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Pathological Case of the Month

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A 17-YEAR-OLD African American girl was seen for severe anemia. She had mild anemia 1 year previously and increased fatigue and shortness of breath for 3 months prior to evaluation. Her history was notable for insulin-dependent diabetes mellitus (IDDM) since age 6 years, Graves disease with hyperthyroidism since age 13 years, and paranoid schizophrenia since age 14 years. Her maternal grandmother and a maternal aunt had vitamin B_{12} deficiency, and a brother and maternal aunt had IDDM. Two weeks prior to presentation, our patient developed peripheral edema and cold extremities, consistent with florid congestive heart failure; on examination she had pallor, peripheral edema, and a cardiac gallop with evidence of moderate pulmonary edema. She was paranoid and delusional and had received chlorpromazine hydrochloride, insulin, and sertraline hydrochloride on a regular basis. Peripheral blood (**Figure 1**) showed profound anisocytosis with macro-ovalocytes, numerous microcytes, polychromatophilic cells, basophilic stippling, and nucleated red blood cells. There were scattered hypersegmented neutrophils. Bone marrow aspiration biopsy (**Figure 2**) revealed hypercellular marrow with profound erythroid hyperplasia, left shift of the myeloid precursors, and marked megaloblastic changes. Her vitamin B_{12} level was low, and lactic dehydrogenase levels were elevated. Hemoglobin electrophoresis was consistent with β-thalassemia trait. A chest radiograph showed mild cardiomegaly and pulmonary edema. The endoscopic appearance of the gastric mucosa (**Figure 3**) revealed marked atrophy with absent rugal folds and a diffuse reticular pattern of the mucosa. Gastric biopsy specimens (**Figure 4**) revealed chronic atrophic gastritis with intestinal metaplasia.

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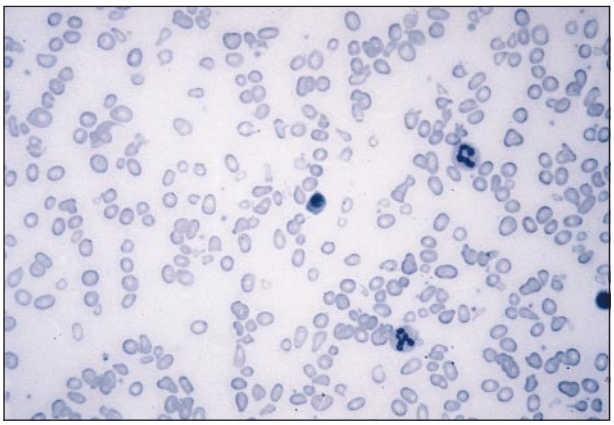
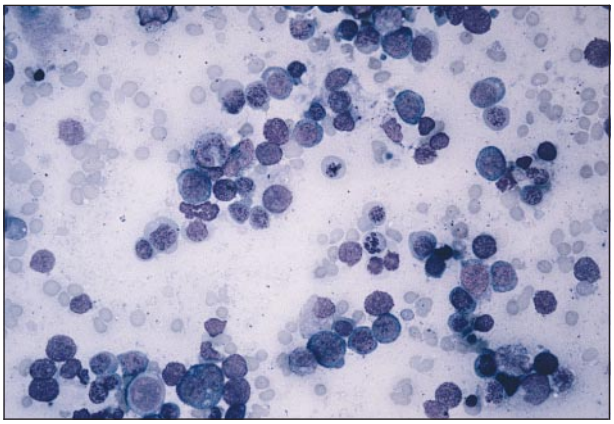


Figure 1.



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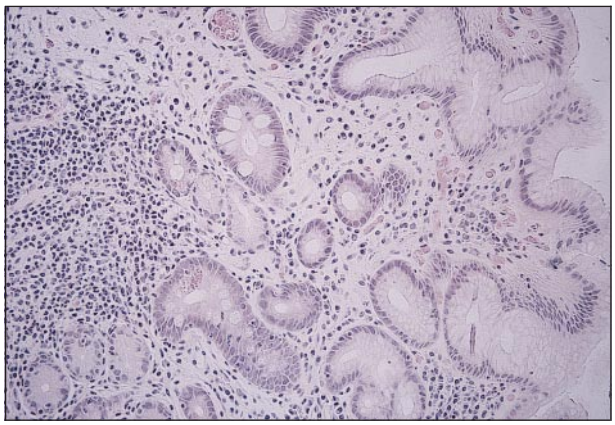

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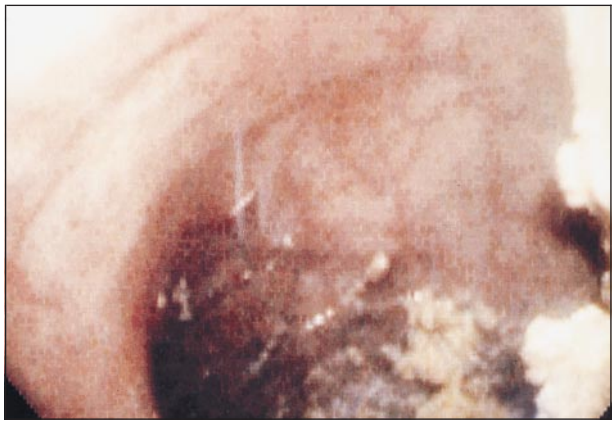

Figure 4.

Diagnosis and Discussion

Pernicious Anemia and Gastric Atrophy in an Adolescent Female With Multiorgan Problems

Figure 1. *Peripheral blood smear showing macrocytosis and hypersegmented neutrophils.*

Figure 2. *Bone marrow biopsy specimen showing typical megaloblastic changes and hypersegmented neutrophils.*

Figure 3. *Gastric mucosal biopsy specimen showing atrophic gastritis.*

Figure 4. *Endoscopic appearance of gastric atrophy. Note the large amount of retained food residue after overnight fasting, consistent with gastroparesis.*

Our patient received a packed red blood cell transfusion and a short course of diuretics with prompt improvement of congestive heart failure. An esophagogastroduodenoscopy was performed after correction of her severe anemia, achieving control of her psychosis. On esophagogastroduodenoscopy, a large amount of retained food was seen after 6 hours of fasting, suggestive of gastroparesis possibly related to her IDDM.

The mucosa was markedly thinned with areas of fibrosis in which glandular elements were absent. Lymphocytes and plasma cells infiltrated the lamina propria. There were focal areas in which goblet cells were prominent among the glandular and surface epithelium, and scattered Paneth cells were present. However, parietal cells were not seen. No *Helicobacter pylori* microorganisms were noted. An upper gastrointestinal tract barium study with small bowel follow-through was unremarkable except for mild gastroesophageal reflux and delayed gastric emptying. The appearance of the terminal ileum was unremarkable.

Recommendations were made to check antiparietal cell and anti-intrinsic factor antibody titers, serum gastrin levels, and to perform a Schilling test. Supplementation with parenteral vitamin B_{12} was started with resolution of her anemia. Her type A atrophic gastritis with pernicious anemia is a predisposing factor for development of gastric adenocarcinoma and gastric carcinoid tumors. Furthermore, because of her gastroparesis with IDDM, long-term follow-up with a gastroenterologist was recommended. It is routine teaching in pediatrics to consider one disease process as a unifying cause of multiple problems. In appropriate circumstances, multiple system problems and diseases must be sought and cared for. This case displays a wide spectrum of autoimmune manifestations, including IDDM, thyrotoxicosis, and pernicious anemia with gastric atrophy and response to vitamin B_{12} supplementation.

Accepted for publication August 15, 2000.

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Submissions

The Editors welcome contributions to *Pathological Case of the Month, Picture of the Month*, and *Radiological Case of the Month*. Those who wish to contribute should send their manuscripts to Dr Gilbert-Barness (*Pathological Case of the Month*), Department of Pathology, Tampa General Hospital, University of South Florida, Davis Island, Tampa, FL 33606; Dr Tunnessen (*Picture of the Month*), The American Board of Pediatrics, 111 Silver Cedar Ct, Chapel Hill, NC 27514-1651; or Dr Wood (*Radiological Case of the Month*), KAM 211, USC-HSC, 1975 Zonal Ave, Los Angeles, CA 90089-9024. Articles and photographs accepted for publication will bear the contributor's name. There is no charge for reproduction and printing of color illustrations.