VIPoma of pancreas in a child

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An eleven-year-old girl had massive watery diarrhea. She was found to have pancreatic VIPoma. It responded favorably to surgical resection of the tumor. There was no tumor recurrence at 18 months of follow-up. [Indian J Gastroenterol 2000;19:194-195]

Key words: Diarrhea, pancreas tumor, vasoactive intestinal polypeptide

Tumor secreting vasoactive intestinal polypeptide (VIP) has been identified as a cause of secretory diarrhea. VIPoma arising from the pancreas is rare in children, most reported cases being ganglioneuromas or ganglioneuroblastomas in the neck, thorax, adrenals or pelvis. We report a child with pancreatic VIPoma.

An eleven-year-old girl presented with intermittent, large-volume diarrhea of two years’ duration. The stool volume was more than two liters per day and diarrhea persisted during fasting. There was no history of laxative abuse or abdominal surgery. The diarrhea was not associated with abdominal cramps, joint pain, fever, skin rash or lymphadenopathy. Physical examination revealed moderate dehydration and her weight was below the third percentile for age.

Investigations: Routine laboratory tests including liver profile were normal. Serum creatinine was 2.6 mg/dL, sodium 150 mEq/L, chloride 109 mEq/L, potassium 2.9 mEq/L (less than 2 mEq/L on three occasions during hospital stay), bicarbonate 14 mEq/L, magnesium 1.82 mEq/L, calcium 10 mg/dL, phosphorus 2.1 mg/dL; normal serum immunoglobulins, lipid profile and prothrombin time. Urine osmolality was 238 mOsm/Kg H₂O. Twenty-four-hour urinary excretion of sodium was 47 mEq/L, potassium 13 mEq/L, calcium 65 mg/dL and phosphate 88 mg/dL. She tested negative for antibodies to human immunodeficiency virus (ELISA). Fasting gastric juice pH was 5.0.

Stool examination was negative for parasites and pathogens on several occasions. Seventy-two-hour fecal fat excretion was 35 g whereas urinary d-xylose excretion test was mildly impaired. Fecal potassium and sodium concentrations were 78.8 mEq/L and 30 mEq/L, respectively, with stool osmolality of 226 mOsm/Kg H₂O.

Barium meal follow-through examination was normal.
Fig: Immunohistochemical stain showing diffuse positivity (arrow) with antibodies for VIP (20 X)

Ultrasonography revealed grade 1 parenchymal changes in the kidney and a cystic area 2 cm in size in the body of the pancreas. Upper gastrointestinal endoscopy was normal. Duodenal biopsy did not reveal any parasites or crypt/ villous changes. MRI (T1W axial imaging) showed a 5 cm x 4 cm x 3.5 cm well-defined soft-tissue mass in the body of the pancreas. Splanchic vessel angiography showed tumor blush in the corresponding area with multiple feeding arteries.

Preoperatively, the diarrhea was controlled with omeprazole. During surgery, palpation of the pancreas revealed a single 4 cm x 3 cm tumor in the body and tail junction. Superior mesenteric artery and portal vein were not involved. Distal pancreatectomy and splenectomy was done. Histology of the resected specimen showed pancreatic tissue with partly circumscribed tumor composed of papillary structures with thin fibrovascular cores and lined by columnar to cuboidal epithelial cells with vesicular nuclei and moderate amounts of cytoplasm. Immunohistochemistry (Fig) showed staining of tumor cells with monoclonal antibodies to VIP. Peripancreatic lymph nodes were also involved by the tumor.

After surgery, diarrhea stopped completely. At 18 months of follow up, she was completely asymptomatic. Repeat CT scan of the abdomen did not reveal recurrence of tumor.

The most consistent features associated with VIPoma are episodic severe secretory diarrhea, hypochlorhydria, hypokalemia and metabolic acidosis. VIP has been shown to stimulate adenylyl cyclase activity and hence secretion by the enteroctyes. Only 20% of patients with VIPoma exhibit flushing. Other abnormalities like hypomagnesaemia and hypophosphatemia, as seen in this case, are possibly secondary to profuse diarrheal losses. Potassium loss is due to its passive movement into the stool water as part of the bulk fluid flow through the small intestine, although some active secretion by the colon may also contribute. Presence of steatorrhea in this patient is unexplained; it is an uncommon occurrence in VIPomas.

The prognosis is good in childhood VIPoma. Diagnosis is often delayed despite radiological and biochemical methods. In our case, two years elapsed before diagnosis and the tumor had already metastasized to the lymph nodes. Greater clinical awareness of this syndrome would lead to earlier detection and improve the chances of successful resection and symptomatic cure.

References

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Acute pancreatitis in a child with idiopathic ulcerative colitis on long-term 5-aminosalicylic acid therapy

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Acute pancreatitis is a rare but known complication of inflammatory bowel disease in adults. In children, only a few cases with this complication have been reported. We describe a 10-year-old boy with ulcerative colitis who developed acute pancreatitis while on long-term treatment with 5-aminosalicylic acid. [Indian J Gastroenterol 2000;19:195-196]

Key words: Inflammatory bowel disease, pancreas

A acute pancreatitis is a rare but known complication of 5-aminosalicylic acid (5-ASA) therapy in adults with inflammatory bowel disease. It classically appears within the first few days or weeks after initiation of therapy. We report a 10-year-old boy with idiopathic ulcerative colitis on 5-ASA who developed a mild clinical episode of acute pancreatitis.

A 10-year-old boy was admitted to our hospital with bloody diarrhea, abdominal pain, and tenesmus of one-year duration.