Huge pelvic retroperitoneal cyst masquerading as hydrosalpinx: A case report with review of the literature

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ABSTRACT

Primary retroperitoneal cysts (RPCs) are a rare surgical entity and are mostly detected incidentally. Usually, they are asymptomatic. At times, they may attain a huge size and may present with a large abdominal lump. Often, they pose a dilemma at diagnosis and for management. Though the laparoscopic approach has been described for their surgical excision, open surgery is still the preferred approach. We herein present a case of a voluminous retroperitoneal pelvic cystic mass in a 40-year-old female, which was diagnosed as hydrosalpinx on a magnetic resonance imaging scan. The patient was referred to us by the specialist in gynecologic laparoscopy after the incidental discovery of the RPC during his surgical intervention, purportedly for large hydrosalpinx. At our hospital, 4 days after the above intervention, she underwent laparoscopic complete excision of the cyst. Her postoperative recovery was uneventful and she was discharged on postoperative day 3 without any complications. Histopathology was suggestive of Mullerian inclusion cyst. This case report aims to share a rare case of a large primary retroperitoneal pelvic cystic lesion, which caused a diagnostic challenge preoperatively but was eventually managed successfully, laparoscopically.

Keywords: Hydrosalpinx, laparoscopy, Mullerian cyst, retroperitoneal cyst

Case Report

A 40-year-old female with no comorbidities was referred to us by the gynecologist due to incidental discovery of a voluminous retroperitoneal pelvic cystic mass in a 40-year-old female, which was diagnosed as hydrosalpinx on a magnetic resonance imaging scan. The patient was referred to us by the specialist in gynecologic laparoscopy after the incidental discovery of the RPC during his surgical intervention, purportedly for large hydrosalpinx. At our hospital, 4 days after the above intervention, she underwent laparoscopic complete excision of the cyst. Her postoperative recovery was uneventful and she was discharged on postoperative day 3 without any complications. Histopathology was suggestive of Mullerian inclusion cyst. This case report aims to share a rare case of a large primary retroperitoneal pelvic cystic lesion, which caused a diagnostic challenge preoperatively but was eventually managed successfully, laparoscopically.

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revealed an anteverted uterus and a minimally tender cystic mass in the pouch of Douglas. She was then subjected to biochemical and radiological investigations. Routine laboratory investigations were within normal limits. Tumor markers including carcinoembryonic antigen (CEA) 1.8 ng/ml, CA 19-9 (15 U/ml), CA-125 (14.6 U/ml), and alpha-fetoprotein (1.1 IU/ml) were measured to assist the diagnosis and were within the normal range. An ultrasonography (USG) scan of the abdomen revealed a large (12.4 × 10.2 × 10 cm) multiloculated, thick-walled cystic mass with low internal echoes noted posterior and left to the uterus separate from both the ovaries, suggesting hydrosalpinx with mild hypervascularity. A magnetic resonance imaging (MRI) scan of the pelvis confirmed the USG findings. The cystic mass was abutting against the rectum and uterus [Figure 1]. The patient was posted for a diagnostic laparoscopy and proceed, keeping in mind the 2 differential diagnoses: i) large hydrosalpinx and ii) large adnexal cyst. At laparoscopy, a large retroperitoneal multiloculated cystic mass measuring 12 × 10 cm was noted more towards the left midline next to the left rectal wall. The mass appeared thick-walled and completely separate from the uterus. Both the ovaries and fallopian tubes were normal and separate from the mass [Figure 2a, b]. Given the uncertain origin of the retroperitoneal mass, the procedure was abandoned and the patient was referred to us. A physical examination revealed a normal soft abdomen with no palpable lump, no organomegaly, and no clinical evidence of free fluid. A per-rectal examination revealed a smooth, cystic, nontender extrinsic mass through the anterior and left lateral walls. We then posted the patient for laparoscopic cyst excision, 4 days after the initial gynecologic intervention. At laparoscopy, the previous findings were confirmed. The peritoneum over the cyst was incised to enter the retroperitoneal space. The cyst was separated from the lateral pelvic wall on the left side and the sigmoid mesocolon and rectum on the right side [Figure 3a-c]. With the caudad progression of the dissection on both sides, the cyst was finally separated from the pelvic floor, thereby freeing it entirely [Figures 3d and 4a]. This being a large cyst with no solid component within, we punctured the cyst and aspirated the contents (transparent, watery fluid), taking due precautions to avoid spillage [Figure 4b]. The fluid was sent for routine microscopy, culture, and cytology. The specimen was retrieved in a plastic bag through the enlarged subumbilical camera trocar site [Figure 4c]. The patient tolerated the procedure well. The postoperative recovery was uneventful and she was discharged on the third postoperative day. Fluid analysis revealed uninfected, transudative nature with no malignant cells. Histopathological examination of the cyst wall was suggestive of Mullerian inclusion cyst [Figure 5a, b]. At the time of writing this paper, 1.5 years after our surgery, a telephonic interview was conducted with the patient. She remains asymptomatic till the present day.

**Discussion**

Primary RPCs, defined as those cysts lying in the retroperitoneal fibro-fatty tissue that have no apparent connections with any adult anatomical structure except by loose areolar tissue, are
rare. RPC masses pose an important diagnostic and therapeutic dilemma to the treating physicians.[9] These develop due to developmental errors during embryogenesis.[5] Based on histologic and embryological division, these cysts are classified as follows:[6]: 1) urogenital cyst: these cysts can appear from the traces of embryonic urogenital structures and are further divided into pronephric, mesonephric, Mullerian cyst, tailgut cyst, and duplication cyst, 2) lymphatic cyst: these cysts can arise from the lymphatics of the intestine, e.g., chylous cyst or lymphangiomas, 3) inclusion cyst: this cyst represents a dermoid cyst that contains sebaceous material, 4) parasitic cyst: most common type is hydatid cyst, and 5) traumatic cyst: these are usually sequelae of a post-traumatic hematoma.

Approximately one-third of the lesions are asymptomatic. However, they become symptomatic once they attain a significant size, which causes a mass effect on the surrounding structures.[9] The patient may present with chronic, vague, nonspecific abdominal pain, distention, back pain, weight loss, lower limb pain, or numbness due to compression of the vessels. Accurate preoperative diagnosis of a RPC remains elusive and poses a great challenge. The diagnostic workup includes careful history taking, a thorough physical examination, routine blood investigations, tumor markers (CEA, CA 125, CA 19-9, and alpha-fetoprotein), and imaging. The cystic fluid if aspirated should be sent for routine microscopy, culture, and cytology to determine the nature of the fluid. However, controversy exists about this and the role of preoperative histological analysis of the cyst walls. The radiologic investigations include the USG abdomen, contrast-enhanced computed tomography (CECT) scan, and/or MRI of the abdomen. Despite the use of the whole diagnostic armamentarium, sometimes these tests do not allow us to unambiguously identify the lesion.[3] Imaging modalities can help us to identify the shape, size, and location of the cyst along with wall thickness, septa, calcification, and fat content. In our case, the MRI pelvis failed to identify the exact nature of the cyst, misdiagnosing it as a hydrosalpinx.

Surgical interventions are indicated when these cysts become symptomatic and when inherent complications such as infection, perforation, or malignancies are suspected. Preferred surgical intervention is through an open approach. However, we present this case as a unique case that was managed laparoscopically. Diligent and complete surgical excision of the cyst with preservation of the surrounding structures remains the gold standard approach. Other less acceptable options are therapeutic aspiration, surgical marsupialization, fenestration, and partial excision.[8,9] In these options, a high recurrence rate is always a cause of major concern. Retroperitoneoscopic surgery is limited to small lesions located in the retroperitoneal region. An intraoperative frozen section is advisable especially when the cyst cannot be excised completely.[10]

A review of the literature shows that being a rare condition, there are very few published case reports/series/reviews on large RPCs and most of them were treated with an open approach. We managed to treat the case with a laparoscopic approach [Table 1].[3,11-21]

Although hydrosalpinx and ovarian cyst are common clinical entities, RPCs are rare. Yet radiological appearances of all these mimic each other, sometimes. Primary care physicians being the first points of contact for patients in many health care systems around the world, have to deal with many such clinical and investigational dilemmas in their day-to-day practice. For them, this paper underscores the importance of carefully discerning multiple differential diagnoses (common and rare), knowing classical imaging appearances, and finally, most importantly, the multispecialty team approaches delivering optimum patient care.

Conclusion

Primary RPCs are rare and have varied presentations. CECT scan is the preferred imaging modality. However, there is a substantial overlap of CECT findings in various RPCs, thereby making an accurate preoperative diagnosis difficult. Complete cystectomy

Figure 4: a) Trilobed cyst in 1 whole piece (black asterisks), freed from all its attachments and ready for extraction. b) Cyst being aspirated carefully to avoid spillage (white arrow), prior to retrieval. c) Cyst wall introduced into a retrieval bag and ready for extraction (white arrow)

Figure 5: Cyst wall lined by simple columnar to cuboidal ciliated epithelium: a) low-power field and b) high-power field
Table 1: Review of the Literature

| Authors [Ref. no.] | Journal (Yr. Of publication) | Study type (No. of pts.) | Mode of surgery | Size | Materials and methods/Imaging appearances | Conclusions/Remarks |
|---------------------|------------------------------|-------------------------|-----------------|------|------------------------------------------|---------------------|
| 1) Yang et al. [3]  | Radio graphics (2004)        | Review article          | Nil             | NA   | Classical CT and clinical features of various types of RPCs studied | *CT provides information of size, shape, location, and involvement of adjacent structures *Substantial overlap of CT findings of various RPCs *Clinical history and certain CT details assist in making a correct diagnosis *Familiarity with CT features leads to accurate diagnosis and treatment |
| 2) Shanbhogue et al. [11] | Radio graphics (2012)       | Review article          | Nil             | NA   | Uncommon primary pelvic masses (cystic and noncystic) in adults studied | *Solid, cystic, vascular, myxoid, calcified, and fatty lesions occur as primary masses in pelvic retroperitoneum *Accurate detection on basis of anatomy, demographic and imaging features allows optimum treatment *Tissue-specific multiplanar capability of high-resolution MR imaging helps better localization and characterization |
| 3) Fdili Alaoui et al. [12] | Case reports in medicine (2012) | Case report (27y/F)    | Open            | 25 cm | Giant retroperitoneal epidermoid cyst excised | *Pelvic epidermoid cysts are very rare *They are retroperitoneal, retro rectal, and presacral in location *Treatment is surgical excision using a cleavage outside capsule to avoid injury to surrounding structures |
| 4) Vilos et al. [13] | Facts, Views & Vision in ObGyn (2020) | Case series (4)        | Open (1) & Laparoscopy (3) | 8.2, 8.5, 6 × 4 and 13 × 8 × 7 (All in cm) | Retroperitoneal pelvic tumors in 4 women studied (2 solid tumors and 2 cystic) | If incidentally detected, better to not attempt excision/biopsy, obtain postop multidisciplinary consults, specific imaging in order to treat these as safely as possible |
| 5) Johan et al. [14] | Visceral surgery (2020)     | Case report (56y/F)     | Open            | 18×14.5×14.6 cm | Preop CT indicated left adnexal mass-possibly ovarian serous cystadenoma, turned out to be benign RPC | *Preop left ureteric stenting underscores the importance of careful anticipation of collateral damage in giant RPCs and multidisciplinary approach *Due to the above, ureteric injury was averted |
| 6) Egawa et al. [15] | International journal of urology (1996) | Case report (63y/M)    | Open            | 11×16 cm | Preoperative retrograde pyelogram-marked displacement of left ureter | *Complete excision should be done *Marsupialization, internal drainage, and simple aspiration to be avoided due to high rate of recurrence and infection |
| 7) Morotti et al. [16] | Oncology letters (2017)     | Case report             | USG guided therapeutic aspiration | 15×7.5 cm | CT angiography showed compression and lateral deviation of Inferior vena cava | *RPCs are a challenging diagnostic dilemma *CT and/or MRI is essential for diagnosis *Unclear whether a biopsy or diagnostic aspiration to be done preop. *This case due to tricky location between IVC and aorta was just aspirated and monitored after ruling out malignancy, infection, and confirming normal fluid pancreatic enzyme levels |
| 8) Mitura et al. [17] | Int journal of surgery case reports (2013) | Case report (41y/F)    | Open            | 17×11×9.3 cm | Rt. Ureter and mesenteric vessels found on anterior surface of cyst | *Suspected to be mesenteric but was epidermoid cyst *RPCs originate from embryologic error *Epidermoid cyst is unilocular and found typically in the presacral space *Recurrence higher in RPCs due to proximity with major blood vessels and other organs, which makes complete excision difficult |
| 9) Yohendran et al. [18] | Asian journal of surgery (2003) | Case report (42y/F)    | Open            | 13 cm | Unilocular thin-walled Mullerian cyst | *RPC is commonest in 4th decade of life *Female preponderance |

Contd...
is the gold standard for the management of patients with RPC. The laparoscopic approach is practical, safe, and cost-effective.

**Key points**

- RPCs are rare and closely mimic commoner abdominal conditions
- Multispecialty team approach delivers optimum care.
- Regardless of size and location, they can be successfully managed by laparoscopy in a well-equipped setup, by an experienced operator
- Primary care physicians need to be aware of these rare differential diagnoses and their classical imaging appearances, in order to best guide their patients.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her names and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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