Case Report

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

Rafique U. Harvitkar[1], Rajendra Sankpal[2], Abhijit Joshi[3]

[1]Department of Surgery, Tameside and Glossop Integrated Care NHS Foundation Trust, Ashton Under Lyne, Manchester, UK, [2]Consultant Gynecologic Endoscopic Surgeon, Dr. L H Hiranandani Hospital, [3]Department of General Surgery, Consultant General, G I and Endo-Laparoscopic Surgeon, Dr. L H Hiranandani Hospital, Powai, Mumbai, Maharashtra, India

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

Introduction

A retroperitoneal cyst (RPC) is a relatively rare clinical entity with an estimated incidence that ranges from 1/5500 to 1/250,000. Approximately one-third of patients with RPCs are asymptomatic and found incidentally.[1] At times, they may attain a huge size and may present with a large abdominal lump and/or with abdominal symptoms due to the local mass effect.[2] These cystic lesions can be differentiated into a) neoplastic lesions (e.g., cystic lymphangioma, mesothelioma, teratoma, Müllerian cyst, epidermoid cyst, bronchogenic cyst, mucinous cystadenoma, cystic change in solid neoplasms, and pseudomyxoma peritonei) and b) nonneoplastic lesions (e.g., pancreatic pseudocyst, lymphocele, urinoma, hematoma and duplication cysts).[2,3]

Address for correspondence: Dr. Rafique U. Harvitkar, Department of Surgery, Tameside and Glossop Integrated Care NHS Foundation Trust, Ashton Under Lyne, Manchester, UK.
E-mail: dr_rafique639@yahoo.com

Received: 11-05-2021 **Revised:** 30-06-2021
Accepted: 05-07-2021 **Published:** 29-11-2021

Access this article online

Quick Response Code:

Website: www.jfmpc.com

DOI: 10.4103/jfmpc.jfmpc_863_21

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 cyst during diagnostic laparoscopy performed by him. She presented to him with chronic, dull, and vague lower abdominal pain for 3 months. There was no history of nausea, vomiting, constipation, or melena. The patient did not have any history of trauma or oral contraceptive use. She had polymenorrhagia and dysmenorrhea for 7 months. On per abdomen examination, she was found to have a soft lax abdomen, central obesity, and no palpable lump. She had a Pfannenstiel scar of a previous lower segment cesarean section in the lower abdomen. A per vaginal examination

How to cite this article: Harvitkar RU, Sankpal R, Joshi A. Huge pelvic retroperitoneal cyst masquerading as hydrosalpinx: A case report with review of the literature. J Family Med Prim Care 2021;10:4293-8.

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 by him 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 of 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.

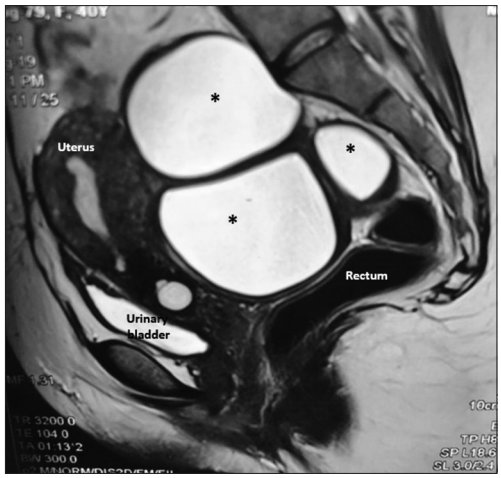

Figure 1: MRI of abdomen and pelvis: sagittal view showing trilobed pelvic retroperitoneal cystic mass

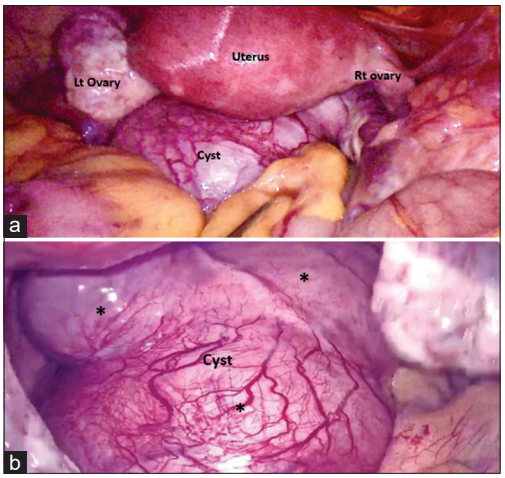

Figure 2: Laparoscopic view of the cyst with internal genitalia. a) Cyst seen separate from the internal genitalia. b) Trilobed cyst posterior to and separate from the internal genitalia (black asterisks)

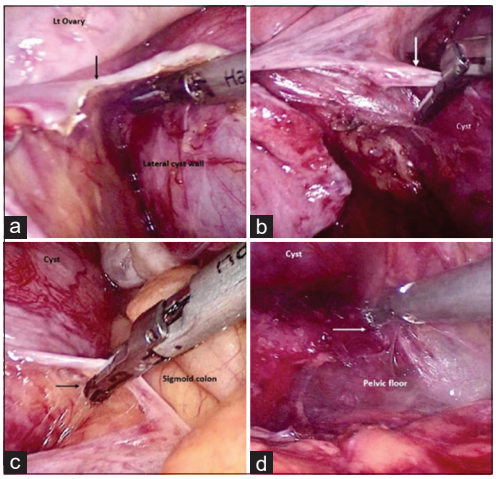

Figure 3: a) Initial dissection: peritoneum overlying the cyst (black arrow) incised thereby exposing the cyst wall. b) The cyst being separated from the left lateral pelvic wall (white arrow). c) The cyst being separated carefully from the sigmoid mesocolon (black arrow) on the right side. d) Pelvic floor attachments of the cyst being lysed (white arrow)

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

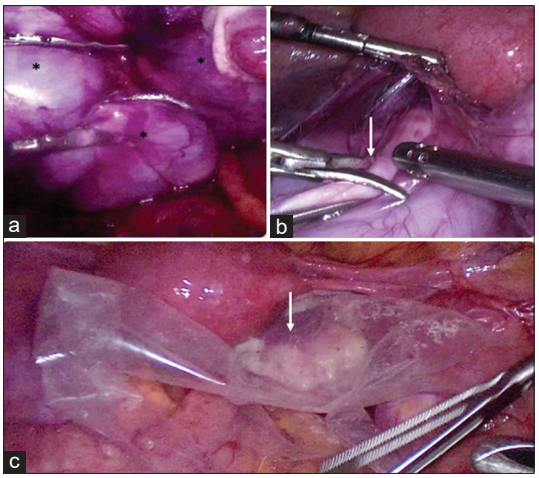

Figure 4: a) Trilobed cyst in 1 whole piece (black asterisks), freed from all its attachments and ready for extraction. b) Cyst being aspirated carefully to avoid spillage (white arrow), prior to retrieval. c) Cyst wall introduced into a retrieval bag and ready for extraction (white arrow)

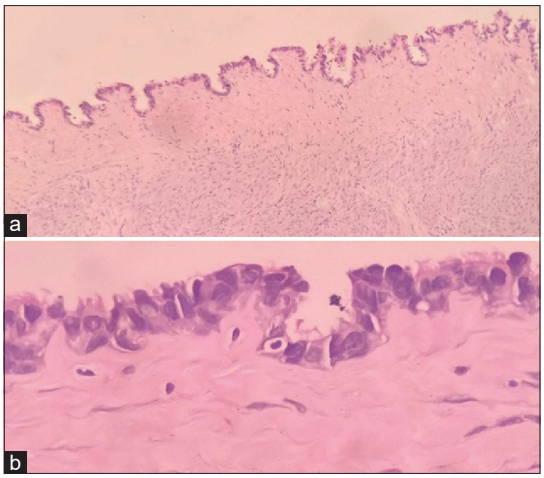

Figure 5: Cyst wall lined by simple columnar to cuboidal ciliated epithelium: a) low-power field and b) high-power field

rare. RPC masses pose an important diagnostic and therapeutic dilemma to the treating physicians.[4] 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.[3] 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.[7] 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

Table 1: Review of the Literature

Authors [Ref. no.]	Journal (Yr. Of publication)	Study type (No. of pts.) Age/Sex	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...

Table 1: Contd...

Authors [Ref. no.]	Journal (Yr. Of publication)	Study type (No. of pts.) Age/Sex	Mode of surgery	Size	Materials and methods/Imaging appearances	Conclusions/Remarks
10) Matthew *et al.* [19]	Female pelvic medicine & Reconstructive surgery (2007)	Case report (33y/F)	Open	17 cm	Upper abdominal Mullerian cyst presenting with nausea and vomiting	*Estimated incidence of RPCs is 1/5750 to 1/250000 *RPCs grow to a large size before becoming symptomatic as the retroperitoneum is a large potential space
11) Naem *et al.* [20]	International journal of surgery case reports (2019)	Case report (23y/M)	Open (Aspirated several times, operated twice)	Large multilocular-20 liters. serous fluid	Recurred twice-operated twice	A rare case of Mullerian cyst in a young male
12) Gopinath *et al.* [21]	Surgical case reports (2020)	Case report (21y/F)	3.5×6 × 5 cm	Laparoscopy	Right hepatic cyst	Highlights the unusual anatomic location and rare occurrence of upper abdominal urothelial RPC

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Guile M, Fagan M, Simopolous A, Ellerkman M. Retroperitoneal cyst of Mullerian origin: A case report and review of literature. J Pelvic Med Surg 2007;13:149-52.
2. Yang DM, Yoon MH, Kim HS, Oh YH, Ha SY, Oh JH, *et al*. Presacral epidermoid cyst: Imaging findings with histopathologic correlation. Abdom Imaging 2001;26:79-82.
3. Yang DM, Jung DH, Kim H, Kang JH, Kim SH, Kim JH, *et al*. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. Radiographics 2004;24:1353-65.
4. Martin R, Sanz E, de Vicente E, Ortega P, Labrador E, Paumard A, *et al*. Differential diagnosis of asymptomatic retroperitoneal cystic lesion: A new case of retroperitoneal bronchogenic cyst. Eur Radiol 2002;12:949-50.
5. Riojas CM, Hahn CD, Johnson EK. Presacral epidermoid cyst in a male: A case report and literature review. J Surg Educ 2010;67:227-32.
6. Alzaraa A, Mousa H, Dickens P, Allen J, Benhamida A. Idiopathic benign retroperitoneal cyst: A case report. J Med Case Rep 2008;2:43.
7. Ros PR, Olmsted WW, Moser RP, Dachman AH, Hjermstad BH, Sobin LH. Mesenteric and omental cysts: Histologic classification with imaging correlation. Radiology 1987;164:327-32.
8. Sharma G, Shreshtha S, Bhatt S, Garg PK. Retroperitoneal cystic mass: A diagnostic challenge. ANZ J Surg 2019;89:E576-7.
9. Sahoo MR, Misra L, Kaladagi RM, Gowda MS, Panda A, Behera SS. A huge retroperitoneal lymphatic cyst presenting as a mesenteric cyst managed laparoscopically. Int J Case Rep Images 2014;5:642-5.
10. Ravo B, Metwally N, Pai B, Ger R. Developmental retroperitoneal cysts of the pelvis. A review. Dis Colon Rectum 1987;30:559-64.
11. Shanbhogue AK, Fasih N, Macdonald DB, Sheikh AM, Menias CO, Prasad SR. Uncommon primary pelvic retroperitoneal masses in adults: A pattern-based imaging approach. Radiographic 2012;32:795-817.
12. Fdili Alaoui FZ, Oussaden A, Bouguern H, El Fatemi H, Melhouf MA, Amarti A, *et al*. Giant pelvic retroperitoneal epidermoid cyst: A rare case report. Case Rep Med 2012;2012:981387.
13. Vilos GA, Vilos AG, Hollett-Caines J, Abu-Rafea B, Jacob GP, Ettler H. Retroperitoneal pelvic tumours in women: Diagnostic and therapeutic challenges. Facts Views Vis Obgyn 2019;11:299-306.
14. Johan S, Haasan MF, Hayati F, Azizan N, Payus AO, Edwin See UH. Huge retroperitoneal cyst masquerading as ovarian tumour: A case report. Front Surg 2020;7:585411.
15. Egawa S, Satoh T, Suyama K, Uchida T, Iwabuchi K, Koshiba K. Giant retroperitoneal cyst in an adult male. Int J Urol 1996;3:304-6.
16. Morotti A, Busso M, Consiglio Barozzino M, Cinardo P, Angelino V, Familiari U, *et al*. Detection and management of retroperitoneal cystic lesions: A case report and review

of the literature. Oncol Lett 2017;14:1602-8.

17. Mitura K, Mikolaj R, Alicja M. An atypical site of a retroperitoneal epidermoid cyst in a middle-aged woman. Int J Surg Case Rep 2013;4:85-7.
18. Yohendran J, Dias MM, Eckstein R, Wilson T. Benign retroperitoneal cyst of Mullerian type. Asian J Surg 2004;27:333-5.
19. Matthew G, Matthew F, Simopolous A, Ellerkman M. Retroperitoneal cyst of Mullerian origin: A case report and review of the literature. Female Pelvic Med Reconstr Surg 2014;20:S151-368. doi: 10.1097/01.spv. 0000451381.90428. b4.
20. Naem A, Dlewati A, Alhimyar M, Ousta MA, Alsaid B. A rare presentation and recurrence of a retroperitoneal Müllerian cyst in a male patient: A case report. Int J Surg Case Rep 2019;65:301-4.
21. Gopinath A, Alkhasawneh A, Baskovich B, Altunkaynak C, Marji N, Awad Z. Upper abdominal primary retroperitoneal cyst with unusual urothelial histogenesis. Surgical case reports 2020. doi: 10.31487/j.SCR.2020.01.01.