm cyst containing 8 l of clear fluid was completely excised. Histopathology confirmed a giant idiopathic retroperitoneal cyst.

Discussion: This case is unique due to the cyst's idiopathic nature, massive size, and successful surgical management despite resource constraints. Complete surgical excision remains the definitive management to prevent recurrence and complications.

Conclusion: This case highlights the importance of surgical intervention in managing giant retroperitoneal cysts in resource-constrained settings, ensuring favorable outcomes despite diagnostic limitations.

1. Introduction

Retroperitoneal cysts are extremely rare, with an incidence ranging between 1 in 5750 and 1 in 250,000 hospital admissions. They originate from embryological remnants, including mesothelial, lymphatic, urogenital, or enteric structures. While most retroperitoneal cysts have a defined histological origin [1,2]. Idiopathic giant unilocular retroperitoneal cysts lack an identifiable epithelial lining, making their etiology uncertain [3].

Giant unilocular retroperitoneal cysts are extremely rare, with only few cases documented. They can manifest as a palpable mass in the abdomen and may pose challenges in diagnosis when compared to other retroperitoneal masses originating from organs like the liver, kidney, and pancreas [4,5]. Idiopathic giant unilocular retroperitoneal cysts are infrequently reported, making this case a valuable addition to medical literature.

Majority of cases are detected incidentally due to their asymptomatic nature and present with nonspecific symptoms such as abdominal distension, vague discomfort, or compression effects on surrounding organs as to our case [7]. The lack of characteristic, clinical features makes early diagnosis difficult, and in resource-limited settings, where access to advanced imaging modalities is restricted, preoperative diagnosis is even more challenging [5,6].

Complete surgical excision of these cysts remains the gold standard for both diagnostic confirmation and treatment. However, in low-resource settings, limitations in intraoperative settings like frozen sections, histopathological studies, and imaging may delay definitive diagnosis and appropriate management planning. This case report highlights the diagnostic and management challenges of an idiopathic

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Abbreviations

CT-Scan	Computed Tomography Scan
KCMC	Kilimanjaro Christian Medical Centre

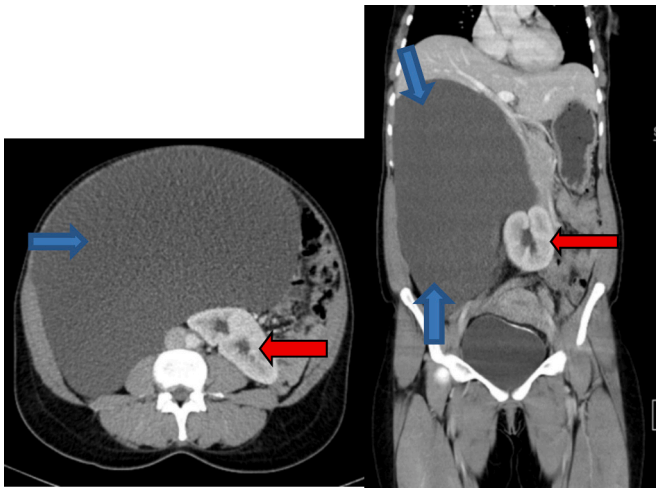

Fig. 1. A contrast CT abdomen and pelvis – axial and coronal views demonstrate a large thin-walled right suprarenal cyst (blue arrows) measuring approximately 23.5 cm (AP) x 20.3 cm (T) x 32.2 cm (CC) in size with a volume of 8 l. The large cyst displaces the right kidney (red arrows) inferomedially across the midline. The cyst exerts pressure effect on the liver, gall bladder, pancreas and small bowels. It extends into the right iliac fossa. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

giant unilocular retroperitoneal cyst in a 38-year-old female, emphasizing the importance of clinical judgment and surgical intervention in resource-limited settings [6].

This case report was prepared following the SCARE Guidelines for reporting surgical case reports to ensure completeness and transparency in reporting [8].

2. Case presentation

A 38 year old woman was admitted in the surgical ward November 2022 with the chief complain of progressive abdominal swelling which was associated with mild abdominal pain, and early satiety for five months. She had no systemic symptoms such as weight loss, constipation, diarrhea, craving for salty foods, easy fatigability or headache.

On examination, she appeared well-nourished but had gross abdominal distension, mild pale, afebrile with sclera tinge of jaundice.

Abdominal palpation revealed a large, non-tender, intra-abdominal mass occupying the entire lower abdomen and extending into the upper abdomen with no signs of peritoneal irritation or organomegaly.

Blood tests showed White cell count (5.7×10^9/l), Haemoglobin (9.8 g/dl), Bilirubin (103 mg/dl) with normal platelet counts (393×10^9/l. Liver enzymes, AST (24.0 U/l, ALT (21.0 U/l), albumin 42.7 g/l. Serum creatinine (53 μmol/l) and ESR (80 mm/h). All other blood tests were normal. An abdominal ultrasound revealed a complex abdominal cyst, both kidneys were normal in size and normal echotexture, the liver appeared normal. Contrast CT abdomen and pelvis – axial and coronal views Fig. 1, demonstrate a large thin-walled right suprarenal cyst (blue arrows) measuring approximately 23.5 cm (AP) x 20.3 cm (T) x 32.2 cm (CC) in size with a volume of 8 l. The large cyst displaces the right kidney (red arrows) inferomedial across the midline and exerts pressure effect on the liver, gall bladder, pancreas and small bowels. It extends into the right iliac fossa. Since there was a tinge of sclera jaundiced, but the mass was separately from the liver, pancreas, biliary trees and the right kidney. Given the size of the cyst and its compressive effects, surgical intervention was planned. An open surgical approach was chosen.

The patient underwent laparotomy. Under general anaesthesia, an abdominal extended midline incision was made, after opening abdominal wall layers, a huge retroperitoneal cyst was identified Fig. 2, that was occupying the entire part of an abdominal cavity.

The cyst was opened to get access for fluid suctioning as demonstrated in Fig. 3. Cystectomy was successfully done, about 8 l of brown-coloured fluid was sucked out from the mass. Cystic wall and fluid samples were sent for histology.

The patient had an uneventful recovery and was discharged on postoperative day 7. Follow-up at 6 and 12 months showed no recurrence or complications. Cytological examination of the sample reported presence of amorphous material, there was no evidence of malignancy. Histological results Fig. 4 (A and B), the cystic wall was reported to have adrenal tissue without atypia, there was cystic wall infiltration by mononuclear inflammatory cells. The inner surface of the wall was not lined by recognizable specific cells. Regarding the intraoperative clinical findings where by the origin of the cyst was uncertained, the findings were more suggestive of a giant idiopathic retroperitoneal cyst.

3. Discussion

Idiopathic retroperitoneal cysts, as seen in this case, are extremely rare, with fewer than 100 cases reported in the literature. The exact etiology of idiopathic retroperitoneal cysts remains unclear. Some theories suggest congenital remnants, degenerative changes, or lymphatic obstruction as potential causes [9]. Most cases remain asymptomatic until the cyst enlarges significantly, leading to symptoms like abdominal distension, pain, or compression of adjacent structures as compared to our case. In most cases, following interventional radiology, image-guided procedures help to achieve different pathological diagnosis in these challenging lesions. This offers the chance of an appropriate treatment; however, the overall clinical assessment of retroperitoneal cysts is highly demanding [10].

3.1. Diagnostic challenges in resource-limited settings

Retroperitoneal cysts present a diagnostic dilemma due to their rarity and nonspecific clinical manifestations. In resource-limited facilities with unavailability of advanced modalities, reliance on clinical examination and basic imaging including ultrasound and CT scan becomes essential [6,10]. This often leads to diagnostic uncertainty, necessitating exploratory surgery for definitive diagnosis. Retroperitoneal cysts are classified based on their presumed embryological origin. Common types include lymphatic (lymphangiomas), mesothelial (mesothelial cysts), enteric (duplication cysts), urogenital, and non-classifiable (idiopathic) cysts. Retroperitoneal cystic lesions are not commonly seen and are considered a rare surgical condition. They can be classified as either neoplastic or non-neoplastic cystic lesions, and they may or may not cause symptoms [2,11]. Neoplastic lesions include various types of cysts such as cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, mullerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst, pseudomyxoma retroperitoneal, and perianal mucinous carcinoma. Non-neoplastic cysts include pancreatic pseudocyst, non-pancreatic pseudocyst, lymphocele, urinoma, and hematoma [11].

3.2. Differential diagnoses

Given its location, a broad range of differential diagnoses had to be considered, including, these include adrenal cysts. Typically present with endocrine dysfunction or hemorrhage, but these was unlike to our

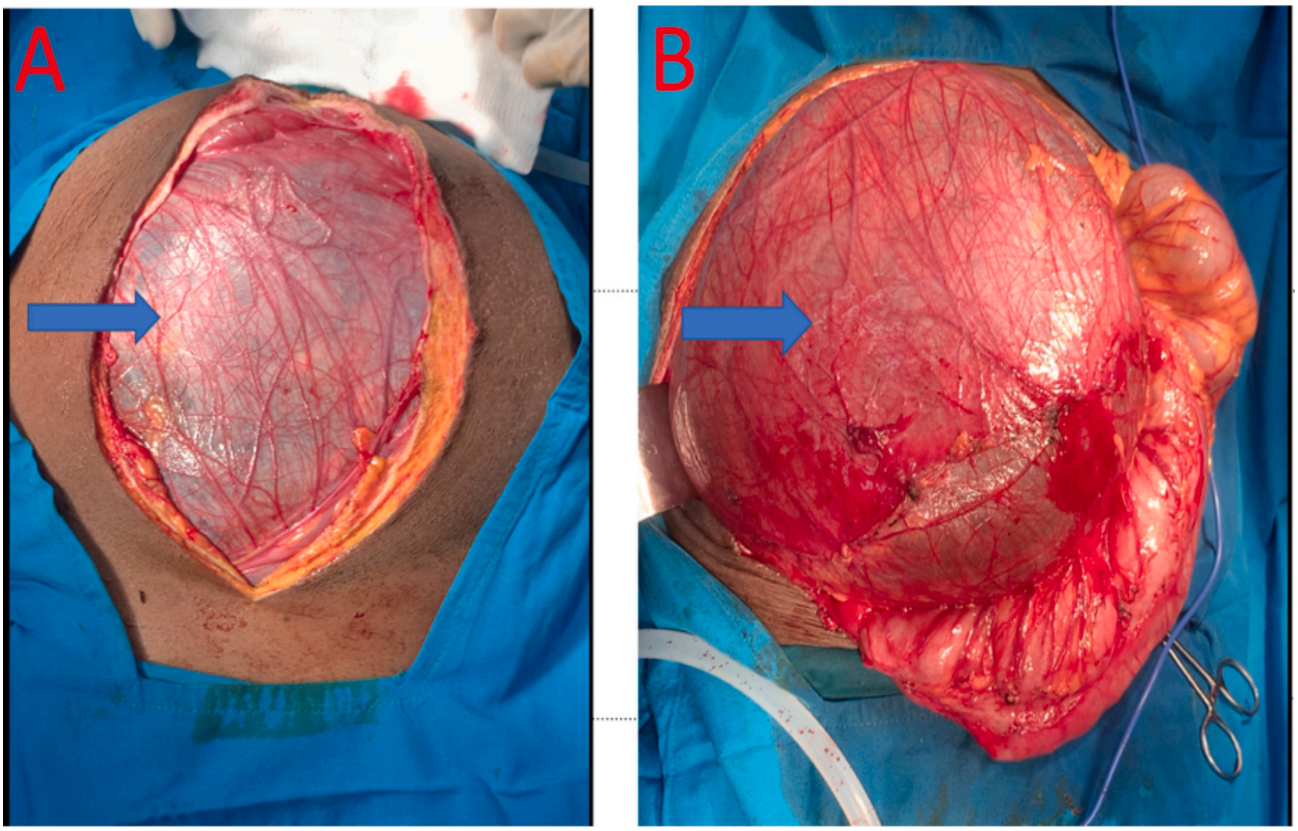

Fig. 2. Blue arrows in image A and B show cystic mass occupying the entire abdominal cavity following laparotomy through an extended midline incision. Image B maximum exposure after retractionof the abdominal wall. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

case as hormonal assays were normal. Lymphangiomas, these are common in pediatric population and they are often multilocular, whereas this cyst was unilocular. Renal Cysts were completely ruled out as the cyst was not connected to the kidney. Mesothelial cysts also were ruled out since they usually have an epithelial lining, which was absent in this case.

3.3. Uniqueness and significance of this case

This case is unique due to the nature and presentation. The patient had no systemic symptoms; the cyst was idiopathic, lacking an identifiable embryological origin, making it extremely rare. The cyst looked giant with the size more than 30cms without significant symptoms is unusual. The case highlights the critical role of surgical intervention in both diagnosis and treatment when preoperative workup is inconclusive. Complete surgical excision is essential as compared to our case to prevent recurrence and complications like infection, hemorrhage, or rupture [6,9]. The decision to excise the cyst rather than marsupialize or fenestrate it was based on minimizing recurrence risk. Laparotomy is the best choice that allows better access and preventing accidental spillage, it reduces chance of leaving behind part of the cyst. But also, minimal invasive procedures have similar outcome, the larger cysts must be aspirated prior to removal allowing better excision and permit mobilization [14]. Aspiration alone risks recurrence and infection. In this case, complete cyst removal was curative, and no recurrence was observed at one-year follow-up. Several case reports described giant retroperitoneal cysts measuring 35 cm reported to be managed laparoscopically, others described cases requiring aspiration before laparotomy [12,13]. Our Case is unlike with the previous cases, this cyst was excised via open surgery in a resource-limited setting with excellent long-term results.

4. Conclusion

This case highlights the rarity of idiopathic giant unilocular retroperitoneal cysts and the challenges in diagnosing and managing such conditions in resource-limited settings. Since their diagnosis is challenging, they are difficult to predict or detect preoperatively. Due to their rarity and the absence of preoperative histological confirmation, surgical excision remains the cornerstone of both diagnosis and treatment. Despite constraints in imaging and surgical resources, cysts are mostly found in routine ultrasound, CT-scans, or MRI studies.

Strengthening diagnostic capacities through improved imaging and cytological analysis would enhance preoperative decision-making, reducing unnecessary surgical interventions. However, in cases where definitive diagnosis is uncertain, timely surgical excision remains the best approach to prevent complications and ensure optimal patient outcomes. This case contributes to the limited literature on idiopathic retroperitoneal cysts and underscores the feasibility of managing complex surgical conditions in low-resource environments.

SCARE guideline

The work has been reported in line with the SCARE criteria.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and the accompanying images. A copy of the written consent is available for review by the corresponding author of this journal.

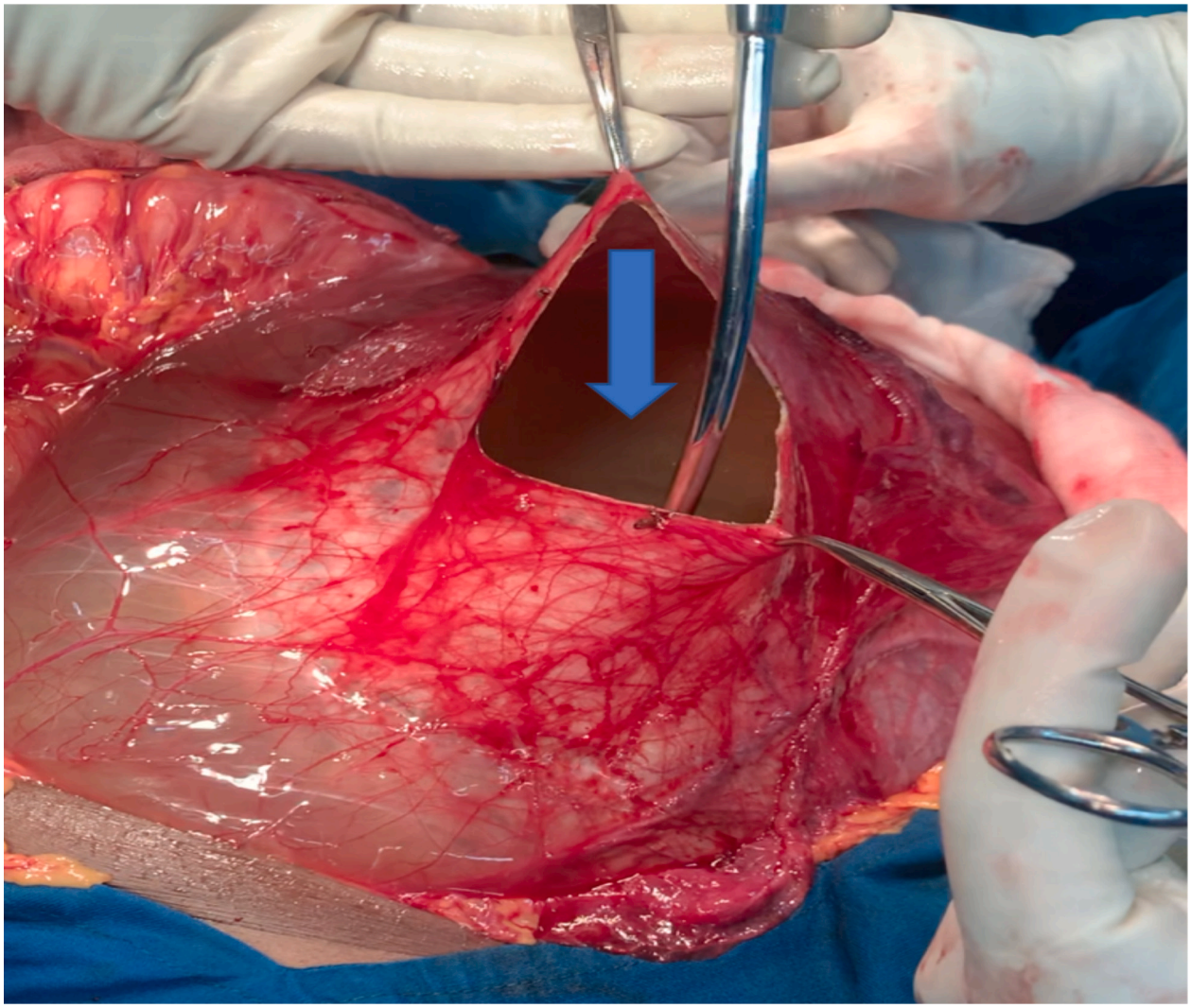

Fig. 3. Shows an opened cystic mass, containing approximately 8 l of brown-coloured fluid (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

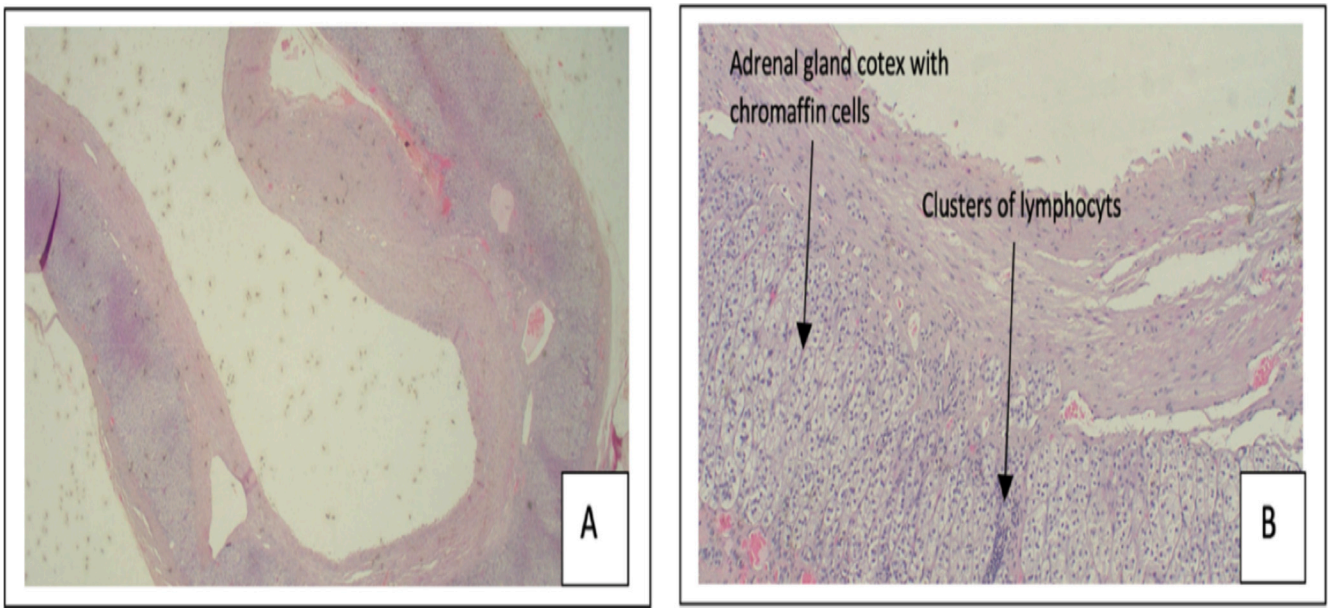

Fig. 4. An adrenal pseudocyst (A and B). H and E section with x20 and x100 magnifications respectively. The sections show a wall of a cystic lesion. The wall shows adrenal tissue without atypia but there is wall infiltration by mononuclear inflammatory cells. The inner surface is not lined by recognizable specific cells.

Ethical approval

Ethical clearance was not necessary at our hospital for a single case report.

Guarantor

All authors in the article accept full responsibility for the work, have access to the patient's information, and decide to publish.

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Declaration of competing interest

All authors declare that there are no conflicts of interest. All authors participated fully during perioperative care, drafting and approval of the article.

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Data availability

The datasets of the present study are available from the corresponding author upon request.

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