Torsion of cecal appendix. Report of the first Italian case and review of the literature

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Abstract

In pediatric patients appendicitis is the most common cause of abdominal pain and surgery. Torsion of vermiform appendix is a rare cause, clinically indistinguishable from appendicitis with usually an intraoperative diagnosis. The first description of vermiform appendix torsion was made by Payne in 1918. Clinical presentation is similar to acute appendicitis. Preoperative investigations play a minimal role. Etiology of this condition is unclear, but is possible to distinguish a primary and a secondary torsion. We report a case of 5-years-old boy who presented with right lower quadrant abdominal pain. His clinical signs, symptoms and investigations mimicked an acute appendicitis. Intraoperatively we found a 720° appendix torsion on its base with its mesentery rotated in counter-clockwise direction. The appendix was gangrenous in appearance. A video-assisted trans-umbilical appendectomy was performed. We describe clinical presentation and management of this rare condition reviewing the literature.

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Introduction

Torsion of vermiform appendix is a rare cause of acute abdominal pain. It was first reported in 1918 by Payne[1] in the British Journal of Surgery, who described the first case of torsion. Two years later Beevors reported a similar case, this time in Lancet.[2] This clinic condition is identical in presentation to acute appendicitis. It is seen more frequently in children than in adults and male:female ratio is 4.5:1.[3] No specific age predominates; the range varies from 50 days[4] to adults[5] with a 76 years old man as oldest patient. Diagnosis of torsion is invariably made intraoperatively. Primary and secondary torsion of the vermiform appendix have been described. Carcinoid tumor and infestation with parasites such as Schistosoma haematobium may also rarely lead to torsion of the appendix.[6,7] Rotation of the appendix has been seen in both clockwise and anti-clockwise directions. The appearance of the appendix varies from twisted with minimal inflammation to severely congested and gangrenous or necrotic. A review of the Entrez PubMed literature documented that totally only 59 cases of volvulus of the appendix have been reported of which 25 are in pediatric patients. Our case will be the 26th and the first Italian case reported.

Case Report

A five-years-old boy presented to Pediatric Emergencies with 12-hours history of continuous periumbilical abdominal pain associated to 6 episodes of vomiting and a diarrhea. Temperature was 38°C. At the initial evaluation the abdominal pain was widespread without signs of peritoneal irritation. Lab results showed mild infective signs (WBC: 18700/mm3, CRP: 0.79 mg/dL) with ketonuria and acetonemic breath. The patient was reassessed after 18 hours of observation, finding an intensified abdominal pain located in the right lower quadrant area with signs of peritoneal irritation, abdominal wall defense/muscular rigidity and positive Blumberg's sign. A complete blood count repeated showed leukocytosis (16,400 wbc/mm3 with 84,7% neutrophils). Abdominal ultrasound showed thin fluid film contouring intestinal loops in both small pelvis and right iliac fossa where they appear distended by liquid. In right iliac fossa also, was described a liquid flap with

a maximum thickness of 8mm and a rounded 12mm hyperechoic area suspected for collection. The appendix was not visualized. An emergency operation was performed under the diagnosis of acute appendicitis. The operation was performed using the conventional video-assisted trans-umbilical laparoscopic technique. Laparoscopy showed inflammatory fluid in pelvic cavity and an indurated area in the right iliac fossa/parietocolic spaces. Lysis of adhesions between omentum, small intestine, and cecum was performed, identifying a gangrenous appendix, 6-7 cm in length and 1cm in diameter, twisted two times (720°, Figure 1) in counter-clockwise at the point of 1.5 cm distal to its base and attached to the abdominal wall of the right parietocolic spaces (Figure 2). The torsion leads to a congestion of appendix vessels wall and venous thrombosis (Figure 3a and 3b). The appendix presented a narrow appendicular mesentery and a movable cecum without fixation. A video-assisted Trans-Umbilical Appendectomy (TULAA) was performed, exteriorizing the appendix and the cecum through the umbilical scar. Operative time was 50 minutes. Feeding was started on 1st postoperative; the boy was discharged on second day. Histological examination of appendix showed transmural inflammation and necrosis with fibrino-leucocytic infarction.

Discussion

Torsion of the vermiform appendix is a rare cause of acute abdominal pain. It occurs as a result of twisting of the appendix along its longitudinal axis. The site of torsion is usually, at least 1 centimeter from the base of the appendix and is less frequent at the base. Torsion degree is usually between 180-1080° while the direction of rotation is variable, but counter-clockwise is more frequent. We are unable to identify any association between the degree of rotation and etiology or presentation in view of the relatively small sample size. All that is known is that a rotation of 120 degrees is enough to compromise vascular supply in some patients. Torsion

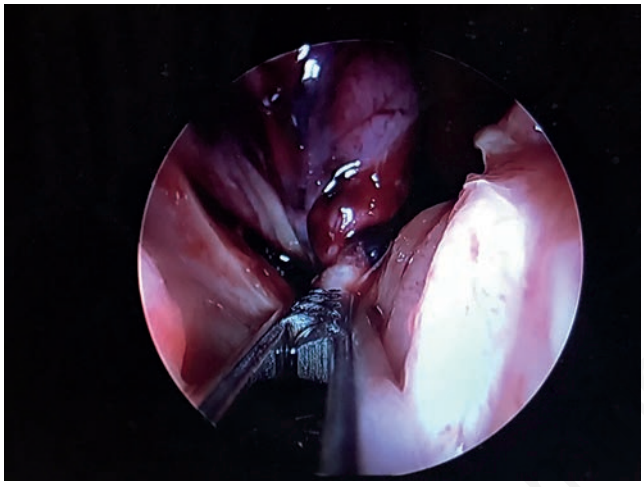

Figure 1. Twisted appendix with a torsion of 720° in counter-clockwise direction.

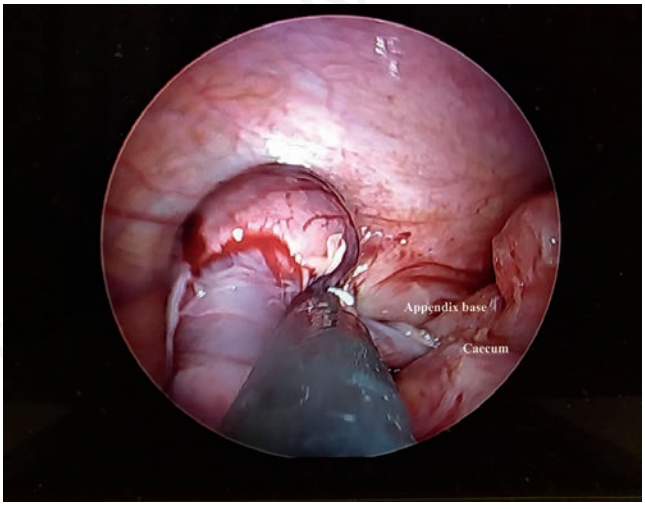

Figure 2. Normal appendix base 1.5 cm distal to the site of torsion.

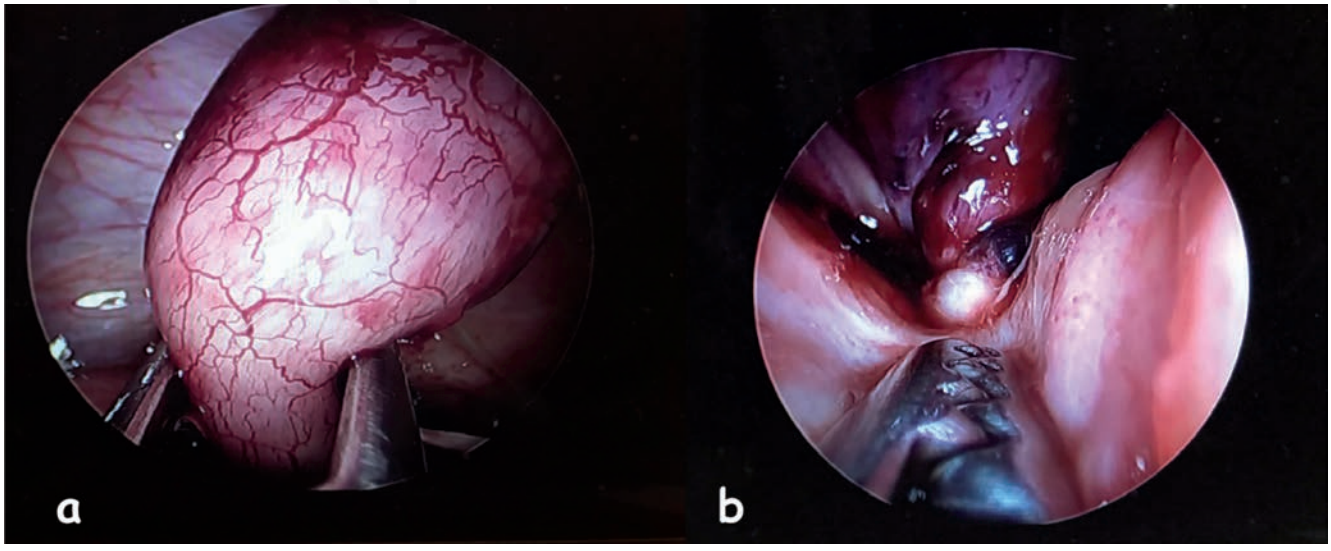

Figure 3. a) Vessel wall appendix congestion; b) Particular of torsion with venous thrombosis.

leads to lumen obstruction compromising the lymphatic drainage and venous return obstructing the arterial supply, resulting in strangulation, hemorrhagic infarct and consequent inflammatory response, which leads clinically to an acute abdomen. From the first description by Payne in 1918,[1] by a review of the Literature we found other papers reporting a description of this condition. Montes-Tapia *et al.* in 2009[6] reported that 15 pediatric patients were describe previously, while in 2012 Dimitriadis *et al.*[8] in reviewing the English literature described a total of 30 cases reported in adult populations including their case and 16 cases in pediatric patients. Adi *et al.*[9] in 2014 wrote that torsion of appendix has been reported 30 times in the last century, half of which were in pediatric patients; Ioannis *et al.*[10] in 2017 cited that 17 cases have been reported in children while Endo *et al.* in 2020 described 22 pediatric cases including his one.[11] We reported our case, the first Italian, and the 26th in pediatric population reported by English literature. Mean age in this pediatric cohort of 26 children is 6.5 years and males are predominant (84,6%); the appendix torsion varies from 270–1080 degrees. Among the predisposing conditions: i) a duplicate appendix was found in one patient, ii) an absent mesoappendix (which was proposed as a predisposing contributory factor) in three; iii) a narrow appendicular mesentery and movable cecum in two patients including our one. Vomiting is present in almost all patients as a presenting symptom, fever in 20 (about 77%). Length of the appendix is reported in 16 patients (approximately 73% of cases) varying from 5 to 15 cm. Only one among the reported etiology was a secondary torsion (simple mucocele); in the remaining cases was a primary torsion. The predominant preoperative diagnosis was acute appendicitis (peritonitis-perforated appendicitis with periappendicular abscess) (Table 1). Is not possible to identify any association between the rotation, its degree and clinical presentation. Clinical presentation is similar to acute appendicitis. The two conditions remain clinically indistinguishable, and their management should be the same.[31] In pediatric cases after admission, observation usually is adopted followed by emergency operations because symptoms did not improve.[11] Etiology of appendix torsion is not known. It is postulated that various pathologies might be responsible. Primary and secondary torsion of vermiform appendix have been described. Primary torsion seems to happen more often in children while secondary torsion is noticed mainly in adults.[10-32] In a review of 33 adults with appendiceal torsion, reported during 1918–2018, only one had an associated congenital anomaly "cecal malposition".[11,30,33] In primary torsion is hypothesized that the axial rotation is due to different possible cause: **i)** to abnormalities of the mesentery, such as a narrow base, as occurred in our case, ii) to the

Table 1. Cases of appendiceal torsion reported in the English literature (UD: undetermined).

N° cases	Author	Year	Age	Sex	Degree	Lenght	Etiology of torsion	Fever (cm)	Nausea/ vomiting	Preoperative diagnosis
1	Carter[12]	1959	8	F	>360°	-	UD	38.3°	Vomiting	UD
2			16	M	>360°	-	UD	37.4°	Nausea	UD
3	Chan[13]	1965	18	F	1260°	10	Simple mucocele	37.3	No	Acute appendicitis
4	Ghent[14]	1966	12	M	360°	7	Primary	37.6°	Nausea	UD
5	Finch[15]	1974	12	M	270°	-	UD	37.2°	Vomiting	UD
6	Willan[16]	1983	4	M	720°	7	UD	37.3°	Vomiting	Acute appendicitis
7	Dewan[17]	1986	3	M	720°	7	UD	37.9°	Vomiting	UD
8			6	F	1080°	7	UD	37.4°	Vomiting	UD
9			16	M	-	-	UD	No	UD	UD
10	Waters et al[18]	1986	3	M	720°	-	UD	38.9°	Vomiting	Exploratory lap
11	Yeung *et al.*[19]	1991	50 days	M	-	-	UD	High fever	-	Generalised peritonitis
12	Merret *et al.*[20]	1992	14	M	720°	14	Normal Appendix	37.5°	Vomiting	Acute appendicitis
13	Gilchrist[21]	1995	6	M	360°	9	Long narrow mesoappendix	37.2°	Vomiting	UD
14	Val-Bernal *et al.*[22]	1996	6	M	>360°	13.5	Primary	37.5°	Vomiting	Acute appendicitis
15	Uroz-Tristan *et al.*[23]	1998	5	M	360°	15	Absent mesoappendix	No	Vomiting	Torsion or mucocele
16	Oguzkurt *et al.*[24]	2004	2	M	270°	10	Duplicated colon and appendix	38°	Vomiting	UD
17	Gopal K *et al.*[25]	2005	9	M	720°	5	Primary	No	Vomiting	Acute appendectomy
18	Sarin[26]	2006	9	M	270°	8	Normal appendix	37.7°	Vomiting	UD
19	Montes Tapia[6]	2009	3	M	1080°	-	Narrow appendicular mesentery and movable cecum	UD	Vomiting	Acute appendicitis
20	Lena Perger[27]	2011	11 w	F	360°	-	UD	Low-grade fever	Emesis	Acute appendicitis
21	D'Souza[28]	2011	2	M	—	6.5	UD	38.3°	Vomiting	Acute appendicitis
22	Hirpara[29]	2018	2	M	720°	7.5	Lymphoid hyperplasia	Low-grade fever	Emesis	Acute appendicitis or Meckel's diverticulitis
23	Endo *et al.*[11]	2019	4	M	720°	8	Primary	37.2	Vomiting	Acute appendicitis
24	Samuk30	2020	40 months	M	-	-	UD	UD	Vomiting	Acute appendicitis
25			23 months	M	1080°	-	UD	UD	Vomiting	Perforated appendicitis with periappendicular abscess
26	Present study	2021	5	M	720°	6-7	Narrow appendicular mesentery and movable cecum	38°	Vomiting	Acute appendicitis

absence of azygotic folds that normally laterally fix the appendix, iii) to cecal malposition, iv) to abnormal peristaltic movements and v) to vigorous physical exercise. Secondary torsion of appendix is rarest. It occurs in association with a mucocele, bilharzia, fecaliths, intussusception, mesoappendiceal lipoma, cystadenoma, duplication of appendix; but also, adhesions and inflammation causing distention could favorite instability and a tendency to twist.[32,34,13] Carcinoid tumors and infestation with parasites such as Schistosoma haematobium may also rarely lead to torsion of the appendix. The features that are commonly associated with torsion of appendix include long appendix and pelvic position of the appendix. An abnormally long appendix was described in most studies with a mean length of 9.6 cm.[8] Clinical presentation of vermiform appendix volvulus may be similar to an appendicular abscess/acute appendicitis with or without perforation both on physical examination and imaging.[28] In our case ultrasound showed in the right iliac fossa, a presence of a liquid flap, an hyperechoic area suspected for collection (abscess). Imaging does not play major role in these cases. Abdominal ultrasound has been used as first step imaging study to find the vermiform appendix, in the differential diagnosis of the most frequent causes of pain in the right iliac fossa, but is difficult to describe an appendicular torsion. In our case, the appendix was not found. In literature, there're no data reported about US-imagine criteria of appendicular torsion. Only Uroz-Tristan and co-workers reported a case where US examination rendered torsion beside the inflammation of the appendix,[23] while Hamada *et al.*[35] reported that target-sign like appearance was a useful finding in identifying appendiceal torsion. Treatment is simple appendicectomy if diagnosis is prompt and intervention is earlier before complications by the conventional approach or by laparoscopy.

Conclusions

In children, the appendix can be source of rare pathological entities that can present as surgical emergencies. Torsion of the vermiform appendix is a very rare disease and difficult to differentiate from appendicitis. The short interval between arrival at the Emergencies Department and the timing of surgery reflects the acuteness of clinical presentation and the need for an emergent operation. Nowadays thanks to minimal invasive surgery, laparoscopy can prove the torsion and the twisted appendix can be removed laparoscopically or by video-assisted trans-umbilical laparoscopy as in our case. There is a consensus that the appendicular volvulus cannot be distinguished from acute appendicitis preoperatively. However, is possible to conclude that laboratory tests (lack of characteristic signs of inflammation), the advancements in US imaging and the rapidly growing use of diagnostic (preoperative) laparoscopy might modify this general view.

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