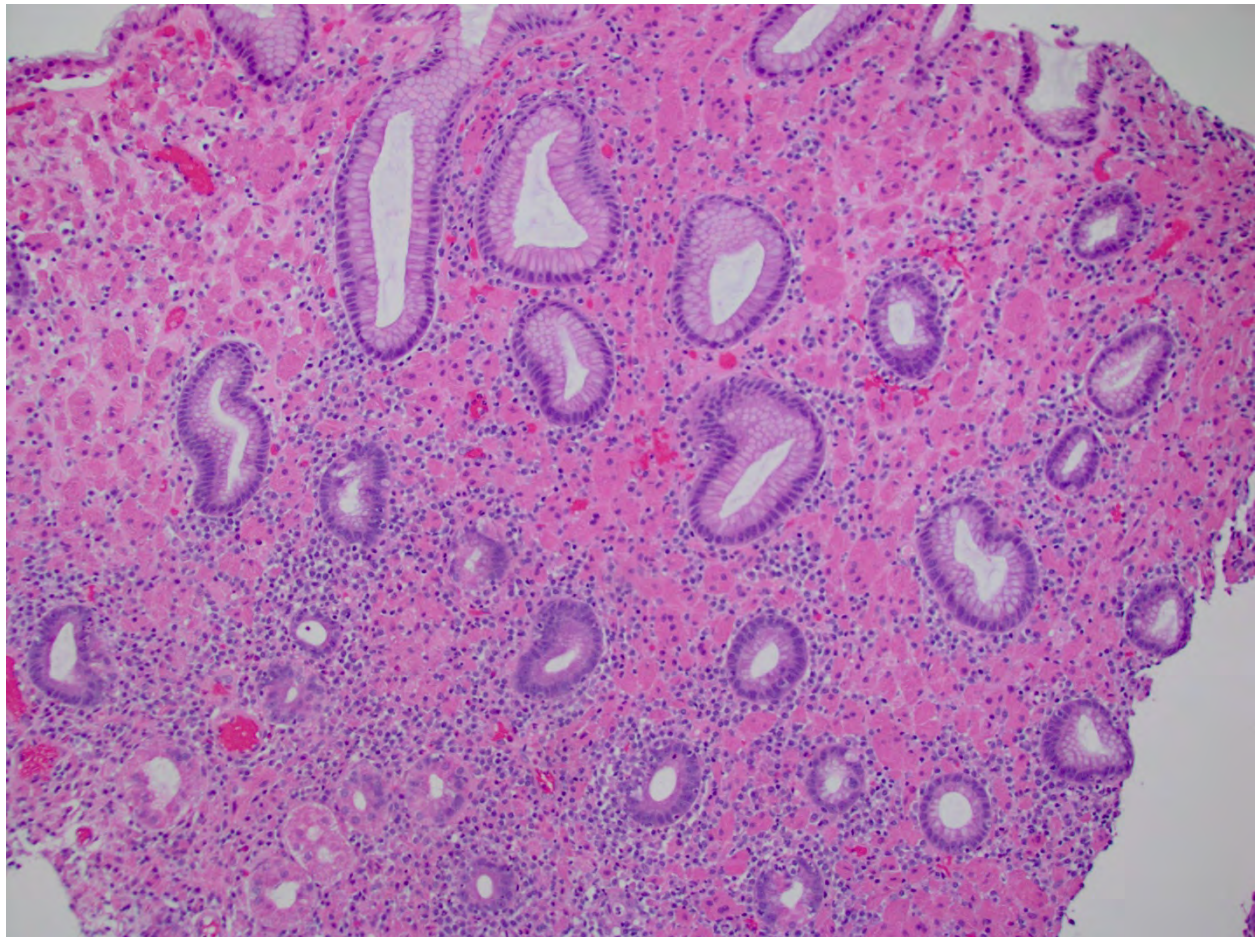
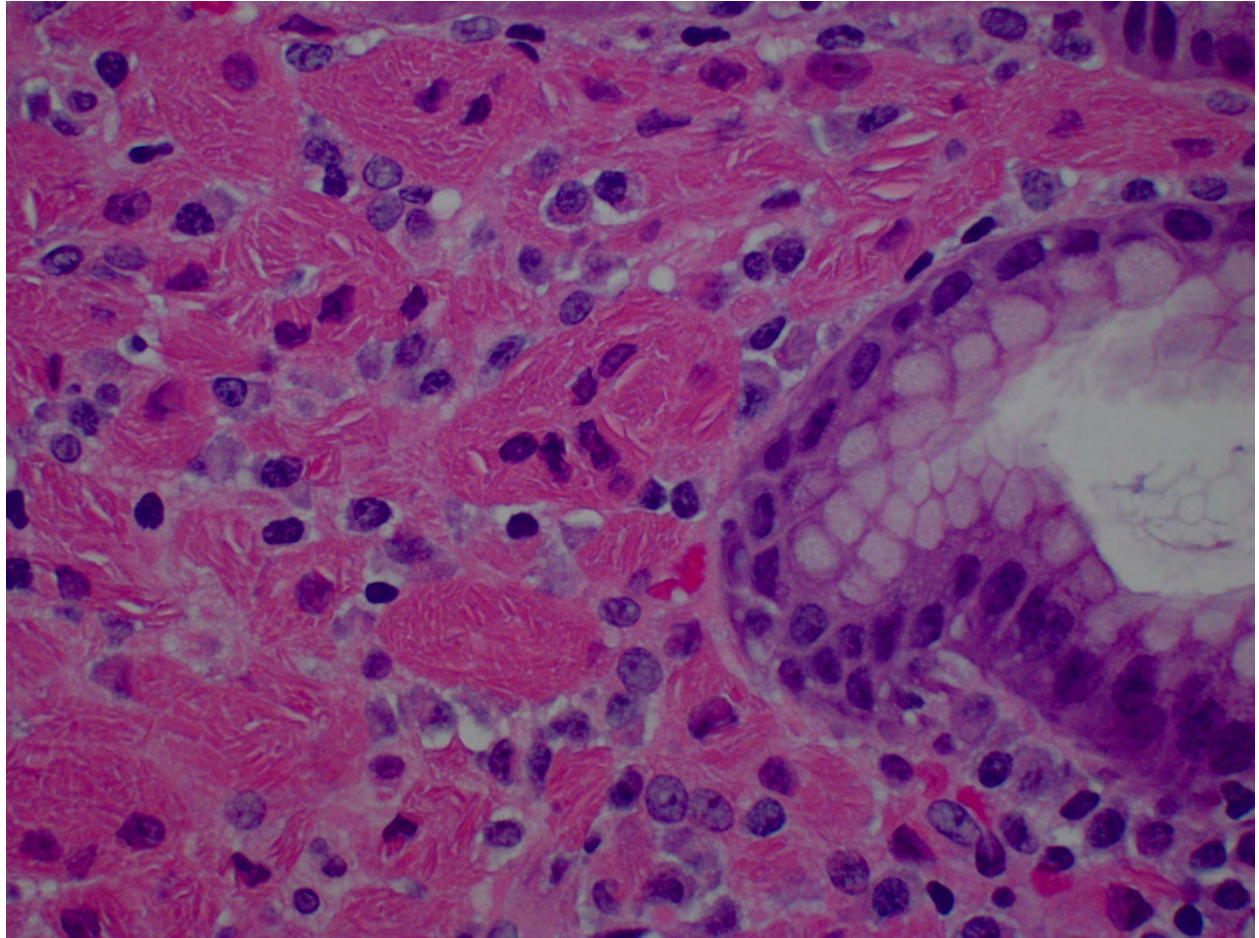
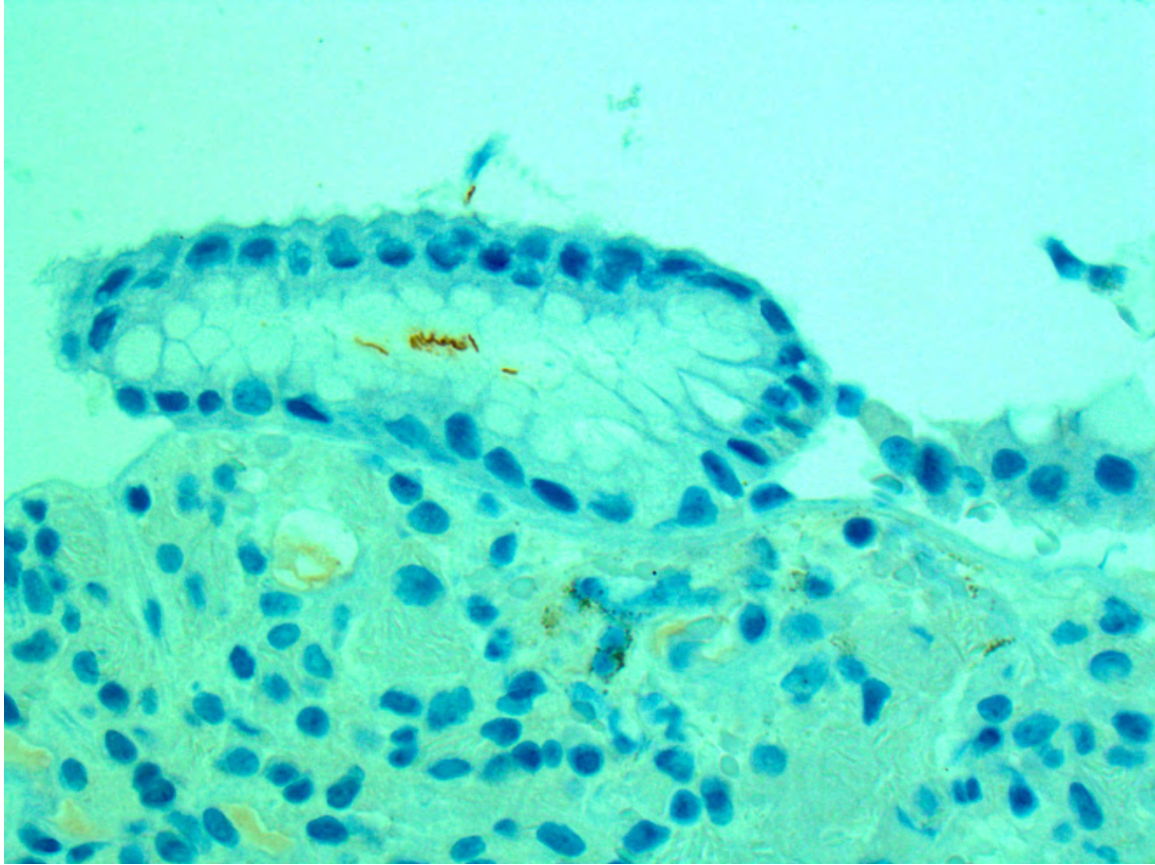


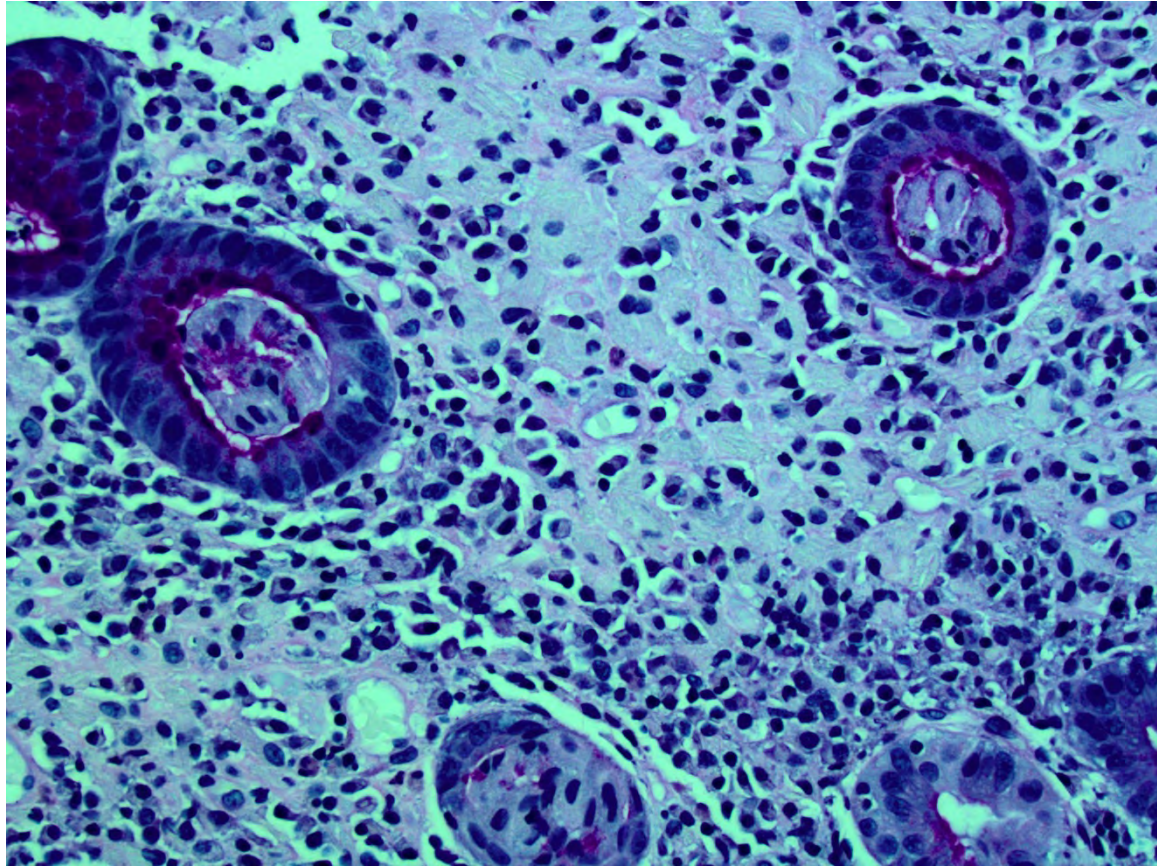
Case: The patient is a 73 year old woman with vague complaints of dyspepsia and abdominal pain. Upper endoscopy showed features of gastritis and a nodular lesion in the body of the stomach. The patient has no significant previous medical history.



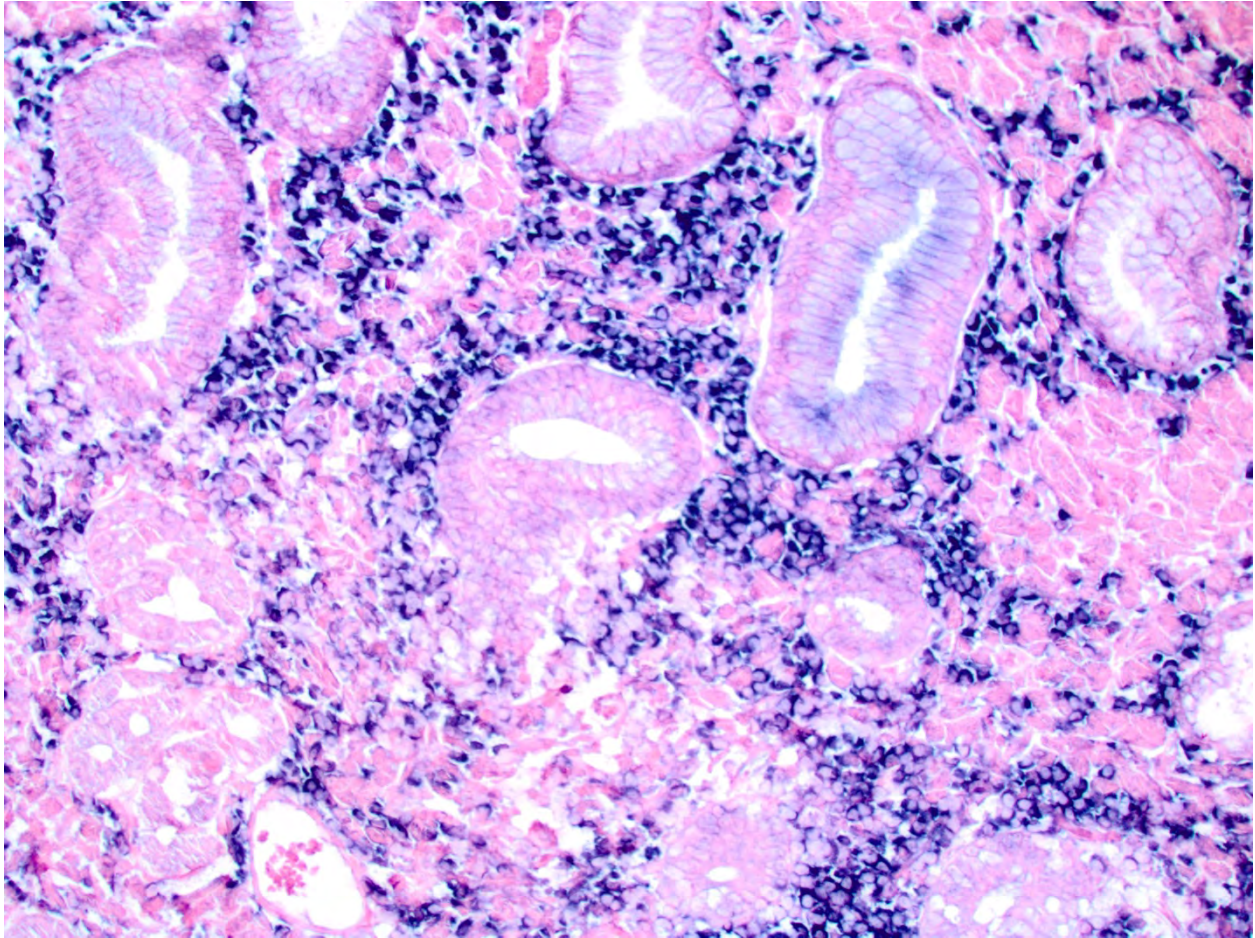




H. pylori immunostain



Alcian blue/PAS



Kappa in situ hybridization

What is your diagnosis?

- A. Russell body gastritis
- B. Mycobacterium avium complex infection
- C. Crystal storing histiocytosis
- D. Gastric adenocarcinoma, diffuse type
- E. Gaucher's disease

Answer and Discussion:

C. Crystal storing histiocytosis

Comment: The photomicrographs demonstrate gastric mucosa with chronic gastritis and *Helicobacter* organisms. The lamina propria contains numerous plasma cells and histiocyte-like cells containing eosinophilic material and apparent crystals. Immunohistochemical staining showed that the histiocyte-

like cells are negative for S-100, smooth muscle actin, desmin, PAS, PAS with diastase, and CAM5.2. Staining for acid fast bacilli was also negative. In-situ hybridization for kappa and lambda light chains showed that the plasma cells are kappa-restricted.

Choice A is incorrect. Russell body gastritis is a recently recognized pseudotumor lesion of the gastric mucosa that is often, but not always, associated with *H. pylori* infection. In Russell body gastritis, the lamina propria is infiltrated by numerous plasma cells which contain large, spherical eosinophilic hyaline globules which push the nucleus to the periphery of the cell. Cytoplasmic crystals are absent.

Choice B is incorrect. *Mycobacterium avium* complex (MAC) infection may result in accumulation of large numbers of histiocytes, most often within the lamina propria of the duodenum. The histiocytes in MAC infection usually appear pale pink and lack the cytoplasmic crystalline structures seen in this case. They contain acid fast bacteria readily identified using special stains.

Choice D is incorrect. Diffuse type gastric adenocarcinoma should be considered in the differential diagnosis in this case. The histiocyte-like cells, however, show no cytologic features of malignancy, and are cytokeratin negative.

Choice E is incorrect. Gaucher's disease is lysosomal storage disease in which glucocerebroside accumulates in histiocytes and other cells. The most common type of the disease may affect adults, but the patient's age makes this diagnosis unlikely. The gastrointestinal tract may be affected. Gaucher cells are histiocytes characterized by abundant finely granular or fibrillary appearing cytoplasm. The material within the cytoplasm is PAS-positive. Crystals are not present.

The correct answer is C. Crystal-storing histiocytosis is a rare disorder, that has been associated with hematopoietic neoplasms, such as multiple myeloma, plasmacytoma, and lymphoplasmacytic lymphoma. It has also been described in association with non-neoplastic plasma cell proliferations, including monoclonal gammopathy of undetermined significance (MGUS). It is presumed to represent accumulation of secreted paraproteins or immunoglobulins within histiocytes and plasma cells which aggregates in crystals. Crystal-storing histiocytosis has been reported in multiple tissues, and may in some cases be restricted to involvement of a single organ. Isolated crystal-storing histiocytosis has been reported in the GI tract, kidneys, spleen, lymph nodes, skin, thyroid, and lungs. Two cases with gastric involvement have been reported; in one the patient had a systemic clonal lymphoproliferative disorder, while in the other the disease was limited to the stomach and was associated with *H. pylori* infection. Since the plasma cell population within the stomach in this case is kappa restricted, this may represent crystal-storing histiocytosis associated with an unsampled MALT lymphoma arising in a background of *H. pylori* gastritis.

References:

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- 2) Joo, M, et al. Localized gastric crystal storing histiocytosis. *Histopathology* 2007;51:114-119.
- 3) Llobet M, et al. Massive crystal-storing histiocytosis associated with low-grade malignant B-cell lymphoma of MALT-type of the parotid gland. *Diagn Cytopathol* 1997;17:148-152.

Submitted by: Dr. Parag Patel, Miraca, Phoenix Arizona