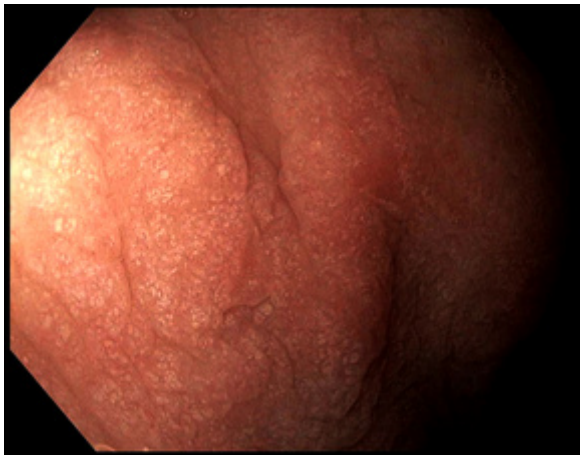
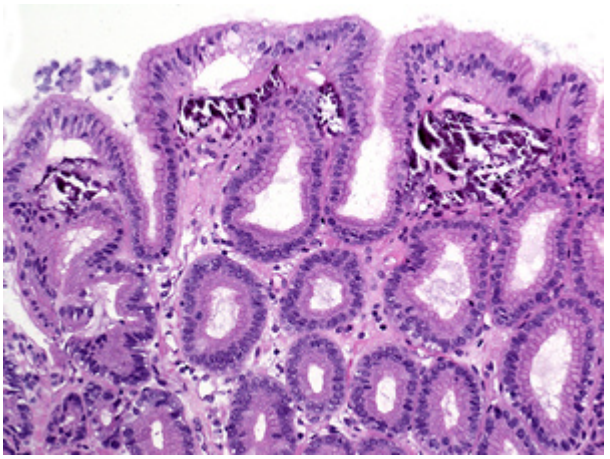
August 2016

Gastric biopsies in an 18-year old male with autoimmune polyglandular syndrome type I (APS-1).

What is your diagnosis?





Diagnosis:

Gastric mucosal calcinosis.

Comment:

An 18-year old male with known autoimmune polyglandular (syn. polyendocrine) syndrome type I (APS-1) underwent endoscopic evaluation of his upper gastrointestinal tract for chronic diarrhoea and unspecific abdominal discomfort.

Multiple white flat plaques, measuring 1 to 3 mm in largest diameter, where detected in the mucosa of the gastric body and antrum (Panel A). Histologic examination of gastric biopsies showed numerous subepithelial microcalcifications (Panel B).

Laboratory testing revealed hypocalcaemia (1.33 mmol/l; normal 2.2-2.65 mmol/l) and hyperphosphataemia (6.12 mg/dl; normal 2.6-4.5 mg/dl), due to the patient's hypoparathyroidism. Creatinine and blood urea nitrogen levels were within normal limits.

Mucosal calcinosis is only rarely encountered in routine biopsies obtained from the stomach. Deposits may be classified as dystrophic, metastatic or idiopathic. Dystrophic calcinosis implies calcification within inflamed, fibrotic or otherwise altered tissue. Metastatic calcinosis is much more common and refers to calcium deposits in the setting of an abnormal serum biochemical environment, usually in otherwise normal tissue. Patients have either hypercalcaemia or hyperphosphataemia, often in conjunction with end stage renal disease. Hyperparathyroidism has been identified as another possible cause, since this disease is able to induce persistent hypercalcaemia. In our patient with severe hypoparathyroidism, the observed mucosal calcinosis can best be explained by a pathological (i.e., elevated) Ca x PO4 product.

Pathologists should mention the presence of gastric microcalcifications in the pathology report, as they may serve as an indicator for generalized metastatic calcifications, especially in organs where they may be fatal, e.g. the heart. It is of further note that some examples of metastatic microcalfications are reversible, with normalization of serum biochemical parameters.

For further reading:

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- Husebye ES, Perheentupa J, Rautemaa R, Kämpe O. Clinical manifestations and management of patients with autoimmune polyendocrine syndrome type I. J Intern Med. 2009; 265: 514-29.
- Yılmaz B, Köklü S, Sökmensüer C. Gastric mucosal calcinosis. Endoscopy. 2013;45 Suppl 2 UCTN:E275.

Presented by:

Dr. Cord Langner and Dr. Christoph Högenauer, Graz, Austria.