Considering the acute course of the disease in our patient, it seems likely that gastro-intestinal aspergillosis preceded the pulmonary and cardiac aspergillosis. The gastro-intestinal ulcers may have been the primary site from which further infective dissemination may have occurred. Subsequently, these ulcers perforated and peritonitis developed. Also, there was no evidence of immune-compromised status of our patient.

Thus, this is a very rare clinical presentation of fatal invasive aspergillosis in an immune-competent elderly male.

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Megacystic microcolon intestinal hypoperistalsis syndrome with mydriasis in a male child

Introduction

Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) is a rare, congenital disease affecting smooth muscle peristalsis mainly in the gastrointestinal tract and urinary bladder. Also known as Berdon disease, as first described by W E Berdon and coworkers[1], is a severe and often lethal form of neonatal pseudo obstruction. It is characterized by hypoperistalsis or a peristalsis of the gastrointestinal (GI) tract, malrotation, microcolon, and genitourinary abnormalities namely, a non-obstructed dilated bladder[2]. The prognosis is extremely poor, with most patients dying within the first 6 months without treatment[3].Although the disease is often diagnosed in female infants, we describe a male child with late diagnosis in childhood with bilateral mydriasis.

Caser report

A 6 year old male child admitted with recurrent vomiting, abdominal pain, and failure to thrive since 1 year of age. Patient had normal growth velocity upto age of 1 year. After that weight and height did not increase as compared to other sibling. However language and motor milestones were normal. Patient had abdominal distention since last 1 year which was painless and more in the central and lower abdomen. His weight was 13 kilograms and height was 80 cm, both of which were below 3rd percentile for his age .His birth weight was 2.7 kilograms and was full term. He had one elder brother, who wasnormal.On examination patient had distended abdomen which was tympanic without evidence of any free fluid or organomegaly. Haematological and biochemical investigations showed mild anaemia. Abdominal X ray showed dilated bowel loops without significant air fluid levels or free gas. Barium study showed slow transit of barium in dilated bowel loops with normal ileocaecal valve and colon. CECT abdomen suggestive of dilated small bowel loops with moderate enlargement of left pelvicalyceal system, left entire ureter & over distended bladder. DMSA renal scan of both right and left kidney showed normal functioning cortical mass with no evidence of any cortical defect. Micturating cystourethrogram showed distended bladder with smooth outline without any evidence of residual urine, filling defect, diverticulum, vesicoureteral reflux with normal anterior and posterior urethral valves. Upper G.I endoscopy showed dilated esophagus, dilated stomach and dilated duodenum. However there was no evidence of any mechanical obstruction. Esophageal manometry was done to

rule out motility disorder which was completely normal. Colonoscopy was normal with no evidence of any mechanical obstruction and mucosa was normal.

Full thickness rectal biopsy taken to rule out any visceral myopathy which showed expanded myentericplexus, increased nerve trunks and mature ganglion cells within myenteric plexus. Muscular layer didn't show any vacuoles. All these histological features were suggestive of megacystis microcolon intestinal hypoperistalsis syndrome.

On retrospective examination we also found bilateral mydriasis which failed to respond to pilocarpine, which was also reported in previous case reports[4]**.** Both parents underwent renal ultrasounds to rule out possibility of an autosomal dominantly inherited condition with reduced penetrance. No renal abnormalities were identified in parents. We have planned for proximal ileostomy to defunction distal colon. Total parenteral nutrition was started.

Discussion

Our patient presented with megacystis and proximally dilated small bowel loops, clinical features of Berdon syndrome. Berdon et al[1], first described what he called megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) in five infant girls, two of whom were sisters. MMIHS is a rare, severe form of functional intestinal obstruction in the newborn, and only more than 200 MMIHS cases have been reported in the literature[5]. The frequency of the disease is observed three or four times more in girls than in boys. The clinical course of MMIHS is complicated, and patients usually become symptomatic early in life[5]. A consensus on the theory of the pathogenesis of the disease does not exist yet. Some of the theories are the lack of nicotinic acid receptor subunits, a defect in fiber synthesis, an inflammatory process of the gastrointestinal and urinary tract, generalized axonal dystrophy in central, peripheral and autonomic nervous system[6]. Treatment is supportive and involves an ileostomy to defunction the colon, with TPN. Because of the gastrointestinal dysmotility, attempts to give enteral feeding may cause fatal pneumonia; on the other hand, to prevent malnutrition, most patients with MMIHS are maintained by parenteral nutrition, which may lead to the loss of venous access, thrombosis of vascular access, catheter sepsis and chronic liver failure[5]. Long term survival is sometimes noted in patients with MMIHS receiving TPN; however, most patients develop complications from prolonged TPN[5]. Therefore, the prognosis of MMIHS is poor, and most patients die early in life. Major causes of death are sepsis, malnutrition, renal insufficiency, and multiple organ failure[1].

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Uncommon presentation of Strongyloidiasis: chronic malabsorption, multiple small bowel strictures and appendicitis in HTLV-1 positive patient