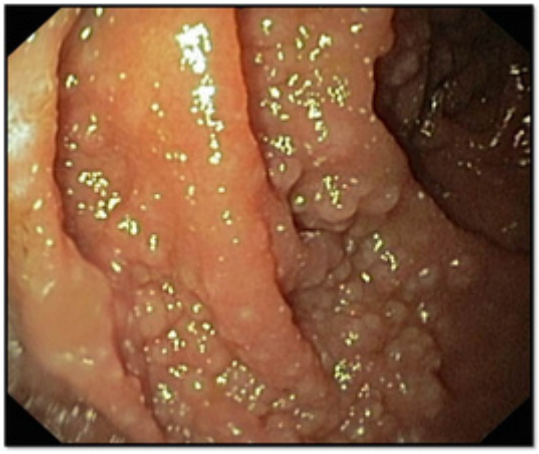
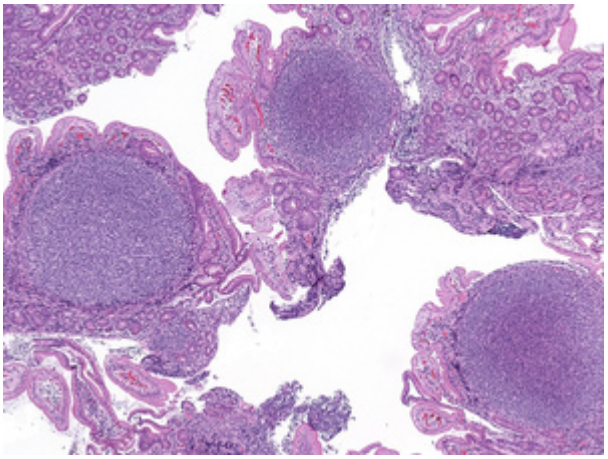
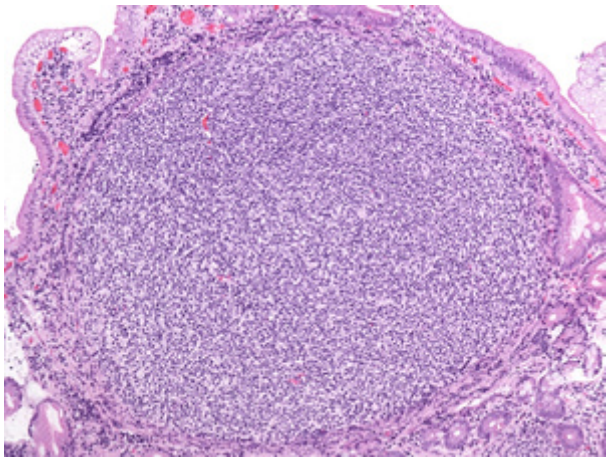
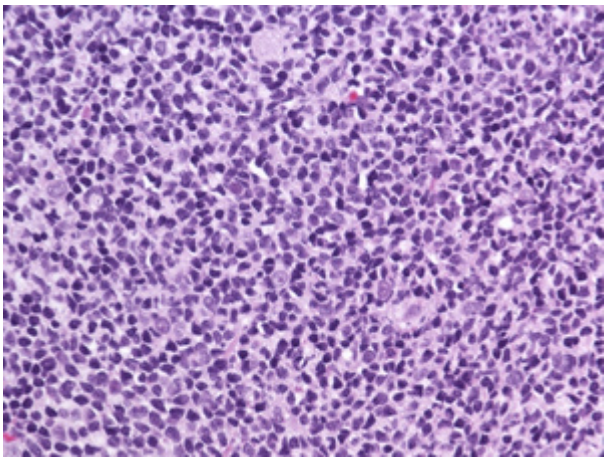
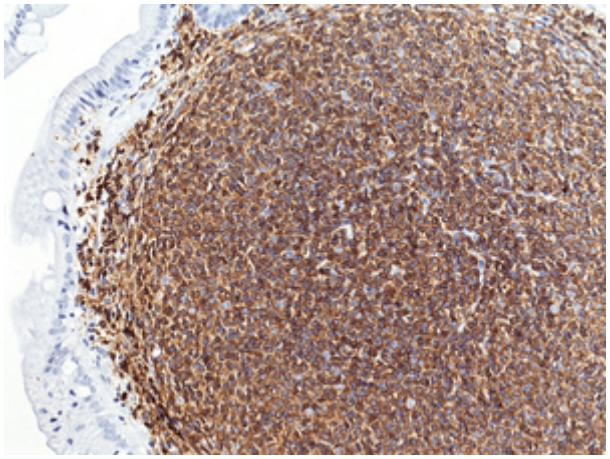
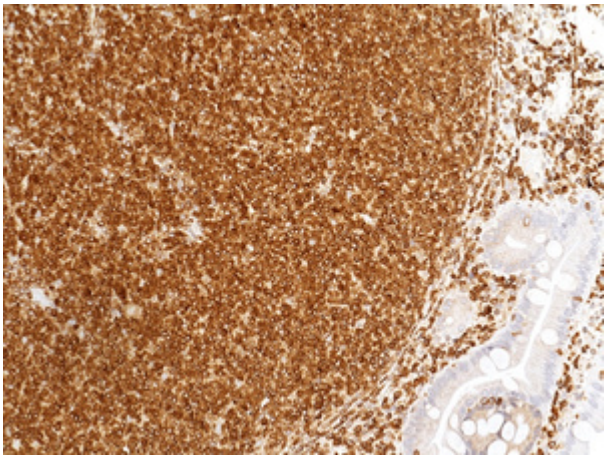
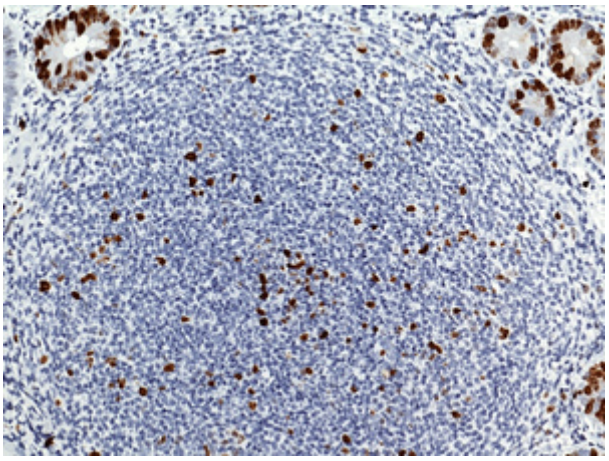
February 2016

"Granulomatous changes" in the duodenum of 37-year-old female.

What is your diagnosis?



Diagnosis:

Primary intestinal follicular lymphoma.

Comment:

A 37-year-old female presented with unspecific epigastric discomfort. The endoscopic evaluation of her upper gastrointestinal showed multiple confluent glassy polyps in the second part of the duodenum (Panel A) and biopsies were taken. Upon histology, uniform neoplastic lymphoid follicles are present in the mucosa, extending in the submucosa. They are composed of a monotonous population of small centrocyte-like cells with slightly irregular nuclei and rare large centroblast-like cells with one or more basophilic nucleoli (Panels B-D). Immunohistochemistry reveals expression for B-cell associated antigen CD20 (Panel E), BCL2 (Panel F), CD10 and BCL6. Cyclin D1 and CD5 are negative. Ki67/MIB1 staining is positive in 15% of the neoplastic cells (Panel G).

Primary intestinal follicular lymphoma (PIFL) is the second most common lymphoma of the small bowel, after diffuse large B-cell lymphoma, but is rarely seen in other locations of the gastrointestinal tract. The disease occurs preferably in young middle-age women, and the most common presenting symptom is abdominal pain. Endoscopically, PILF can have several different appearances, described by Tari et al. as whitish granules, multiple small nodules, fold swelling and thickening, mass-forming, ulcer with irregular margin or just rough mucosa. As in our case, the most frequent endoscopic appearance is as whitish granules, often as an incidental finding.

Morphology, immunophenotype and genetic features are similar to those of nodal follicular lymphoma. Thus, PILFs demonstrate a monotonous pattern of neoplastic follicles, with compressed or absent mantle zones, and typically non-polarized germinal centers. Upon high magnification, germinal center B-cells, typically both centrocytes and centroblasts are seen, randomly distributed and usually without admixed tingible body macrophages. The immunophenotype reveals positivity for B-cell associated antigens (CD19, CD20, CD22), BCL2, BCL6 and CD10, with characteristically low proliferative rate (Ki67/MIB1 usually <20%).

The main differential diagnoses comprise other malignant (low grade) lymphomas, and also follicular hyperplasia. In follicular hyperplasia follicles vary in size and shape, with present mantle zone and polarized germinal centers. Tingible body macrophages and centroblasts are common. In contrast to follicular lymphoma, reactive follicular hyperplasia lacks expression of BCL2, and the Ki67/MIB1 proliferative rate is high (usually >50%).

Regarding treatment and prognosis, most patients with PILF have localized disease. The course is often indolent, with excellent survival, even without treatment. "Watch and wait" seems to be the most sensible strategy.

For further reading:

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