CASE REPORT

An unusual presentation of a retroperitoneal cyst

Debashis Sarkar,^{1,2} Dev Gulur,² Snehal Patel,² Thiagarajan Nambirajan³¹MSc Advanced Surgical Practice, Cardiff University, Cardiff, UK²Department of Urology, Wirral University Hospital, Wirral, UK³Wirral University Teaching Hospital, Wirral, UK**Correspondence to**Debashis Sarkar,
debu4726@yahoo.co.uk

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SUMMARY

A 34-year-old woman presented to the surgical assessment unit with severe right loin to groin pain. An ultrasound scan of the abdomen revealed a complex cyst in the right iliac fossa and a subsequent CT scan revealed a 7.5 cm retroperitoneal cystic lesion below the lower pole of the right kidney. The patient also had MRI of the kidneys, which confirmed the finding. The image showed the cyst was not attached to the kidneys and was clearly separate. She underwent a laparoscopic excision of the cyst. Histopathology revealed a cyst lined by a single layer of mucinous epithelium of endocervical type with foci of calcification and hyalinisation on the wall. The cyst was thought to be a benign cyst of Mullerian origin.

BACKGROUND

Mullerian ducts are paired urogenital ducts formed in females during the developmental period in utero. During the period of embryogenesis and developmental period, these ducts transform into the fallopian tube, cervix and upper portion of the vagina. Mullerian duct cysts are derived from the remnants of the fused caudal end of the Mullerian ducts, which normally regress in utero. However, failure to regress can develop into Mullerian duct cysts, which present in adult age around the female pelvis close to the genital organs. In this case, the patient presented with a huge retroperitoneal cyst, which makes the diagnosis difficult.

CASE PRESENTATION

A 34-year-old woman presented with a history of severe right loin to groin pain associated with nausea not dissimilar to ureteric colic. There were no urinary tract symptoms, no haematuria and no signs of infection. The pain was intermittent with no exacerbating factors. The patient had no history of trauma. However, she did have a history of cervical intraepithelial neoplasia grade 2, under gynaecological review. She was otherwise fit and healthy and not taking any medication. There was no significant family history. She has a single child from a normal delivery and lives with her husband.

INVESTIGATIONS

- USS of the abdomen-right iliac fossa complex mass ([figure 1](#)).
- CT of the abdomen and pelvis ([figures 2 and 3](#)).
- MRI of the abdomen ([figure 4](#)).
- Histopathology ([figures 5 and 6](#)).

Figure 1 Right iliac fossa complex mass.

TREATMENT

In view of the symptoms, the patient opted for laparoscopic excision. A transperitoneal approach revealed a cyst with clear fluid in the retroperitoneum separate from ureter, gonadal vessels and kidney. The cyst was completely excised. The fluid was aspirated and cyst extracted through the port site.

OUTCOME AND FOLLOW-UP

Histology revealed a cyst lined by a single layer of mucinous epithelium of endocervical type. Foci of calcification and hyalinisation of the cyst wall were noted. The cyst was thought to be a benign cyst of Mullerian origin. The patient attended follow-up and is doing well and is pain free.

Figure 2 Mass originates from lower pole of the right kidney.

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Figure 3 Mass originates from lower pole of the right kidney.

Figure 4 Mass is separated from the kidney.

Figure 5 Low-power and high-power views showing cyst wall lined by a single layer of benign Mullerian type (mucinous) epithelium on histopathology.

Figure 6 Low-power and high-power views showing cyst wall lined by a single layer of benign Mullerian type (mucinous) epithelium on histopathology.

DISCUSSION

Mullerian cysts usually present in women during the third to fourth decade. The incidence of retroperitoneal cysts is reported in the literature from 1 in 5750 to 1 in 250 000 (average, 1 in 105 000). Among these, Mullerian duct cysts are very uncommon. The usual clinical presentation for Mullerian duct cysts is in the female pelvis area, but around the pancreas and near the gastric area has also been reported in the literature. The retroperitoneal area near the kidney is an uncommon presentation. In males, they present as prostatic utricle cysts, which are also uncommon.

Learning points

- ▶ Retroperitoneal cysts can be Mullerian cysts, which can mimic pyelonephritis or ureteric colic and can present as other differentials such as:
 - Epithelial neoplasms: Mucinous cystadenoma or cystadenocarcinoma, serous cystadenocarcinoma.
 - Germ cell neoplasms: Cystic teratoma.
 - Neuroendocrine neoplasms: Paranglioma.
 - Neural neoplasm: Schwannoma.
 - Sarcoma: Necrotic areas can appear cystic.
 - Lymphovascular: Lymphangioma, lymphangiomatosis, lymphangioleiomyoma.
 - Developmental: Epidermoid cysts, tailgut cysts, bronchogenic cysts (subdiaphragmatic location).
- ▶ Miscellaneous fluid collections: Haematoma, urinoma, abscess, pseudocyst (pancreatic and non-pancreatic), and pseudomyxoma retroperitonei (caused by a ruptured mucocele of the appendix).
- ▶ Laparoscopic excision would be the best treatment option as laparoscopic cyst decortication is an effective and durable treatment for symptomatic renal cysts, as assessed over a long-term follow-up. Its minimal invasiveness and greater success rate and less recurrence favour it over other techniques (open surgical or percutaneous drainage).
- ▶ It does not routinely need frozen section if it arises from the kidneys, as the published literature so far does not suggest any malignant changes.

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