Frontiers in Medical Case Reports, ISSN: 2582-8142

A Rare Case of an Unusual Complication of Appendicitis in A Child with Cystic Fibrosis

Valentina Angelini[1] | Antonella Tosco[2] | Piero Trovato[1] | Angela Sepe[2] | Gianfranco Vallone[3] | Valeria Raia[2] | Maria Grazia Caprio[4]*

***Correspondence:** Maria Grazia Caprio

Address: [1]Department of Oncoematologia, Diagnostica per immagini e morfologica e Medicina Legale, University of Naples Federico II, Naples, Italy; [2]Paediatric Unit, Department of Translational Medical Sciences, Regional Cystic Fibrosis Centre, University of Naples Federico II, Naples, Italy; [3]Department Life and Health "V. Tiberio", University of Molise, Francesco De Sanctis st 1, 86100, Campobasso, Italy; [4]Institute of Biostructure and Bioimaging IBB, CNR, Via De Amicis, 95 80145 Naples, Italy

e-mail ✉: mariagrazia.caprio@ibb.cnr.it

Received: 11 May 2020; **Accepted:** 19 May 2020

ABSTRACT

This case regards of an unusual complication of appendicitis in a child with Cystic Fibrosis (CF) who presented delayed presentation and appendiceal abscess formation. In pediatric CF the incidence of appendicitis is lower than in unaffected children, but the diagnosis is often made already in the complicated phase. In these patients, it would be useful to remember the different conditions of differential diagnosis in abdominal pain to reduce the number of misdiagnoses and to set up timely medical therapy by reducing the number of acute surgical abdomens. Ultrasonography is a rapid, non-invasive technique that should be considered when patients suffering from CF present with new-onset abdominal pain.

Keywords: *Cystic Fibrosis, Appendicitis, Abdominal Pain*

Introduction

CF is a disease of secretory glands caused by mutation of the Cystic Fibrosis Transmembrane Regulator (CFTR) gene. The clinical manifestations of CF are repetitive lung infections, biliary cirrhosis, pancreatic abnormalities and gastrointestinal disorders (Chen *et al*., 2012). The clinical consequences of absent or reduced CFTR ion channel function result in complex, multiorgan characteristics of the CF phenotype, which are site-specific but vary considerably in severity and age of onset (Wilschanski and Durie, 2007). Gastrointestinal manifestations in CF may be divided into two groups: those where the pathobiology is directly related to the basic defect of CF and those which arise as a secondary complication of the disease or its treatment (Wilschanski and Durie, 2007). Some studies supposed that, except for the exocrine pancreas, manifestations of CF disease in the liver and the intestinal tract are primarily associated with patients who have classic CF disease, most of whom carry severe class of CFTR mutations on both alleles (Wilschanski and Durie, 2007). These mutations may lead to dysregulation of

the inflammatory responses, too. It is biologically plausible that this altered inflammatory response should be present not only in the lungs, but in other organs where CFTR is expressed (Smyth *et al*., 2000). Several studies have shown evidence of intestinal inflammation in CF.

Acute appendicitis develops in up to 1.5% of patients with CF, a rate significantly lower than 7% observed in the general pediatric population (Shields *et al*., 1991). Diagnosis is difficult, often delayed: abdominal pain in children with CF is frequent and often attributed to other causes, such as intussusception (Shields *et al*., 1991; McCarthy *et al*., 1984; Coughlin *et al*., 1990).

Methods and Results

An 11-year-old Caucasian child with CF presented with an 8-day history of intermittent migrating abdominal pain. The pain was constant, initially presenting in the epigastric region, then radiating to the right and left upper quadrants and the right lower quadrant. Back and right thigh pain had been noted in the 6 days before abdominal pain presentation. There was the suspect of subocclusion because of the absence of evacuation from 4 days and concomitant nausea and vomit. There was no history of abdominal rigors or sweats. No similar attacks had been experienced in the past. The patient was taking pancreatic enzyme supplements and pulmonary antibiotics prophylaxis because of the presence of some typical CF findings on CT, such as bronchiectasis and COBP (Fig. 1).

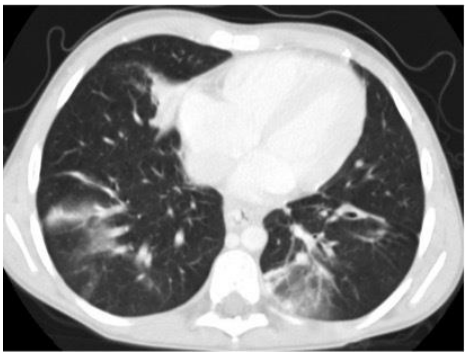

Figure 1: Axial frame of CT. At the lung bases there are bilateral disventilatory strips, to be attributed to the COBP.

Laboratory findings revealed a white cell count of 15.1 with a neutrophilia. Chest and abdominal X-rays reported irregular representation of intestinal meteoric aria with poor representation of this in sigmoid-rectum colon tract associated with hydro-air level in these regions.

Abdominal ultrasound revealed a high wall thickening of vermiform appendix (DMax:13mm) and the terminal ileum (one wall Diameter: 8mm) with a moderate bordering fluid component and with a diffuse phlegmonous inflammatory peri-appendicular mass. This mass presented an ana-ipoechoic aspect and multiple septa. Mesenteric fat had a hyperechoic aspect with some reactive lymph nodes within (Fig.

2-4).

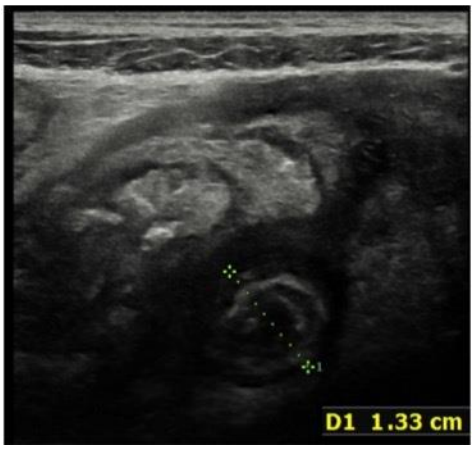

Figure 2: US scan (linear probe 12MHz) in the right iliac fossa showing the appendix in cross-section. The wall stratification is altered and the transverse appendicular diameter is higher than normal (greater than 6 mm).

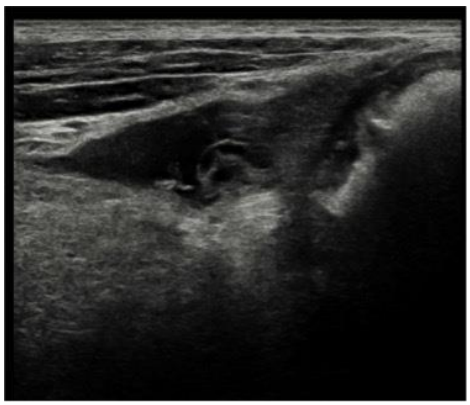

Figure 3: US scan (linear probe 12 MHz) showing fluid flap between the intestinal loops.

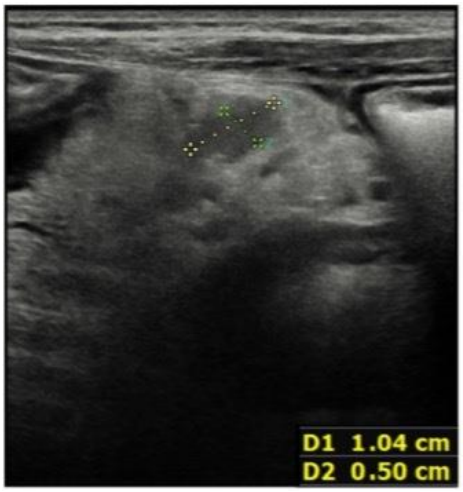

Figure 4: US scan (linear probe 12 MHz). Mesenteric fat had a hyperechoic aspect with some reactive lymph nodes within.

This aspect is suggestive of appendicitis complicated by abdominal abscess.

The abdomen CT performed to the little patient confirmed the congestion and tumefaction and the high wall thickening of vermiform appendix and the presence of a peri-appendicular fluid concentration with multiples air nucleus within. These evidences were suggestive of peri-appendicular abscess in complicated appendicitis (Fig. 5).

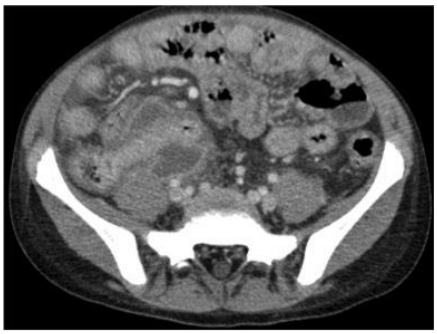

Figure 5: axial frame of CT abdomen post-mdc (venous phase). There are congestion and tumefaction and the high wall thickening of the vermiform appendix and the presence of a peri-appendicular fluid concentration with multiples air nucleus within. These shreds of evidence are suggestive of periappendicular abscess in complicated appendicitis.

The patient was treated with abdominal percutaneous drainage guided by ultrasound and with intravenous cefotaxime, metronidazole and meropenem. Aggressive chest physiotherapy was implemented. The patient progressed rapidly from oral intake of fluids to a normal diet after 15 days.

An MRI enterography was performed after 3 months and showed a dimensional reduction of the abscess and a fibrotic evolution of this one (Fig. 6).

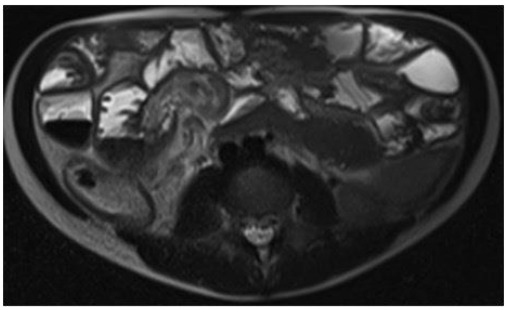

Figure 6: Axial T2 frame of entero-MR after 3 months from the acute phase. There is a dimensional reduction of the abscess and a fibrotic evolution of this one.

The ultrasound exam of bowel was repeated about 6 times in a year and after 12 months the abscess appeared completely commuted in fibrotic tissue and the appendix wall thickening was normal and of regular wall stratification.

Discussion

Despite the fact that the appendix is frequently swollen and distended with inspissated eosinophilic secretions in patients with cystic fibrosis (Lardenoye *et al*., 2004) the reported incidence of acute appendicitis in CF patients is lower than that in the general population (Jaffe *et al*., 1966; Holsclaw *et al*., 1974; Barker, 2002).

Diagnosis of appendicitis in CF patients may be difficult and is often delayed. The antibiotic treatment prescribed for respiratory infections may mask a patient's general pathological condition until perforation or abscess formation occurs (McCarthy *et al*., 1984); the delay may be due to a more indolent or atypical presentation, too (Shields *et al*., 1991). For this reason, and since CF patients generally have a poor respiratory status, their morbidity and mortality following appendicectomy is significantly higher (Shields *et al*., 1991).

The migrating nature of our patient's abdominal pain, probably due to the length of the inflamed appendix irritating different parts of the peritoneum, contributed to the diagnostic delay.

Other more common causes of abdominal pain in these patients, such as meconium ileus equivalent, intussusception, faecaliths, muscular aches associated with coughing, cholelithiasis, and volvulus may be proposed earlier before the correct diagnosis is made. Delay in seeking medical attention after the onset of symptoms is also a contributing factor (Barker, 2002; Martinez-Garcia *et al*., 2005).

The study of the gastrointestinal tract by imaging, in particular by ultrasound, is a necessary tool for the diagnosis of acute and chronic gastrointestinal pathologies in children. In particular, ultrasound has a primary role in the evaluation of the gastrointestinal tract in pediatric patients both in routine and in emergency (Esposito *et al*., 2019).

After a correct diagnosis, conservative treatment was carried out in that child. Following laparotomy in CF patients, in fact, prolonged postoperative ileus is not uncommon and hence a conservative approach with or without total parenteral nutrition should be considered (Koletzko *et al*., 1989).

Conclusion

In conclusion, patients with the refractory or unexplained intestinal symptoms in pediatric and adult CF should not be misunderstood and these patients deserve more investigations. Ultrasonography is a rapid, non-invasive technique that should be considered when patients suffering from CF present with new-onset abdominal pain, mass or fluid discharge from the right flank; in fact, with a prompt US

evaluation, the correct diagnosis and a consequent non-invasive therapy could be easily achieved.

Funding Sources: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of Competing Interest: There are no conflicts of interest to disclose.

Acknowledgements: None.

References

Barker AF. "Bronchiectasis". *N Engl J Med* 2002; 346: 1383-1393.

Chen CH, Chang CC, Yang BY, Lin PY, Wang CS. Acute appendicitis mimicking intestinal obstruction in a patient with cystic fibrosis. *J Formos Med Assoc* 2012; 111: 580-3.

Coughlin JP, Gauderer MW, Stern RC, Doershuk CF, Izant Jr RJ, Zollinger Jr RM. The spectrum of appendiceal disease in cystic fibrosis. *J Pediarr Surg* 1990; 25: 835-839.

Esposito F, Di Serafino M, Mercogliano C, Ferrara D, Vezzali N, Di Nardo G, Martemucci L, Vallone G, Zeccolini M. The pediatric gastrointestinal tract: ultrasound findings in acute diseases. *J Ultrasound* 2019; 22: 409-422.

Holsclaw DD, Rocmans C, Shwachman H. "Abdominal complaints and appendiceal changes leading to the diasis of cystic fibrosis." *J Pediatr Surg* 1974; 9: 867-873.

Jaffe BF, Graham WP, Goldman L. "Postinfancy intestinal obstruction in children with cystic fibrosis." *Arch Surg* 1966; 92: 337-343.

Koletzko S, Stringer DA, Cleghorn GJ, Durie PR. "Lavage treatment of distal intestinal obstruction syndrome in children with cystic fibrosis." *Pediatrics* 1989; 83: 727-733.

Lardenoye SW, Puylaert JB, Smit MJ, Holscher HC. Appendix in children with cystic fibrosis: US features. *Pediatric Imaging* 2004; 232: 187-189.

Martinez-Garcia MA, Perpina-Tordera M, Roman-Sanchez P, Soler-Cataluna JJ. "Quality-of-life determinants in patients with clinically stable bronchiectasis". *Chest* 2005; 128: 739-745.

McCarthy VP, Mischler EH, Hubbard VS, Chernick MS, Di Sant'Agnese PA. Appendiceal abscess in cystic fibrosis A diagnostic challenge. *Gastroenterology* 1984; 86: 564-568.

Shields MD, Levison H, Reisman JJ, Durie PR, Canny GJ. Appendicitis in cystic fibrosis. *Arch Dis Child* 1991; 66: 307-310.

Smyth RL, Croft NM, O'Hea U, Marshall TG, Ferguson A. Intestinal inflammation in cystic fibrosis. *Arch Dis Child* 2000; 82: 394-399.

Wilschanski M and Durie PR. Patterns of GI disease in adulthood associated with mutations in the CFTR gene. *Gut* 2007; 56: 1153-1163.