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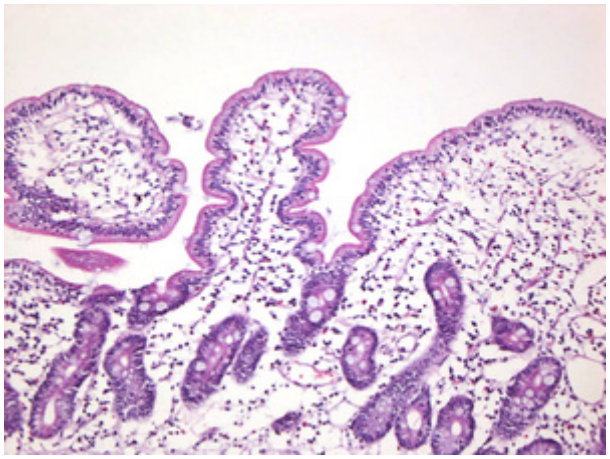
Biopsies of the descending duodenum from a 23-year-old female. Clinical information: “confirmation of celiac disease!”

What is your diagnosis?



Diagnosis

Pseudo-celiac disease in jejunal stenosis.

Microscopic findings: Normal cell count in the tunica propria with slightly elevated number of eosinophilic granulocytes. Elevated number of intraepithelial lymphocytes (27 lymphocytes per 100 epithelial cells). Flattened mucosal surface with mild atrophy (shortening) of villi.

Diagnosis: A diagnosis of “potentially” celiac disease was made since 40 intraepithelial lymphocytes per 100 epithelial cells were not reached. In addition, the cell count in the tunica propria was not increased. On the other hand the surface of mucosa was definitively flattened. The clinical colleagues were asked to perform additional serological test to confirm or exclude the diagnosis of celiac disease.

Follow-up: After three weeks we got a phone call by the clinical collegue stating that celiac disease serology was negative but the patient suffered from familial adenomatous polyposis and had jejunal adenocarcinoma with luminal stenosis. In the meantime, the patient had been operated upon and a follow-up endoscopy of the duodenum showed completely normal mucosa without any signs of inflammation and atrophy, endososcopically and histologically. The number of intraepithelial lymphocytes had dropped down to 3 per 100 epithelial cells.
The operation specimen showed a pT2, N0 (0/25), M0, L0, V0, Pn0, R0 well differentiated adenocarcinoma. The patient is well now three years after the operation and an additional proctocolectomy with J-pouch anastomosis.

Comment

A diagnosis of celiac disease should always be made with care. In cases with atypical or incomplete histology, e.g. when some features, such as villous atrophy or increased number of intraepithelial lymphocytes, are missing or only poorly developed, it is recommended to ask for results of serological testing. This is of eminent importance, particularly as guidelines are available that accept as few as 20 lymphocytes per 100 epithelial cells (instead of 40 lymphocytes) as being diagnostic for the disease. Differential diagnosis of duodenal lymphocytosis include Helicobacter induced duodenitis, drug induced lesions due to ASA or NSAID

medication, autoimmune duodenitis, and stenosis by what ever reason in deeper parts of the small bowel. On the other small bowel cancer has rarely been described to occur in patients with celiac disease and should therefore always be excluded if affected patients present with unusual gastrointestinal symptoms.

For further reading

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