

Velika retroperitonealna masa pri 56-letni ženski

Large retroperitoneal mass in a 56-year old female

Avtor / Author
Ustanova / Institute

Nuhi Arslani^{1,2}

¹Univerzitetni klinični center Maribor, Oddelek za abdominalno in splošno kirurgijo, Maribor, Slovenija; ²Univerza v Mariboru, Medicinska fakulteta, Maribor, Slovenija

¹University Medical Centre Maribor, Department of Abdominal and General Surgery, Maribor, Slovenia; ²University of Maribor, Faculty of Medicine, Maribor, Slovenia

Ključne besede:
Retroperitonealni prostor, kongenitalne napake, benigne ciste, ciste Müllerjevega voda

Key words:
Retroperitoneum, congenital abnormalities, benign cyst, Müllerian ductal cysts

Članek prispel / Received
8.11.2016

Članek sprejet / Accepted
20.4.2017

Naslov za dopisovanje / Correspondence

Dr. Nuhi Arslani, dr. med.
Univerzitetni klinični center Maribor,
Oddelek za abdominalno in splošno kirurgijo, Maribor, Slovenija.
E-pošta: arslani.nuhi@gmail.com

Izvleček

Namen: Ciste Müllerjevega voda so redke kongenitalne napake regresije Müllerjevega sistema, ki se običajno nahajajo v medenici. Njihova prisotnost v retroperitoneju je zelo redka. Incidenca retroperitonealnih cist različne morfologije in etiologije je približno 1 na 100 000 odraslih bolnikov. Razvrstimo jih lahko v limfne ciste, mezotelne, enteralne in urogenitalne ciste. Urogenitalne ciste so nadalje razvrščene v pronefrične, mezonefrične, metanefrične in Müllerjeve tipe. Pri obeh spolih je najpogostejše mesto za Müllerjeve ciste medenica, pri moških večinoma v bližini prostatičnega utrička in modih. Pri ženskah jih lahko pogosto zamenjujemo s primarnimi cistami jajčnikov in jajcevodov.

Poročilo o primeru: V tem poročilu predstavljamo primer benigne retroperitonealne ciste Müllerjevega tipa pri 56-letni bolnici.

Abstract

Purpose: Müllerian ductal cysts are rare congenital abnormalities of Müllerian system regression, typically located in the pelvis. Their presence in retroperitoneum is extremely rare, as the incidence of retroperitoneal cysts, even of diverse morphology and etiology, is approximately 1 in 100,000 adult admissions. Moreover, they can be classified into lymphatic, mesothelial, enteric, and urogenital cysts. Urogenital cysts are further categorized into pronephric, mesonephric, metanephric, and Müllerian types. However, in both sexes the most common location of the Müllerian cyst is in the pelvis; in men they reside mostly near the prostatic utricle and the appendix of the testis. In women, they may be often confused with primary ovarian or tubal cysts.

Case Report: In this case report, we present a case of a benign retroperitoneal cyst of the Müllerian type in a 56-year old female.

CASE REPORT

A 56-year old female was admitted to the urological department for percutaneous biopsy of a kidney cyst. Further investigations showed that there was no connection between the kidney and the cyst; thus, a mesenteric cyst was suspected. She complained of nonspecific pain under the left costal margin, a digestive disorder, nausea, and flushing. In the past, she was anemic due to gynecological bleeding, for which she underwent endometrial ablation. She also had a history of uterine myomas. She was on medication for elevated blood pressure. Her physical examination was unremarkable.

Laboratory examinations, including total blood count, biochemical profile, and tumor markers were within normal range. A nativ computer tomografi (CT) scan and ultrasound of the abdomen were performed, which confirmed a round cystic lesion, likely mesenteric filled with 480 ml of sludge, located under the spleen with no connection to other organs. Moreover, a biliary hepatic cyst in the fifth lobe and a 6 cm large uterus myoma were described. Her urea breath test was positive for *Helicobacter pylori*, for which she underwent eradication therapy without consequence.

After one year, follow-up abdominal computer tomografi scan showed enlargement of the abdominal

mass to 12 × 16 × 15 cm in size. The homogeneous hypodense mass was located between the lower margin of the spleen and lesser pelvis, and had radiologic features of a mesenteric cyst with close relationship to and compression of other organs (Figures 1, 2). Extirpation of the cyst was offered.

A laparotomy was performed. After the mobilization of the line of Toldt, the cyst became apparent in the retroperitoneum (Figure 3). There were no attachments to other organs in the abdomen or pelvis. The cyst was evacuated in its entirety from the retroperitoneum, mostly with hand manipulation. Drainage was left in situ. Macroscopically, the cyst was homogenous, measuring 23 × 15 × 13 cm and weighing 2040 g. The epithelium was approximately 0.1 cm thick and the cyst contained yellow clear fluid (Figures 5, 6). Microscopic examination revealed Müllerian duct origin. The cystic wall was composed of tubular epithelium with connective tissue stroma. There were no complications in the early postoperative period or during follow-up.

DISCUSSION

Retroperitoneal cyst of Müllerian type is a rare entity (5, 6). The precise mechanism of its development is not known. One theory postulates that the retroperitoneum includes aberrant, embryologic-derived

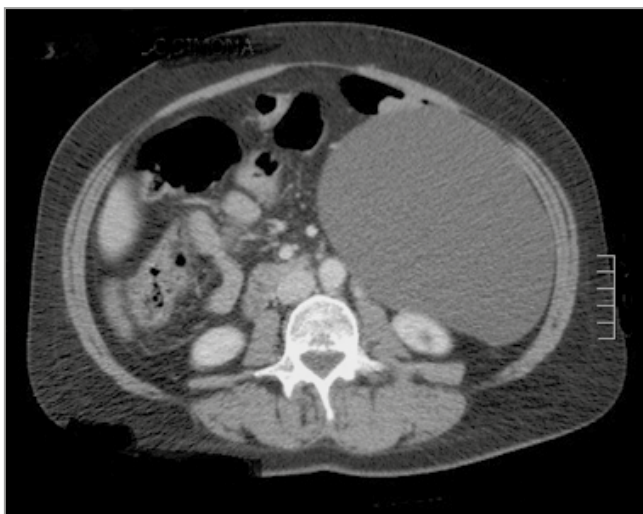


Figure 1. Transverse abdominal CT scan showing the retroperitoneal cyst on the left.

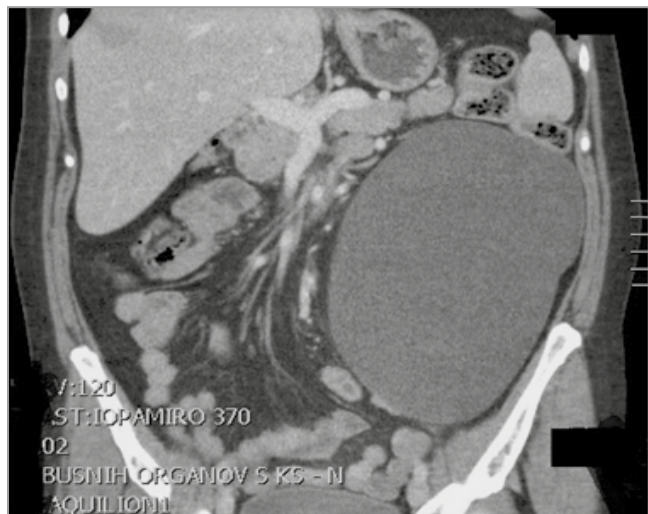


Figure 2. Coronal CT scan showing the retroperitoneal cyst on the left.

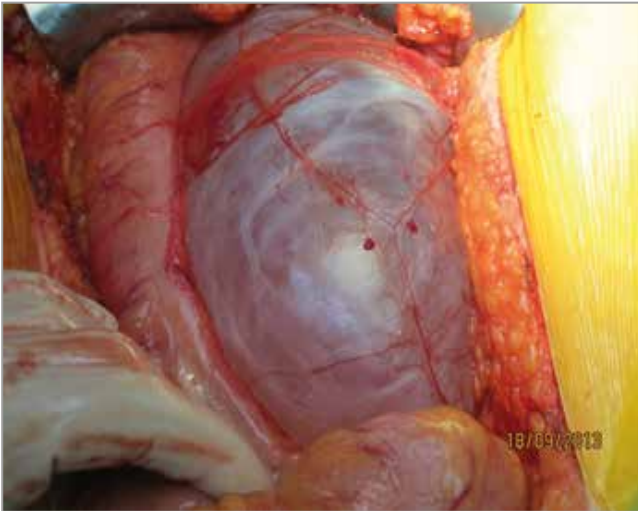


Figure 3. Exposure of the cyst after mobilization of the line of Toldt.

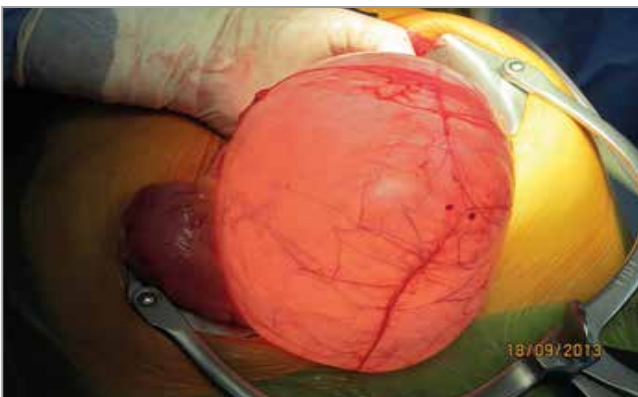


Figure 4. The macroscopically homogenous müllerian cyst containing yellow clear fluid

Müllerian duct tissue. This tissue might grow later in life under the influence of abnormal hormonal stimuli. The other theory suggests that the peritoneum might have undergone differentiation to become a serous/tubal-type with invagination of the underlying tissue, producing a cystic structure (7).

Retroperitoneal mMüllerian cysts are mostly found in women after the third decade. In most cases, retroperitoneal cysts are clinically silent; but they can cause abdominal discomfort, pain, nausea, and vomiting (8, 9) In our case, epigastric discomfort and nausea were present, probably because of the *H. pylori* infection. Furthermore, Müllerian duct cysts

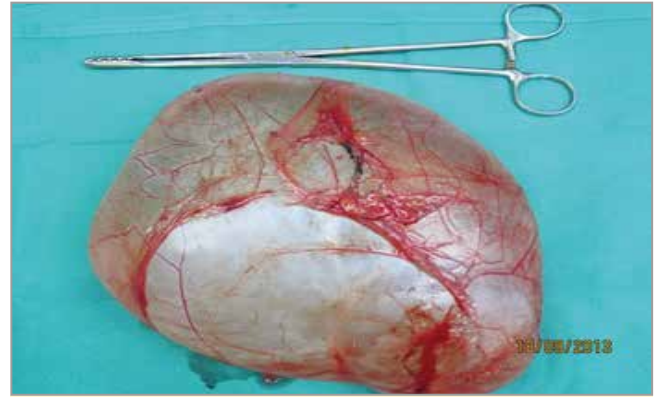


Figure 5. Removed cyst after the procedure, measuring 23 × 15 × 13 cm and weighing 2040 g.

can cause obstruction and irritation of the urinary system with the development of hydronephrosis and hydroureter (10,11). Retroperitoneal cysts can also be present as a palpable abdominal mass, in approximately 50% of cases (12).

The diagnosis of retroperitoneal cyst is difficult to establish preoperatively due to the lack of pathognomonic clinical features. Laboratory data are of no value, except for cancer antigen 125 that can indicate the benign nature of the cyst. Radiologic investigations such as ultrasound, CT scan, and magnetic resonance imaging of the abdomen can determine the location, relationship to other organs, and the nature of the cystic lesion, whether serous or blood-filled. The definitive diagnosis can be confirmed after pathologic examination of the cyst. In our case, the cystic formation first resembled a renal cyst. Further radiologic diagnostic investigations proved there to be no connection between its formation, and the kidney and the pancreas.

Invasive treatment of Müllerian cysts is only indicated when symptoms appear. The best treatment choice is surgical extirpation with preservation of other normal surrounding structures. This procedure can be performed either laparoscopically or open. Open excision is recommended for large pelvic or abdominal masses. Segmental bowel resection may be necessary due to involvement of the mesenteric vessels (12).

REFERENCES

1. Tayyebi Meibodi N, Mahdavi R, Montazer M. Large pelvic cystic mass of müllerian duct origin with involvement of rectal wall: a case report. *Iran Red Crescent Med J* 2011; 13(2): 149–50.
2. De Perrot M, Bründler M, Tötsch M, Mentha G, Morel P. Mesenteric cysts. Toward less confusion? *Dig Surg* 2000; 17(4): 323–8.
3. De Peralta MN, Delahoussaye PM, Tornos CS, Silva EG. Benign retroperitoneal cysts of müllerian type: a clinicopathologic study of three cases and review of the literature. *Int J Gynecol Pathol Off J Int Soc Gynecol Pathol* 1994; 13(3): 273–8.
4. Shayan H, Owen D, Warnock G. Surgical images: soft tissue. Müllerian cyst of the upper abdomen: a lesion mimicking pancreatic cystadenoma. *Can J Surg J Can Chir* 2004; 47(5): 369–71.
5. Konishi E, Nakashima Y, Iwasaki T. Immunohistochemical analysis of retroperitoneal Müllerian cyst. *Hum Pathol* 2003; 34(2): 194–8.
6. Ray M, Bose B, Honore L. A 53-year-old woman with abdominal pain and fullness. *CMAJ Can Med Assoc J J Assoc Medicale Can* 2005; 172(2): 184.
7. Lauchlan SC. The secondary müllerian system revisited. *Int J Gynecol Pathol Off J Int Soc Gynecol Pathol* 1994; 13(1): 73–9.
8. Minor TX, Yeh BM, Horvai AE, Abrahams HM, Meng MV, Stoller ML. Symptomatic perirenal serous cysts of müllerian origin mimicking renal cysts on CT. *AJR Am J Roentgenol* 2004; 183(5): 1393–6.
9. Lee J, Song SY, Park CS, Kim B. Müllerian cysts of the mesentery and retroperitoneum: a case report and literature review. *Pathol Int* 1998; 48(11): 902–6.
10. Burkett JS, Pickleman J. The rationale for surgical treatment of mesenteric and retroperitoneal cysts. *Am Surg* 1994; 60(6): 432–5.
11. Walker AR, Putnam TC. Omental, mesenteric, and retroperitoneal cysts: a clinical study of 33 new cases. *Ann Surg* 1973; 178(1): 13–9.
12. Liew SC, Glenn DC, Storey DW. Mesenteric cyst. *Aust N Z J Surg* 1994; 64(11): 741–4.