ELECTRONIC CLINICAL CHALLENGES AND IMAGES IN GI

Ectopic Tissue in the Stomach Wall Harbors Stunning Diagnosis

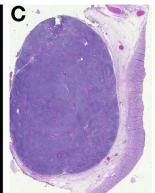


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Question: A 48-year-old patient was referred for gastroscopy and colonoscopy because of anemia. Personal history was otherwise unremarkable except for Wolf-Parkinson-White syndrome. Gastroscopy showed a submucosal 2×4 -cm tumor in the antrum with a central retraction (Figure A, endoscopic finding). On endosonography and MRI, a submucosal tumor was evident, confined to the antral wall, without invasion of the muscularis propria.

We performed an endoscopic submucosal dissection (Olympus GIF – H 180). Tumor margins were marked with argon plasma coagulation. For submucosal injection, we used adrenaline (Bichsel, Interlaken, Switzerland), methylene blue (Hänseler, Herisau, Switzerland), and physiogel (gelatine, B. Braun, Melsungen, Germany). In toto resection of the 2×4 -cm tumor (Figure B, resection procedure) was done both with dual knife (Olympus, Volketswil, Switzerland) and hook knife (Olympus, Volketswil, Switzerland).

Surveillance and 4-day hospitalization were uneventful. Anemia and iron deficiency resolved completely within few days postoperatively after initial iron replacement therapy. Gross pathologic examination showed a well-circumscribed firm submucosal tumor (maximum. 2.5 cm) with a greyish-white cut surface, covered by a normal-appearing gastric mucosa. The histology surprisingly revealed an ectopic lymph node, located in the submucosa, with infiltration of an atypical lymphoid cell population (Figure *C*, overview). The overlying mucosa was normal, without signs of acute or chronic gastritis.

What is the diagnosis?

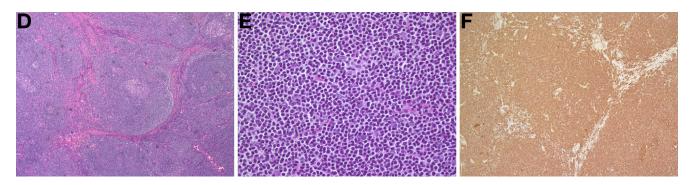
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Conflicts of interest
The authors disclose no conflicts.

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Answer to: Image 2: Nodal Marginal Zone B-Cell Lymphoma



Histologic sections show an enlarged submucosal gastric lymph node with incomplete architectural effacement by a patchy interfollicular neoplastic infiltrate of a nodal marginal zone B-cell lymphoma. In the medium- and high-power views (Figure *D*, *E*), small B-cells are found in the interfollicular area, expanding the marginal zones and filling up sinuses, intermingled with centroblast-like cells throughout the neoplastic infiltrate. Immunohistochemistry highlights strong membranous staining of CD20, indicating the B-cell phenotype of the tumor cells (Figure *F*, immunostain).

There are different types of gastric lymphomas, subdivided into primary and secondary gastric lymphomas. The well-known extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT) type is the most common primary gastric lymphoma and accounts for about 50% of all primary gastric lymphomas. Marginal zone lymphomas are indolent B-cell non-Hodgkin lymphomas and classified into 3 distinct types: MALT, splenic, and nodal types.

MALT lymphomas manifest in various mucosa-associated lymphoid tissue, most frequently in the gastrointestinal tract. Other sites of involvement include salivary gland, lung, thyroid or head and neck, ocular adnexa, skin, and breast.¹

Primary nodal marginal zone lymphomas are very rare B-cell non-Hodgkin lymphomas of unknown etiology and account only for 1.5%–2% of all lymphoid neoplasms.² It is an indolent lymphoma similar to other clinically indolent B-cell lymphomas and affects both women and men in the sixth or seventh decade (median age around 60) with a slight female preponderance.³ Clinically, patients show localized or widespread lymphadenopathy, amongst others of neck, axilla, or femoral region. B symptoms and marrow infiltration are possible features of nodal marginal zone lymphomas, but a leukemic phase is rare.³ Histomorphologically and immunohistochemically, these neoplasms resemble MALT or splenic-type marginal zone lymphomas. In other words, a diagnosis of nodal marginal zone lymphoma strictly requires exclusion of synchronic MALT lymphoma, because involvement of nearby lymph nodes by MALT lymphomas is possible. Hence, to confirm a strictly nodal entity in our patient and to rule out MALT lymphoma, we did gastric mapping as well as computed tomography, both without indication of local or systemic lymphoma spread. Further, to rule out a concomitant *Helicobacter pylori* infection, we did an additional antibody test for anti-Hp immunoglobulin G levels with a negative result. In the previous follow-up 2 months and 4 months after initial diagnosis, the patient has undergone endoscopic surveillance, both endoscopically and histologically in each case without local recurrence, new intragastric focus, or tumor detection. The next endoscopic and histologic control examination will be performed 16 months after diagnosis.

To the best of our knowledge, this is the first case of a strictly nodal marginal zone lymphoma described in a heterotopic lymph node in the gastric wall.

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