Intestinal Transmesenteric Plication in a Cystic Fibrosis Child with Recurrent Distal Ileal Obstruction Syndrome

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Abstract

Recurrent intestinal obstructions are frequently reported in Cystic Fibrosis patients at any age. The most frequent diagnoses are constipation and distal ileal obstruction syndrome, however in previously operated patients, post-surgical adhesions must be considered in the differential diagnosis of recurrent abdominal pain. We report the case and the follow-up of a 4-years-old Cystic Fibrosis child with recurrent intestinal obstruction after neonatal abdominal surgery for meconium ileus. The decision making process to treat symptoms is described. The chosen surgical procedure proved to prevent any further occlusive episode in a ten years follow up.

ABBREVIATIONS

CF: Cystic Fibrosis; DIOS: Distal Ileal Obstruction Syndrome; MI: Meconium Ileus

INTRODUCTION

Cystic Fibrosis (CF) patients frequently suffer from recurrent intestinal sub-occlusive episodes during their lifetime; constipation and distal ileal obstruction syndrome (DIOS) are largely the most common diagnosis. Nevertheless these clinical events are more frequently reported in CF patients who presented at birth with meconium ileus (MI) requiring surgery in the first days of life [1].

MI, a complete intestinal obstruction caused by the intraluminal impaction of dehydrated and thickened meconium, is the earliest manifestation of CF [2]. Hyper echogenic fetal bowel with dilation detected by ultrasound during the second trimester of pregnancy must place the suspect for CF [3].

European CF patients’ registry reports an 18% frequency of MI for European CF population in 2009, while a recent European multicenter study showed that about 40% of all CF patients suffering from DIOS presented at birth with MI [4].

These data confirm the high prevalence of DIOS in CF patients with MI as both these conditions probably share the same pathophysiology, such as slow intestinal transit and impaired intestinal secretion flow leading to sticky mucus.

In CF patients who underwent surgery for MI, differential diagnosis should include also postoperative adhesions syndrome, which is reported as a late complication of intraperitoneal surgery with a variable incidence depending both on the primitive disease and the surgical procedure [5].

Every recurrent occlusive episodes in CF/MI patients show grossly the same clinical presentation, making sometimes hard to differentiate between DIOS and a possibly post surgical adherence syndrome.

We report the case of a CF male patient with MI requiring surgery in the first day of life, who presented several intestinal occlusive episodes since the age of two years. An extended adhesiolysis together with a Childs-Phillips procedure (transmesenterical plication) as a prophylaxis against recurrences solved the episodes of bowel obstruction.

CASE REPORT

L.C. was born at full term with proper weight for gestational age and presented a neonatal intestinal obstruction requiring surgery in the first day of life. Abdominal exploration showed a meconium ileus which required about 20 cm ileal resection with a temporary Bishop-Koop ileostomy.

Cystic Fibrosis was suspected by a positive newborn screening program (IRT/DNA) with two CFTR mutations (N1303K/711+1G→T). The infant was therefore referred to our...
Central age, whose recurrent intestinal occlusive episodes dramatically...