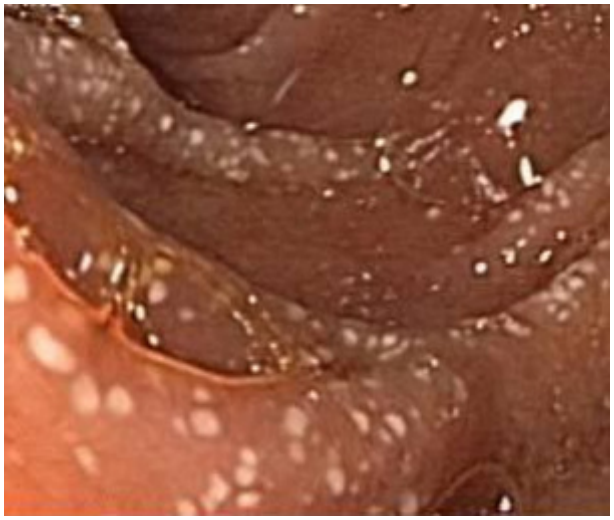
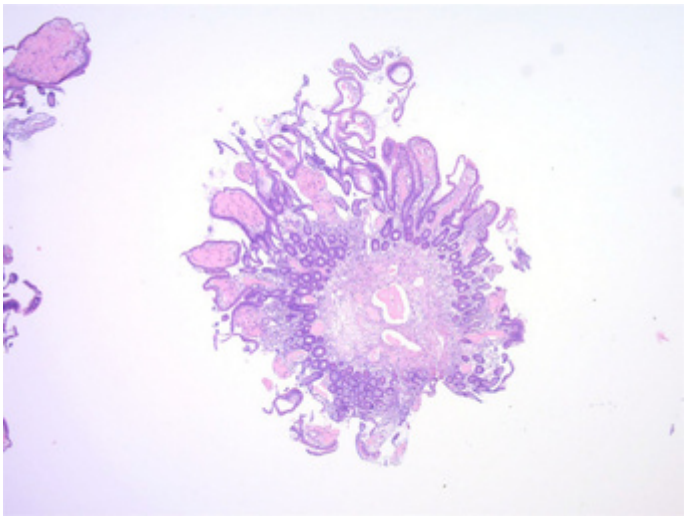
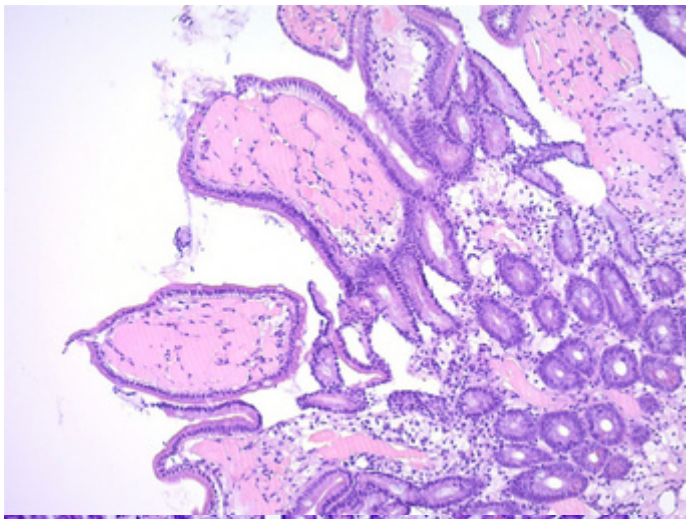
February 2018

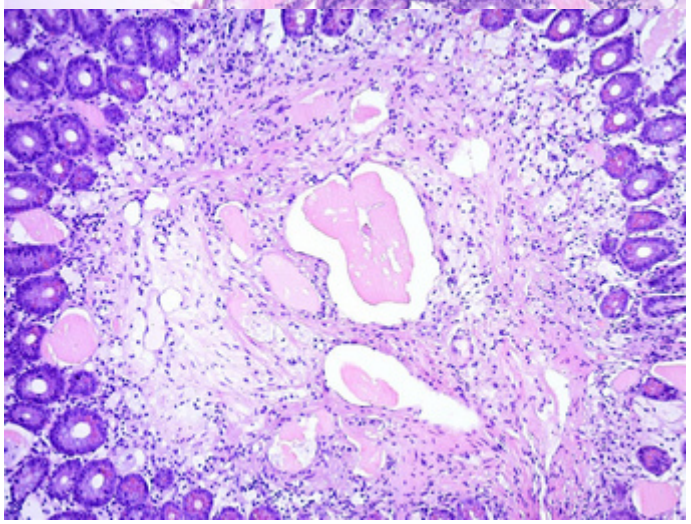
Duodenal biopsy in a 63-year-old female with night sweats, watery diarrhea and newly acquired shin hyperpigmentation.

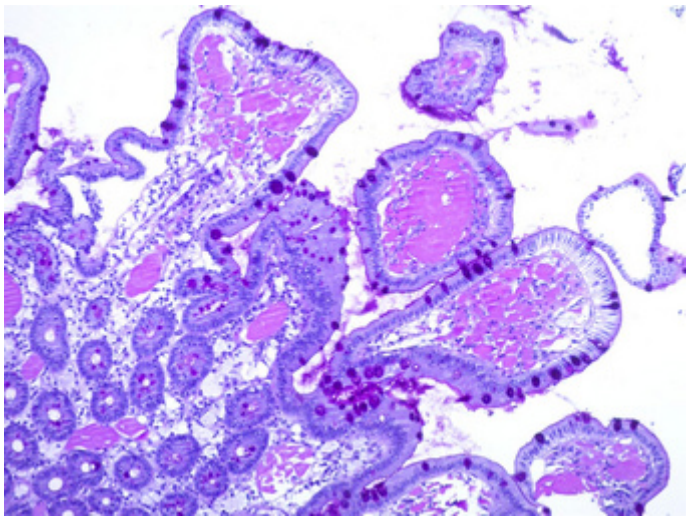
What is your diagnosis?











Diagnosis:

Lymphangiectasiae due to Waldenström macroglobulinemia.

Comment:

A 63-year-old female patient presented with a three month history of weight loss (20 kg), headache, watery diarrhea, nausea, generalized weakness, night sweats, newly acquired shin hyperpigmentation and splenomegaly. Upper endoscopy revealed whitish dots in the duodenal mucosa (Panel A). Histology from the duodenal bioptic material shows intestinal mucosa with preserved villi and distended lamina propria with numerous dilated lymphatic vessels (Panels B and C) that are also present in the submucosa (Panel D). The vessels contain eosinophilic, PAS-positive content (Panel E).

A bone marrow smear and bone marrow biopsy were performed, followed by flow cytometry, all revealing bone marrow infiltration by lymphoplasmacytoid lymphoma. Immunoelectrophoresis showed monoclonal IgM kappa.

Lymphoplasmacytic lymphoma is a B-cell neoplasm that usually presents in the sixth or seventh decade of life. More than 90% of cases are associated with a MYD88L265P mutation. A substantial fraction of the tumor cells undergo terminal differentiation to plasma cells, which commonly secrete large quantities of monoclonal IgM. When abundant enough, IgM can cause a hyperviscosity syndrome known as Waldenström macroglobulinemia. Intestinal symptoms of the hyperviscosity syndrome include mild to severe diarrhea, sometimes with malabsorption, occasionally presenting as the main complaint that ultimately leads to the correct diagnosis. The pathomorphological substrate are lymphangiectasiae, which appear as whitish specks on endoscopy. The most striking histologic finding is the deposition of acellular, eosinophilic PAS positive material within lymphatic channels in the tips of villi and in the base of the mucosa, corresponding to IgM.

Amyloidosis, primary lymphangiectasia, Whipple's disease and Mycobacterium avium-intracellulare should be excluded in the differential diagnosis, which can readily be done by the use of special stains and PCR for infectious microorganisms.

For further reading:

- Waldenström J. Incipient myelomatosis or “essential” hyperglobulinemia with fibrinogenopenia: A new syndrome? Acta Med Scand. 1944; 117: 216-47.
- Cooke RE, Kalnins RM, Ho WK. Waldenström macroglobulinaemia and intestinal lymphangiectasia. Br J Haematol. 2014; 167: 292.
- El-Ayoubi A, Wang JQ, Hein N, Talaulikar D. Role of plasma cells in Waldenström macroglobulinaemia. Pathology. 2017; 49: 337-45.

› Abeykoon JP, Paludo J, King RL, et al. MYD88 mutation status does not impact overall survival in Waldenström macroglobulinemia. Am J Hematol. 2017; doi: 10.1002/ajh.24955 [Epub ahead of print].

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