ABSTRACT
The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms. Mucinous cystadenoma of the appendix is a rare condition that develops as a result of proliferation of mucin-secreting cells in an occluded appendix and found in 0.2–0.3% of resected appendices in Europe and the United States. Therapy is fundamentally surgical and several options have been reported, ranging from simple appendectomy to right hemicolectomy.

In this case, mucinous cystadenoma of the appendix was removed by used laparoscopic retroperitoneoscopic technic.

Key Words: Retroperitoneal mass, Appendix mucinous cystadenoma, Retroperitoneoscopy

INTRODUCTION
The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms that can be either primary or metastatic lesions. Malignant tumours of the retroperitoneum occur four times more frequently than benign lesions. Sarcomas comprise a third of retroperitoneal tumours. Other retroperitoneal neoplasms include primary lymphoproliferative tumours (Hodgkin’s and non-Hodgkin lymphoma) and epithelial tumours (renal, adrenal, pancreas) or might represent metastatic disease from known or unknown primary sites (germ cell tumours, carcinomas, melanomas). The most common benign pathologies encountered in the retroperitoneum include benign neurogenic tumours (schwannomas, neurofibromas), paragangliomas (functional or non-functional), fibromatosis, renal angiomyolipomas, benign retroperitoneal lipomas and mucinous cystadenoma (1).

Mucocele of the appendix caused by a retention cyst, mucosal hyperplasia, mucinous cystadenoma or mucinous cystadenocarcinoma is found in 0.2–0.3% of resected appendices in Europe and the United States (2). Therapy is fundamentally surgical and several options have been reported, ranging from simple appendectomy to right hemicolectomy. Surgical excision of the tumor without rupture is of paramount importance because rupture of the lesion causes pseudo-myxoma peritonei (3). We present a case of an appendiceal mucocele caused by a mucinous cystadenoma clinically presented as a giant retroperitoneal mass. The retroperitoneal mass was removed laparoscopic retroperitoneoscopic technic. Diagnosis was postoperatively made, after pathological study of the surgical sample. This is, to our knowledge, the first case of retroperitoneoscopic by laparoscopic resection for retroperitoneal mucocele of the appendix described in the indexed literature.
CASE

An 77-year-old man was referred to our hospital with lower right back pain experienced for a few days. He did not have any significant history of bloating or diarrhea, constipation or any melena or hematochezia. He denied any recent episodes of fever, nausea or vomiting and reported a stable appetite and weight. On physical examination, he was afebrile and hemodynamically stable. The abdominal examination was normal. There was no focal tenderness over McBurney’s point and rebound tenderness on palpation. He was not found to have a right iliac fossa mass. Routine laboratory examination on admission showed no abnormalities. Laboratory data were unremarkable and tumor markers including CA-125, CA-19.9, and CEA were within normal limits. Ultrasound showed a 9 cm in diameter fluid-filled mass in the right retroperitoneal space. A contrast-enhanced computed tomography (CT) scan of the abdomen revealed a hypodense right retroperitoneal mass (8x5 cm). (Figure 1)

Laparoscopic resection of the tumor by retroperitoneal approach was carried out. The procedure was performed under general anesthesia. After bladder catheterisation, the patient was placed in the standard right lateral kidney position. We used the open (Hasson) technique for obtaining initial access. A 12 mm incision was made in the lumbar (Petit’s) triangle below the 12th rib at the lateral border of paraspinalis muscles. The balloon dilator was then inserted into the opening. Distension of the balloon with air rapidly and atraumatically displaces the adjacent fat and peritoneum, thereby creating an adequate working space for retroperitoneoscopic surgery within that area. A 10mm port was then placed in this opening and used as the camera port. The 2nd and 3rd ports were inserted under direct vision. The psoas muscle and ureter acts as a landmark and was sought immediately on entry with the laparoscope. The mass was found lower pole of the right kidney, superior of the ureter (Figure 2). The mass was dissected gently starting from its upper pole. The mass was attached to the retroperitoneal area. Following the dissection of the upper pole, the relationship between peritoneum and lower pole of the mass was determined. Then peritoneum was opened gently. It appeared to be a mass of the appendix from the cecum. The mass was excised from cecum using an endoscopic linear stapler (Figure 3). The specimen is kept in an endobag and delivered through a small muscle-splitting incision in the right iliac fossa (Figure 4, 5). Histopathologic diagnosis was mucinous cystadenoma of appendix.

There were no complications in the postoperative period. The patient was released from the clinic on the fourth day. After two years follow-up the patient is stable.
Therefore, when a mucinous tumor is diagnosed intra or retroperitoneal, careful search for the origin(s) has to be performed. Most likely they are metastases from the ovaries, fallopian tubes, uterus, cervix, appendix, bladder, urachus, breast, ureter, pancreas, bowel, or gall bladder (5). In our case, it was shown that the retroperitoneal mass arised from appendix.

Mucinous cystadenoma is a rare primary epithelial retroperitoneal tumor that is believed to arise secondary to invagination of peritoneal mesothelium with subsequent mucinous metaplasia and cyst formation, as a result of ectopic ovarian tissue, from a teratoma with single mucinous cell lineage, or from remnants of the embryonic urogenital apparatus (4).

**DISCUSSION**

The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms that can be either primary or metastatic lesions. The differential diagnosis includes a primary neoplasm arising from a retroperitoneal visceral structure (eg, pancreas, adrenal glands, kidneys, and duodenum), a retroperitoneal sarcoma, a lymphoma, or a metastatic lesion (4).

Mucinous cystadenoma of the appendix is a rare condition that develops as a result of proliferation of mucin-secreting cells in an occluded appendix (2). The tip of the appendix may lie in a variety position. The most common location is retrocecal 77%. It is pelvic in 30% and retroperitoneal in 7% of the population (6). These anatomic variation might be responsible for atypical presentations of appendix mucocele. Therefore retroperitoneal appendix mucocele may seem like a retroperitoneal mass.
Preoperative diagnosis of appendicular mucocele is very important for the selection of an adequate surgical method to prevent peritoneal dissemination, to prevent intraoperative and postoperative complication, and repeated surgery (2).

Moreover, radiological imaging, such as CT and MRI, plays an important role in describing and assessing the disease characteristics and involvement of adjacent/distant structures of the mass, but cannot exclude the malignant potential of the retroperitoneal masses (7). In our case, CT scan of the abdomen revealed a hypodense right retroperitoneal mass. Laboratory studies, including serum tumor markers and cytology study of cystic fluid are not helpful in making diagnosis of the tumors. However, Motoyama et al. (8) reported that measurement of CEA level in the cystic fluid may be useful in making the diagnosis. In our case, tumor markers [CA-125, CA-19.9, and CEA] were within normal limits.

Provided resection margins are clear, appendectomy is curative for unruptured appendiceal mucoceles of benign origin (i.e mucosal hyperplasia and mucinous cystadenoma). On the other hand, partial cecectomy may be indicated for a broad-based benign mucocele that protrudes into cecal lumen. If either cecal wall or ileum is invaded by tumor or adequate surgical margins cannot be secured, ileocecal resection or right hemicolecotomy may be required (3). In our case, only the appendiceal mass was removed using endoscopic linear stapler.

The optimal treatment modality for appendiceal mucocele is still controversial. Surgical management of this entity differs primarily depending on the characteristics of the mass (location and size) and clinical presentation, whereas the approach used (open or minimally invasive techniques) depends partly upon the preference and experience of the surgeon (9).

Some authors feel that a laparoscopic approach should be avoided in this setting because the change of rupture is increased, predisposing to this complication. Moreno et al. reported a case of nonperforated mucinous adenocarcinoma resected via laparoscopic appendectomy where diffuse peritoneal carcinomatosis later developed, citing appendiceal mucocele as a contraindication to laparoscopic resection (10).

Navarra et al. (11) reported successful laparoscopic resection of an appendiceal mucocele and suggested that conversion to a laparotomy should be considered if the lesion is grasped traumatically or if the tumor clearly extends beyond the appendix. Lau et al. (12) also reported the laparoscopic resection of a 12 cm large mucocele and Chiu et al. (9) reported a case involving the successful laparoscopic resection of an 8 cm appendiceal mucinous cystadenoma.

The advancement of laparoscopic surgery offers surgeons a useful way of removing retroperitoneal cystic lesions, with further advantages including less postoperative pain, lower morbidity, shorter hospitalization, and an earlier recovery (13).

In our case, the retroperitoneal mass was removed by laparoscopic retroperitoneoscopic technique. There was no complication.

CONCLUSION

Mucocele of the appendix should be considered in the differential diagnosis of cystic lesions in the retroperitoneum. Laparoscopic appendectomy for mucocele should be considered as the primary choice. Laparoscopic technique is a good alternative to open surgery for appendix mucocele and significantly reduced the morbidity of surgery. A retroperitoneal approach is feasible, despite the large amount of retroperitoneal space occupied by this appendix mucocele. This is, to our knowledge, the first case of retroperitoneoscopic laparoscopic resection for mucocele of the appendix described in the indexed literature.

REFERENCES

