Acute Gastroenteritis as Initial Clinical Presentation of Autoimmune Pancreatitis

125

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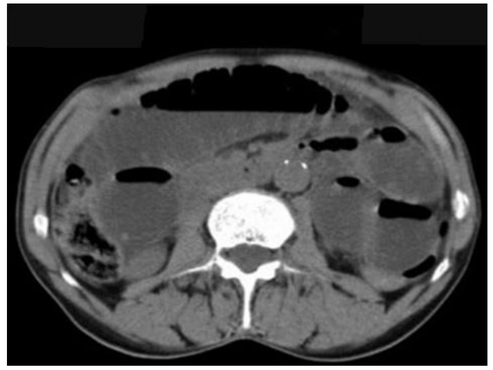
Background

Autoimmune pancreatitis (AIP) is a distinct subtype of complex immune-mediated inflammation of the pancreas that can also occur in children. Disease-specific diagnostic criteria have been developed for AIP in adults, but not for children. Pediatric AIP has a distinct presentation with features similar to type 2 AIP in adults. Abdominal pain along with obstructive jaundice are the most common symptoms and serum IgG4 is rarely elevated in children although increased serum IgG4 is very suggestive of AIP.



Abdominal CT scan
Distended and meteoristic intestinal loops and present aeroliquid levels

Methods

Six years old girl presented with symptoms of acute gastroenteritis acompained with stomach pain. On admission subfebrile, pale, moderately dehydrated, tachycardic with difuse abdominal pain on palpation. Laboratory tests revealed hypoglycemia=3,2 mmol/l, elevated levels of serum amylase=707 U/L and lipase=1097 U/L and urine amylase=1776.22 U/L. Abdominal ultrasound and CT with distended and meteoristic intestinal loops and present aeroliquid levels. Serology tests for HAV, HBC, HCV, TORCH, EBV were negative, antibodies to tissue transglutaminase within normal limits. Parenteral rehydration and nutrition were started, and additionally included a third-generation cephalosporin, proton pump inhibitor, Somatostatin.

Results

Due to the persistence of high pancreatic enzyme values (Amylase=707 U/L, Lypase= 1097 U/L) despite the therapy, MR cholangiopancreatography was performed with normal findings. Genetic testing for hereditary pancreatitis was negative, but we found elevated IgG4 levels=312.2 mg/dL which is why a systemic corticosteroid was additionally included in the therapy and oral nutrition was reintroduced, with gradual reduction of pancreatic enzymes. Because endoscopic ultrasound imaging is not available at our center, we decided to forego pancreatic biopsy in our patient. Further follow-up of the child was outpatient with normal pancreatic enzyme values with a gradual reduction in the dose of systemic corticosteroid. One year after cessation of therapy, the child has normal pancreatic function and pancreatic enzyme values and normalization of IgG4.

Conclusion

AIP should be studied in children because clinical manifestations can be heterogeneous and serum IgG4 is rarely elevated. Even though the frequency of the disease in the pediatric population is low, multicenter studies are needed to characterize the clinical presentation, diagnosis, and progression of AIP in children.

References

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