

# Torsion of an appendiceal mucinous cystadenoma

## Report of a case and review of literature



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### Torsion of an appendiceal mucinous cystadenoma. Report of a case and review of literature

*Vermiform appendix torsion is rare, and even more uncommon is the volvulus of appendiceal mucinous tumors. To our knowledge, only nine cases of torsion of the vermiform appendix associated with appendiceal mucinous neoplasms have been reported up to date. We report a case of secondary torsion of the vermiform appendix with mucinous cystadenoma in a 30-year-old man. The symptoms were consistent with acute appendicitis. Diagnostic laparoscopy revealed a mucinous tumor of the appendix with a 360 degrees twisted appendix. In order to avoid peritoneal dissemination open appendectomy was performed. The final pathologic diagnosis was a mucinous cystadenoma of the appendix. The postoperative recovery was uneventful.*

*Secondary torsion of vermiform appendix with mucinous cystadenoma is a rare entity. Correct preoperative diagnosis is unlikely. Open appendectomy is the standard of care for benign mucinous appendiceal tumors. Furthermore, it is important to prevent spillage of the appendiceal mucocele content.*

KEY WORDS: Appendix, Mucocele, Torsion.

### Introduction

Appendiceal mucocele (AM) is a very rare clinical condition, characterized by distension of the appendiceal lumen due to accumulation of mucinous substance<sup>1</sup>. The spontaneous and surgery induced complications of AM include intestinal obstruction, intussusception, intestinal bleeding, fistula formation, and volvulus<sup>2</sup>. The most severe complication is pseudomyxoma peritonei (PMP), characterized by peritoneal dissemination caused by iatrogenic or spontaneous rupture of the mucocele<sup>3</sup>. Acute torsion complicating AM is exceedingly rare, with only nine cases reported in the English literature<sup>4-12</sup>.

Although preoperative diagnosis is important to avoid rupture of a mucocele, the diagnosis is often established during surgery.

We present an additional case of secondary volvulus due to AM (mucinous cystadenoma) as well as the relevant literature review.

### Case report

A 30-year-old Caucasian male presented 6 hours after onset to our department complaining right lower abdominal quadrant pain and nausea. There was no significant past medical history. On physical examination his body temperature was 36.6 °C, heart rate 95 beats/min, blood pressure 120/60 mm Hg. Rebound tenderness and muscular guarding in the right lower abdominal quadrant were positive. Peripheral blood test showed a WBC of 9,600/mm<sup>3</sup>. No other abnormalities were detected. The Alvarado's score for acute appendicitis was five points. Diagnostic laparoscopy was performed demonstrating a twisted mucinous tumor of the appendix with an appar-

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Fig. 1: Distended and 360° clockwise twisted appendix (arrow).

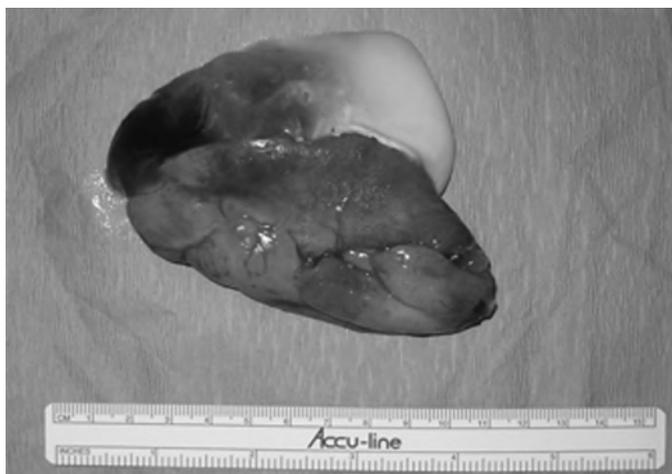


Fig. 2: Macroscopic findings: appendiceal lumen filled with mucin with dark color at the distal part.

ently gangrenous distal part. In order to avoid peritoneal dissemination conversion to an open appendectomy was decided. At laparotomy a cystic mass of the appendix was found with gangrenous change in distal part without perforation. The appendix was twisted 360 degrees clockwise at the point of 1cm distal to its base (Fig. 1). A simple appendectomy was performed without spillage of the appendiceal content. During revision a moderate amount of serous fluid was found in Douglas' pouch. There was no regional lymphadenopathy. Macroscopically the removed appendix was 11 cm in length and 4 cm in diameter. The specimen contained yellow-colored

mucin with dark color in the distal part (Fig. 2). The final pathologic diagnosis was a mucinous cystadenoma of the appendix. Postoperative recovery was uneventful and postoperative colonoscopy failed to reveal other colonic abnormalities. The patient was scheduled for close follow-up and is disease free during 1 year.

## Discussion

Appendiceal mucocele is a general term describing the swollen appendix due to abnormal accumulation of mucus within the appendiceal lumen<sup>1</sup>. Mucocele of the appendix was recognized as a pathologic entity by Rokitansky in 1842 and was formally named by Feren in 1876<sup>10</sup>. Up today, few series have been published on this type of appendiceal lesion<sup>1</sup>. Mucocele of the appendix is an infrequent event, representing 0.3%-0.7% of appendiceal pathology and 8% of appendiceal tumors<sup>13</sup>.

Currently the AM includes four histological groups: simple mucocele or retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma<sup>14</sup>. The preoperative diagnosis of AM is often difficult, since its presentation is variable. In a recently published study from Mayo Clinic was found that of 135 patients diagnosed with AM only 48% presented some clinical symptoms: 27% abdominal pain, 16% abdominal mass, 10% weight lost, 9% nausea, vomiting, or both, and 8% had clinical signs of acute appendicitis. Moreover, presence of symptoms was associated with a higher incidence of cystadenocarcinoma<sup>1</sup>.

Unusual AM complications include intestinal obstruction, intussusception, intestinal bleeding, fistula formation, and volvulus<sup>2</sup>.

Since its first description by Payene JE, primary and secondary appendiceal torsion is distinguished<sup>15</sup>. Torsion of the vermiform appendix is a rare disorder inducing clinical signs indistinguishable from acute appendicitis and about 50 cases being reported up to date<sup>4,16</sup>. Primary appendiceal torsion is characterized by secondary ischemic or necrotic change with luminal distension distally to the torsion site in the absence of any primary lesion. Secondary torsion is much more rarely reported and is believed to be induced by an appendiceal abnormality (cystadenoma, mucocele, fecalith impaction, malformation or parasites infestation). The mechanism of AM torsion seems to be similar to that of torsion of the ovary, testicle or appendix epiploicae<sup>4,16</sup>.

Only nine well-documented cases of secondary torsion of the vermiform appendix associated with AM have been reported in the English literature up to date<sup>4-12</sup>. The clinico-pathological characteristics of ten cases of AM torsion are summarized in Table I. The volvulus of AM tend to occur in adults (mean  $\pm$  SD age of  $41.3 \pm 5.9$ , range 18-79 years) sex ratio M:F=1:1. The appendiceal lesions were evenly distributed – simple mucocele and mucinous cystadenoma 1:1.

TABLE I - Summary of secondary torsion of the vermiform appendix associated with AM (English literature).

Case	References	Year	Age/Sex	Tumor size (cm)	Degree/ Direction of rotation	Appendiceal lesion
#1	Dickson DR [5]	1953	60/F	11.5	720/NR	simple mucocele
#2	Chan KP [6]	1965	18/F	10x4	1260/C	simple mucocele
#3	Legg NG [7]	1973	291M	10	360/NR	simple mucocele
#4	Abu Zidan FM [8]	1992	32/F	3.2x3.5x5.9	NR/C	simple mucocele
#5	Moten AL [9]	2002	44/F	NR	360/CC	mucinous cystadenoma
#6	Ruddloff U [10]	2007	28/F	3.3x5	720/C	simple mucocele
#7	Hebert JJ [11]	2007	59/M	12x6x4	NR	mucinous cystadenoma
#8	Kitagawa M [4]	2007	34/M	10x4	180/C	mucinous cystadenoma
#9	Hamada T [12]	2007	79/M	6x3	180/CC	mucinous cystadenoma
#10	Present case	2011	30/M	11x4	360/C	mucinous cystadenoma

C = Clockwise; CC = counterclockwise; M = male; F = female, NR = not reported

As reported by Val-Bernal JF et al., the counterclockwise direction of appendiceal rotation (primary or secondary) is reported more frequently<sup>17</sup>. In cases of volvulus of AM the clockwise rotation is reported more frequently, as in the present case<sup>4-12</sup>. The mean degree of AM rotation was  $517.5 \pm 129.25^\circ$  varying from  $180^\circ$  to  $1260^\circ$ .

Despite specific endoscopic, ultrasound and radiologic features described for AM the correct preoperative diagnosis is rare<sup>4,12,18-20</sup>. It is even more difficult in case of a twisted AM since the clinical signs are indistinguishable from acute appendicitis, the correct preoperative diagnosis was established only in two previously described cases<sup>11,12</sup>.

Simple appendectomy appears to be sufficient for AM of benign origin, while cystadenocarcinomas or involved cecum require right-sided hemicolectomy<sup>1,21</sup>. In the described cases of AM torsion, open or laparoscopic appendectomy was performed<sup>4,6,8-12</sup>. We support the opinion that if a mucocele is visualized during laparoscopy, conversion to open appendectomy should be performed<sup>21</sup>.

The outcome of simple mucocele, mucosal hyperplasia, and mucinous cystadenoma after appendectomy is excellent, 91% survival being reported during a 10-year period<sup>13</sup>. The lack of follow-up data in many case reports regarding AM torsion does not allow us to determine the exact long-term outcome. Taking in account the potential progressing to PMP, our patient was scheduled for close follow-up.

In conclusion, secondary appendiceal torsion due to an appendiceal mucinous cystadenoma is extremely rare and the correct preoperative diagnosis is difficult. Simple appendectomy is the treatment of choice for benign appendiceal mucinous lesions. Open appendectomy should be performed with minimal handling in the uninvolved appendiceal portion in order to limit the possibility of spillage of appendiceal mucocele content.

## Riassunto

La torsione dell'appendice vermiforme è una rara eventualità, ed ancora meno rara è la torsione di tumori mucinosi dell'appendice. A nostra conoscenza fino ad oggi ne sono stati riferiti in letteratura soltanto nove casi di torsione dell'appendice vermiforme associata a tumori mucinosi.

Riferiamo allora un caso di torsione secondaria dell'appendice vermiforme sede di un cistadenoma mucinoso in un uomo di 30 anni. I sintomi erano gli stessi di una appendicite acuta. La laparoscopia diagnostica ha rivelato trattarsi di un tumore mucinoso dell'appendice torta a  $360^\circ$ . Per evitare una disseminazione peritoneale l'intervento fu convertito ad appendicectomia ad addome aperto. La diagnosi istopatologica definitiva fu appunto quella di cistoadenoma mucinoso dell'appendice. La guarigione postoperatoria fu regolare e privo di eventi di rilievo.

La torsione secondaria dell'appendice vermiforme con cistoadenoma mucinoso è una eventualità rara e la diagnosi preoperatoria improbabile. L'appendicectomia ad addome aperto rappresenta la procedura standard per il trattamento dei tumori mucinosi benigni dell'appendice. Inoltre è importante evitare la contaminazione del cavo peritoneale con il contenuto del mucocele appendicolare.

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