Perforation in a jejunal duplication lined by ectopic gastric mucosa

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We report a 3-year-old child with jejunal duplication with localized perforation and bleeding in the adjacent normal intestine. The entire duplicated jejunal segment was lined by gastric mucosa. This was successfully treated by mucosal excision of the duplicated intestine and resection of a short segment of normal jejunum. [Indian J Gastroenterol 2004; 23: 152]

Key words: Intestinal duplication

Duplication of the intestinal tract lined entirely by ectopic gastric mucosa is a rare cause of bleeding in children. Perforation is known but is unusual in the adjacent normal jejunum.

A 3-year-old boy was admitted with complaints of severe abdominal pain with distention, occasional vomiting and several dark tarry stools. He was anemic and in shock. Abdominal distension and guarding. Per rectal examination revealed melena. Post-resuscitation X-ray abdomen showed gas under the diaphragm. After blood transfusion the child was taken up for surgery.

Laparotomy revealed a long tubular duplication of the jejunum on the mesenteric side (Fig). This was communicating distally with the normal intestine. A localized perforation was noted in the adjacent normal jejunum. Total mucosal excision of the duplicated jejunal segment with resection of a small segment of the normal jejunum bearing the perforation was done. End-to-end anastomosis was done in two layers; the mesenteric defect was closed. The specimen showed ectopic gastric mucosa in the entire length of the duplicated jejunum. The patient recovered smoothly and was discharged on the 10th day.

Intestinal duplications can occasionally be multiple and may not communicate with the adjacent normal bowel. In our case it was communicating at the distal end. The duplications lie on the mesenteric border of the intestine, often sharing a common blood supply and muscular cover. Our case had a common blood supply.

Hypertrophic gastric mucosa in the entire duplicated jejunal segment is extremely rare. The entire duplicated bowel in our case was lined by ectopic gastric mucosa.

Bleeding from ulceration in duplicated bowel or adjacent normal intestine is well known and occurs in about 20% of cases. Perforation in duplicated bowel

Fig: Long tubular single duplication of jejunum on mesenteric side (arrow)
is also known, but perforation of normal small bowel mucosa secondary to duplication of jejunum lined by heterotrophic gastric mucosa is very rare. We have come across only one such case report. In cases with small length of duplicated bowel, the normal bowel is sacrificed. In cases with long duplications, opening both ends of the abnormal structure to form a double bowel intestine is done, or the duplication is separated from normal bowel, or only mucosal excision is done. Our case had a long duplication, and to minimize bowel and blood loss only mucosal excision of the duplicated jejunum was done.

References

Good's syndrome: an unusual cause of chronic diarrhea

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A 50-year-old man presented with recurrent episodes of pulmonary infections over a period of 5 years, chronic
small bowel diarrhea and weight loss of 6 months' duration. On evaluation he was found to have a thymoma, intestinal infection with giardia, oral candidiasis, and low immunoglobulin levels. He was diagnosed to have Good's syndrome. The patient refused further management. [Indian J Gastroenterol 2004;23:152-153]

Key words: Immune deficiency, primary; thymoma

Good's syndrome (thymoma with immunodeficiency) is a rare cause of combined B and T cell immunodeficiency in adults. Clinically the patient has increased susceptibility to bacterial infections with encapsulated organisms and opportunistic viral and fungal infections. Hypogammaglobulinemia and reduced or absent B cells are the most consistent findings in these patients.

A 52-year-old man was admitted with history of small bowel diarrhea for six months and cough with expectoration for 5 days. There was history of significant weight loss over the last six months. He had had repeated episodes of pulmonary infections during the preceding 5 years, which responded to various antibiotics. On examination the patient had a BMI of 16.7, pallor and oral candidiasis. Chest examination revealed bronchial breath sounds in the left infracapsular area.

Investigations: hemoglobin 10 g/dL, leukocyte count 3400/mm³ (80% neutrophils). Blood sugar and kidney function tests were normal. Liver profile was normal except for low level of globulin. Chest X-ray showed upper mediastinal widening and left lower lobe pneumonia. The patient was treated with intravenous amoxicillin-clavulanic acid and ceftriaxone oral solution. Ultrasonography of the abdomen did not reveal any abnormal finding. Upper gastrointestinal endoscopy did not show esophageal candidiasis. Biopsy from the third part of the duodenum showed normal crypt-villus ratio, chronic inflammatory cells in the submucosa, and tropohyphocytes of Giardia lamblia. Injectable metronidazole was added to the treatment. Sputum culture grew Streptococcus pneumoniae. The patient responded to treatment and was symptom-free after 5 days. CT scan chest showed a thymoma. Serology for HIV infection (ELISA) was negative twice. Immunoglobulin levels revealed a decrease in IgA (0.8 g/L), IgM (0.4 g/L) and IgG (6 g/L) levels.

The patient was diagnosed to be suffering from Good's syndrome. He was advised resection of the thymoma, bone marrow examination and immunoglobulin replacement, which he refused.

Good's syndrome is classified as a distinct entity, though diagnostic criteria for this disorder are absent. The pathogenesis of this disorder is not known. There is evidence that the basic defect may be in the bone marrow. Patients usually present in the 4th or 5th decade of life. The immunodeficiency may precede or follow the diagnosis of thymoma.

The patient may present with asymptomatic anterior mediastinal mass on chest X-ray, compressive symptoms secondary to thymoma, or with mass in the neck.

Patients may present with infection as a result of the defects in humoral and cellular immunity. The most common infection is recurrent sinopulmonary infection secondary to encapsulated organisms, or bacterial urinary tract and skin infections. Diarrhea has been reported in almost 50% of patients with Good's syndrome. There is also a suggestion about presence of an idiopathic inflammatory diarrhea in these patients; Good's syndrome may be associated with other autoimmune conditions.

Hematological abnormalities include anemia, low white blood cell count and thrombocytopenia. Pure cell aplasia, aplastic anemia, hemolytic anemia, and pernicious anemia have been reported. Presence of monoclonal gammapathies and T cell tumors have been described.

The dominant immunological findings are hypogammaglobulinemia, few or absent B cells, abnormal CD4+:CD8+ T cell ratio, decreased CD4 T cells, and impaired T cell mitogenic responses.

The treatment of thymoma is surgical removal or debulking of the tumor. Patients with stage 3 or 4 tumors require radiotherapy and combination chemotherapy. Removal of the thymoma does not reverse the immunological abnormalities. Antibody deficiency requires immunoglobulin replacement treatment. The prognosis of Good's syndrome is poor. The principal causes of death are infections, other autoimmune diseases, or associated hematological complications.

References

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