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Giant Rugal Hyperplasia of the Stomach: Menetrier's Disease

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Menetrier's disease is an uncommon clinical entity, marked by protein losing enteropathy and hypertrophic gastropathy that has predilection for the proximal portion of stomach(l). The entity in the pediatric age group is known as "Pediatric Hypertrophic Gastropathy" which signifies

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different etiopathogenesis and course in children(2).

Case Report

A 16-year-old black girl was admitted with a three week history of epigastric pain, recurrent vomiting and swelling of both lower extremities. During this period, she was afebrile but anorexic and suffered weight loss of 6 lbs. She did not have smoky urine, oliguria, dyspnea on exertion or other physical complaints. Physical examination revealed stable vital signs, pale nonicteric sclera, mild periorbital edema and edema of both lower extremities. The remaining systemic examination was unremarkable.

The total leucocyte count was 10500/cu mm with a DLC of P:50, L:42, M:5 and E:3. The absolute eosinophil count was 160/cu mm. Sickle cell preparation was negative. Urine analysis, electrolytes and culture were unremarkable. The 24 hour urine albumin was within normal limits. Serum electrolytes were Na^+ 137 meq/L, K^+ 3.6 meq/L, Cl-105 meq/L and HCO_3, 26 meq/L. Blood urea nitrogen was 22 mg/dl. Blood sugar and serum creatinine were 119 mg/dl and 1.1 mg/dl, respectively. Total serum proteins were 3.8 g/dl with albumin 2.1 g/dl. Serum cholesterol was 168 mg/dl. Hepatic enzyme values were in normal range with non-reactive hepatic serology. Rheumatoid factor and anti-nuclear antibody were negative. Thereafter, an upper gastrointestinal series, abdominal scan and endoscopy were done as a part of the workup (*Figs. 1 & 2).* With diagnosis of Menetrier's disease, the patient was treated with ranitidine. She quickly became symptom free and had a slight rise in serum proteins within a ten days period. She was followed as an

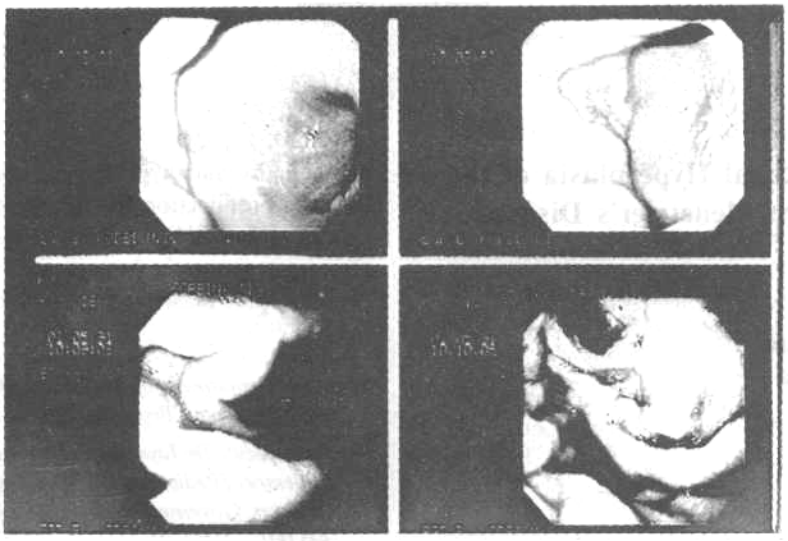

Fig. 1. Gastroscopy shows thick gastric folds in the body and fundus of stomach with virtually normal antrum.

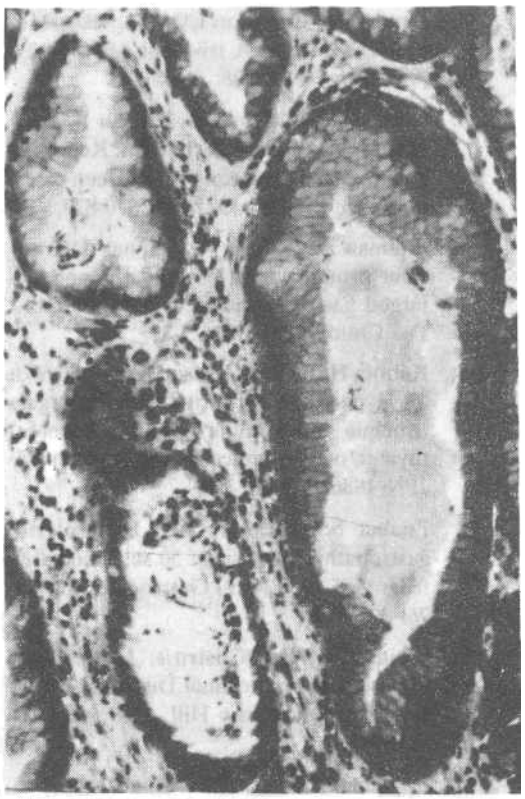

Fig. 2. Gastric mucosal biopsy shows intestinalization of the gastric mucosa with microcystic changes consistent with Menetrier's disease.

outpatient and edema completely resolved over the next three weeks.

Discussion

Menetrier's disease is a rare clinical entity and the exact data regarding the incidence in children is not available due to nebulous definition. Menetrier's disease is also known as "Trivalent Gastropathy" which signifies a triad of thick gastric folds, typical mucosal histology and hypoalbuminemia(3). In contrast to adults, the disease has a benign course in children with spontaneous resolution over a few weeks(l).

The majority of cases are idiopathic in nature but several causes which can invoke reversible disease process in children, are allergy(4), infection specifically with cytomegalovirus(5) and immune phenomenon^). Cytomegalovirus infection in children is linked with hypertrophic gastropathy mainly because of presence of cytomegalic inclusions in gastric biopsy, biopsy culture positive for CMV and cytomegalouria(2,5). Herpes virus and *Campylobacter pylori (Helicobacter)* are suggested as other possible infective agents in pathogenesis. The allergic hypothesis is based on peripheral eosinophila, eosinophilic mucosal infiltration and self limiting course of protein losing enteropathy(4).

The clinical presentations of Menetrier's disease are recurrent abdominal pain, anorexia, vomiting and weight loss accompanied by manifestations of hypoalbuminemia, mainly edema and frequent infections. The hypoalfcuminemia is the hallmark and consistent feature of the entity(3). The other biochemical variables are serum gastrin and gastric acid secretion study which are important to subclassify other hypertrophic gastropathics. Gastric acidity is usually low(7) possibly due to decreased parietal cell mass, neutralization of the acid by mucus and exuded plasma proteins and back diffusion of acid. In the roentgenographic evaluation, upper gastrointestinal series and abdominal scan are helpful in supporting the diagnosis but are not specific. Barium study shows thick gastric folds, strikingly sluggish peristalsis and delayed emptying. In many cases, apparently thick gastric folds, seen on barium study, are not found thickened on endoscopy(8). Gastro'scopy shows thick gastric folds which are most promi-

nent in the fundus and body of stomach along the greater curvature with virtually normal antral region. Typical histological features on gastric biopsy include elongation and tortuosity of pits, often associated with prominent cystic dilatations(9) with marked foveolar hyperplasia. There is an accompanying extensive reduction in numbers of parietal and chief cells with replacement by mucus glands. The differential diagnosis of asymptomatic child with thick gastric folds include idiopathic hypertrophic gastropathy, hypertrophic hypersecretory gastropathy, Zollinger-Ellison syndrome, Cronkhite-Canada syndrome and diffuse infiltrating neoplasm of stomach.

Menetrier's disease in children is us ually a self limiting disorder. H2 receptor blockers like cimetidine and ranitidine, tight en gastric cell junctions and thereby limit paracellular protein loss(10). Parenteral nutrition and surgical treatment are rarely indicated in children but often necessary in adults.

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