Ménétrier Disease

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A previously healthy 16-month-old boy was hospitalized because of vomiting of 10 days' duration, fever of 4 days' duration (temperature up to 38.6°C [101.4°F]), and watery diarrhea. He also had had a maculopapular rash, which resolved the day before presentation. Family history was unremarkable.
A previously healthy 16-month-old boy was hospitalized because of vomiting of 10 days' duration, fever of 4 days' duration (temperature up to 38.6°C [101.4°F]), and watery diarrhea. He also had had a maculopapular rash, which resolved the day before presentation. Family history was unremarkable.

Physical examination, including abdominal examination, was normal. No rash was noted. Weight was 12.5 kg (90th percentile), and development was appropriate for age. On the third hospital day, significant pitting edema (mainly on the trunk) and periorbital swelling developed; the albumin level was 1.8 g/dL, and total protein level was 3.5 g/dL. Urinalysis results, liver enzyme levels, prothrombin time, partial thromboplastin time, urea nitrogen level, and creatinine level were normal. A protein-losing enteropathy/gastropathy was suspected. Gastroscopy revealed hypertrophic gastric folds and severe gastritis (A). Histopathological examination of the gastric mucosa showed foveolar hyperplasia and inflammatory changes (B), consistent with Ménétrier disease, and Cytomegalovirus (CMV) inclusion bodies (C, arrows).

Ménétrier disease in children is a rare, self-limited disorder that is characterized by marked protein-losing gastropathy associated with enlarged and thickened gastric folds.\textsuperscript{1-3} Abnormal regulation of gastric epithelial growth, probably triggered by an infectious agent (CMV or \textit{Helicobacter pylori}), may be a cause of this disorder.\textsuperscript{4-9} In this case, enzyme immunoassay testing was positive for CMV-specific IgM antibody and urine culture was positive for CMV antigen. The clinical, radiological, and histological features of Ménétrier disease in children resemble those of the adult form. However, there are important differences. Generalized edema occurs in fewer than 25\% of adult cases but is the most common finding in children. In addition, the clinical course is usually unfavorable in adults; many require partial or total gastrectomy for persistent symptoms.
The prognosis for children, on the other hand, is excellent. This patient was treated with omeprazole; within 3 weeks, his symptoms had resolved and the serum albumin level had normalized.

References:

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