

Retroperitoneal ovarian cyst presenting as acute abdomen – successfully managed with pre-operative prophylactic ureteric stenting and complete excision

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Mullerian Cyst , Retroperitoneum tumour , Ureteral Stent , Acute abdomen

Introduction

Cyst that arises from the retroperitoneum space are uncommon. Despite their possible common similarities in location and symptoms, they have known to be morphologically diverse. The incidence of such lesions is approximately 1 in 100,000 adult admissions. Retroperitoneum Mullerian cyst is rare whereby most patients are asymptomatic and usually present as an incidental finding of an abdominal mass [1]. Here, we report a case of retroperitoneum Mullerian cyst presenting as an acute abdomen.

Case Presentation

A 48-year-old lady presented with an acute onset of lower abdominal pain for 3 days associated with abdominal distention and vomiting. Physical examination revealed tenderness and fullness over the right iliac fossa and suprapubic region. White cell counts, haemoglobin, platelet count and renal profile were within normal limit and urine

pregnancy test was negative. Contrast enhanced computed tomography (CT) Scan of the abdomen revealed a large and well-defined retroperitoneal cystic tumour mass measuring about 8cm x 9cm x 15cm (AP x W x CC) with its medial side in close proximity to the right ureter and gonadal vessel (Figure 1A). There was streakiness over the Gerota's fascia above the lesion. There were no obvious paraaortic nodes or free fluid. She had worsening and persistent pain which raised the suspicion of impending tumour rupture. An early laparotomy with the intention for complete excision of cystic tumour was scheduled. The right ureter was prophylactically stented following general anaesthesia. Intraoperatively, the retroperitoneal cyst was mobilized from retroperitoneal attachments with ease. It was lying over the right lumbar muscles, inferior to the right kidney and lateral to the right ureter. It had no shared blood supply to any pelvic organs (Figure 1B). She recovered uneventfully and was discharged home on the second day of surgery and she remained well throughout her follow ups.

Histopathological examination revealed that the cyst wall was lined with cuboidal epithelium and showed fibrocollagenous tissue interspersed with smooth muscle cells; features are consistent with a Müllerian duct cyst

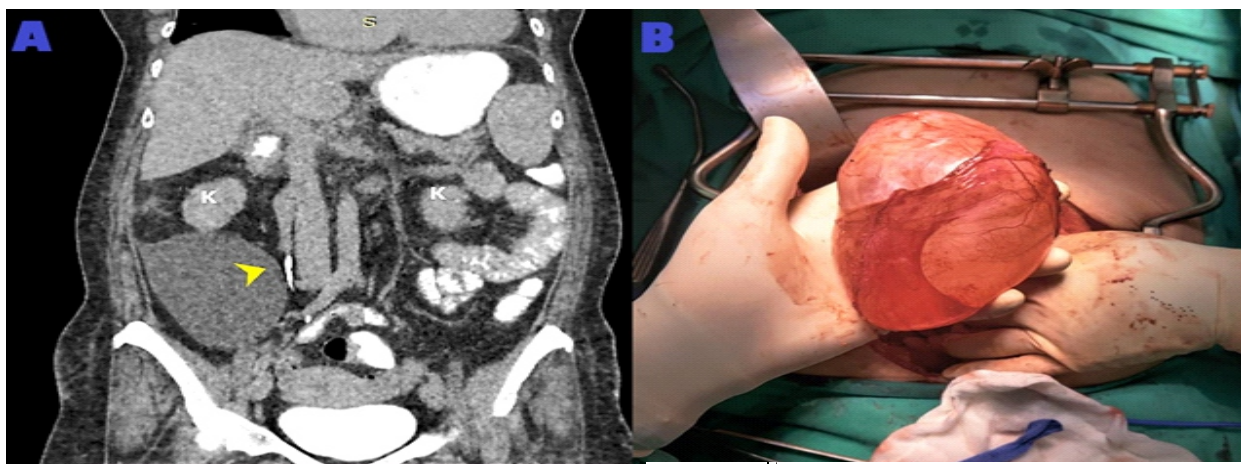


Figure 1A: Contrast CT in delay phase revealed a large and well-defined retroperitoneal cystic mass with its medial side in close proximity to the right ureter (arrowed head). K: kidney

Figure 1B: Intraoperative specimen of the retroperitoneal cyst

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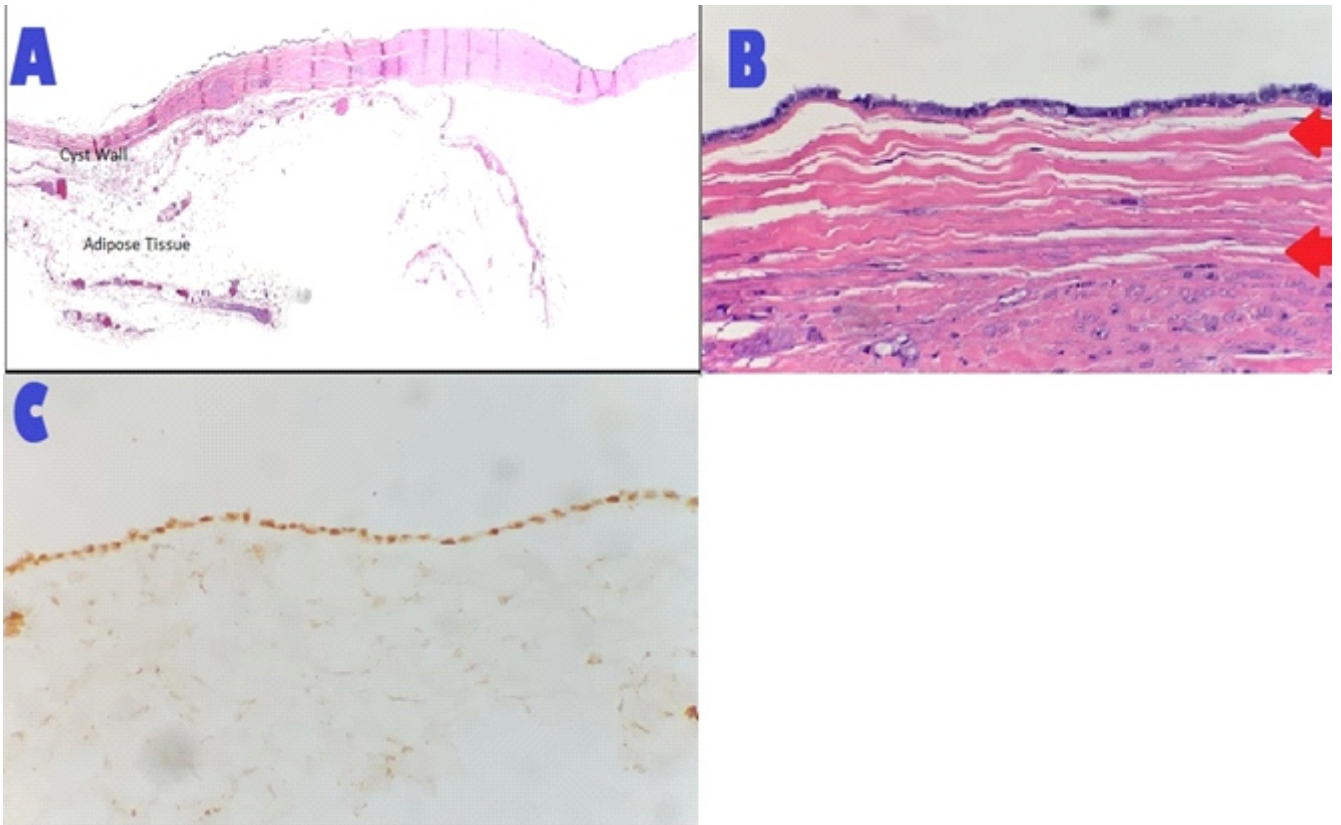


Figure 2A: Sections show a thin cyst wall lined by a single layer of flattened to ciliated cuboidal-columnar epithelium. No abnormal epithelial proliferation or cellular atypia seen. The wall is fibrocollagenous with occasional smooth muscle bundles noted. Minimal adipose tissue is also seen covering the capsule.

Figure 2B: Upper arrowhead – A Thin cyst wall lined by a single layer of ciliated cuboidal-columnar epithelium
Lower arrowhead – Fibrocollagenous wall with occasional smooth muscle bundles

Figure 2C: Immunohistochemistry: PAX 8 is positive

(Figure 2A, Figure 2B). No atypical cells seen. Immunohistochemical analysis showed positive for PAX8 (Figure 2C)

Discussion

Embryonically the retroperitoneal Mullerian Cyst originates from the remnants of the Mullerian system. It is uncommon for an acute presentation if is uncomplicated [2]. Imaging clue from CT scan are usually the most objective way to differentiate the lesion. There are wide variety of differential diagnosis of a retroperitoneal cystic mass, it can be either neoplastic or non-neoplastic. Neoplastic lesions include cystic lymphangioma, mucinous cystadenoma, cystic teratoma, Mullerian cyst. Nonneoplastic lesions include pancreatic pseudocyst, nonpancreatic pseudocyst and lymphocele. In this current case, the initial diagnosis was either a lymphangioma or cystic neoplasm. The indication of surgical excision in this case was due to abdominal pain and the concerns of tumor rupture. As there was an uncertain malignant potential with its large size and adjacent

compression, a complete excision was planned rather than drainage.

The incidence of malignant retroperitoneal cyst reported is low at less than 3% [3]. There were reports that described the option of percutaneous drainage. However, it carries higher risk of recurrence and delayed recovery [4]. Judging by the risk of recurrence, even if it may be a benign cyst, the option of total surgical resection was performed. The excision can be performed with laparoscopy access but we embarked in laparotomy manner due to logistics circumstances (available surgeon experience and staffing at the time point of emergency time schedule).

Conclusion

Large retroperitoneal cyst is uncommon and usually benign especially when it is clear cystic content. It may present as acute abdomen though uncomplicated as in current case. However, if the patient is symptomatic, early complete surgical excision is the best option especially in a fit patient.

References

1. Yohendran J, Dias MM, Eckstein R, Wilson T. Benign retroperitoneal cyst of Mullerian type. *Asian Journal of Surgery*. 2004 Oct 1;27(4):333-5. DOI: 10.1016/S1015-9584(09)60062-2
2. Singh A, Shree S, Mishra V, Radhakrishnan G. Mullerian duct cyst misdiagnosed as ovarian cyst: a rare case report. *International Journal of Reproduction, Contraception, Obstetrics and Gynecology*. 2016 Apr 1;5(4):1260-3. DOI: <http://dx.doi.org/10.18203/2320-1770.ijrcog20160900>
3. Kurtz RJ, Heimann TM, Holt JA, Beck AR. Mesenteric and retroperitoneal cysts. *Annals of surgery*. 1986 J a n ; 2 0 3 (1) : 1 0 9 . DOI:10.1097/00000658-198601000-00017
4. 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. *International Journal of Surgery Case Reports*. 2019 Jan 1;65:301-4. DOI : 10.1016/j.ijscr.2019.11.001

Learning Points:

- Large retroperitoneal cyst can be benign and has a clinical course that mimics an acute abdomen
- Preoperative diagnosis of retroperitoneal cyst can be a difficult task as they are morphologically diverse.
- Best interest of the patient should always be included when formulating treatment options.