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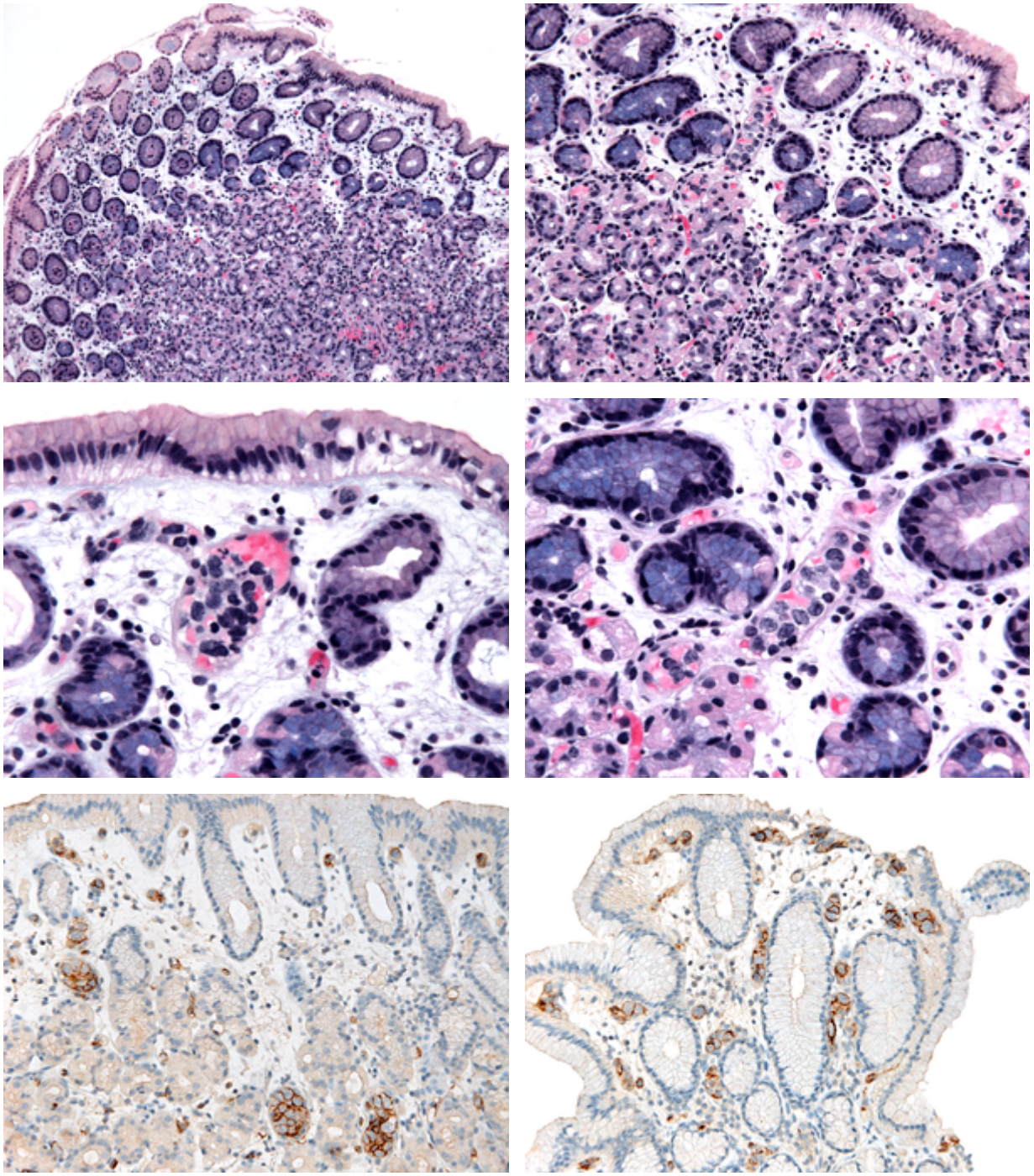
Gastric biopsies from a 79-year-old woman with a 2 month history of weight loss and fever.

What is your diagnosis?



Diagnosis:

Intravascular B-cell lymphoma.

Comment:

A 79-year-old woman was admitted for diagnostic work-up because of unexplained weight loss and fever. Gastroscopy didn't show any abnormalities. Nevertheless, random biopsies were obtained from antrum and corpus. Microscopically, the mucosa appeared almost normal (Panel A). However, careful examination showed

atypical large cells in the capillaries in the lamina propria (Panels B, C, D). Immunohistochemically, they were positive for CD20 (Panels E and F) and negative for S100 and cytokeratin. The patient died 2 weeks later due to pneumonia and sepsis. At autopsy, massive involvement of small blood vessels by lymphoma cells was found in the brain, gastrointestinal (GI) mucosa, lungs, liver and kidneys, confirming the diagnosis of intravascular lymphoma.

Intravascular B-cell lymphoma (IVBCL) is a rare type of extranodal large B-cell lymphoma characterized by the selective growth of lymphoma cells within the lumina of the small- to medium-sized blood vessels, without an obvious extravascular tumor mass or detectable circulating lymphoma cells in the peripheral blood. Different organs and tissues may be involved, including the central nervous system, skin, lung, kidney and GI tract. It rarely affects the liver, spleen and lymph nodes. Clinical presentation is highly variable and non-specific due to occlusion of small vessels and capillaries in different organ systems. The most common clinical manifestations include central nervous system presentation, cutaneous lesions, fever and hemophagocytic syndrome. IVBCL is an aggressive lymphoma with a poor prognosis. The diagnosis is often delayed due to varied clinical presentation. Biopsies are mandatory for the accurate diagnosis, showing the classic appearance of large malignant lymphoid cells filling small vascular lumina.

IVBCL is mostly diagnosed from skin or bone marrow biopsies. Studies of autopsy cases have shown involvement of the GI mucosa in the majority of patients. Few cases of IVBCL diagnosed from GI endoscopic biopsies have been reported, indicating that a correct diagnosis can be achieved from random GI biopsies. It is therefore important to be aware of this entity and to recognize the subtle, yet diagnostic histologic appearance of IVBCL in GI biopsies.

For further reading:

- Ponzoni M, Ferreri AJ. Intravascular large B cell lymphoma: widespread but not everywhere. Acta Haematol. 2014; 131:16-7.
- Shimada K, Kinoshita T, Naoe T, Nakamura S. Presentation and management of intravascular large B-cell lymphoma. Lancet Oncol 2009; 10: 895-902.
- Shimoyama Y, Sugimoto K, Kotake M, Uehara D, Kamide Y, Kuribayashi S, Kawamura O, Kusano M, Handa H, Hirato J, Yamada M. Two cases of intravascular lymphoma diagnosed by gastrointestinal endoscopic biopsy. Intern Med. 2015; 54: 3145-9.

Presented by:

Dr. Nina Zidar and Dr. Anton Cerar, Ljubljana, Slovenia.