AIRP Best Cases in Radiologic-Pathologic Correlation

Gastroblastoma: A Rare Biphasic Gastric Tumor[1]

Teresa Fernandes, MD
Roberto Silva, MD
Vítor Devesa, MD
José Manuel Lopes, MD, PhD
Fátima Carneiro, MD, PhD
Bárbara Viamonte, MD

RadioGraphics 2014; 34:1929–1933

Published online 10.1148/rg.347130103

Content Codes: GI OI

[1]From the Departments of Radiology (T.F., B.V.), Pathology (R.S., J.M.L., F.C.), and General Surgery (V.D.), Centro Hospitalar de São João, Alameda Prof. Hernâni Monteiro 4200, 319 Porto, Portugal; and Medical Faculty and Institute of Molecular Pathology and Immunology, University of Porto (IPATIMUP), Porto, Portugal (J.M.L., F.C.). Received November 4, 2013; revision requested January 22, 2014, and received March 2; accepted March 6. All authors have disclosed no relevant relationships. **Address correspondence to** T.F. (e-mail: *te_fernandes@hotmail.com*).

History

A 19-year-old woman sought medical attention for diffuse abdominal pain that had been present for 5 months. The pain was worse at the inferior quadrants of the abdomen, radiated to the back, and was relieved with the use of oral analgesics. The patient reported that no diarrhea, constipation, anorexia, or weight loss was present. At physical examination, a mass was palpated in the right quadrants of the abdomen. Laboratory findings and medical history were unremarkable.

Imaging Findings

Abdominal ultrasonography revealed a well-defined complex cystic lesion of indeterminate origin that was centered in the right abdominal quadrants with multiple cystic loculi of variable sizes and innumerable irregular internal septa. The contents of the loculi were anechoic, with increased through transmission.

Computed tomography (CT) of the abdomen and pelvis was performed after administration of intravenous contrast material (iobitridol). CT images showed a lobulated complex cystic mass that was 13 cm in the largest dimension and appeared to protrude from the gastric antrum. The wall of the lesion was well defined with some foci of punctiform calcification. Multiple cystic loculi of variable sizes were also seen and were separated by internal septations with a maximal thickness of 10 mm (Fig 1). The wall and internal septa demonstrated moderate enhancement after administration of intravenous contrast material. No nodular enhancing components were detected. Contrast-enhancing adenopathy was seen along the greater curvature of the stomach, with short-axis measurements as much as 8 mm. The lesion was considered to be a protruding mass of the gastric antrum or a primary mesenteric tumor.

Magnetic resonance (MR) imaging of the abdomen with intravenous contrast material (gadobenate dimeglumine) was performed to further evaluate the relationship of the mass with adjacent organs. Again, the lesion seemed to originate from the wall of the gastric antrum (Fig 2). MR images better displayed the cystic loculi compared with CT. The dimensions of the loculi varied from a few millimeters to 8 cm. The fluid content of the loculi was homogeneously hypointense on T1-weighted images and markedly hyperintense on T2-weighted images. The wall and internal septa were isointense relative to muscle both on T2-weighted images and unenhanced T1-weighted images and demonstrated moderate enhancement after administration of intravenous contrast material. Contrast-enhancing adenopathy along the great curvature of the stomach was observed. There were no signs of invasion of adjacent organs or mesenteric vessels, and no parenchymal lesions or peritoneal implants were detected.

Figure 1. Axial **(a)** and coronal **(b)** contrast-enhanced CT images of the abdomen show a multiloculated cystic mass (*) with septal enhancement and scattered calcifications (white arrow in **b**) that measures approximately 13 × 7 × 10 cm in its longitudinal, anteroposterior, and transverse diameters, respectively, and is touching (black arrow in **b**) the gastric antrum *(S)*.

Pathologic Evaluation

Explorative laparotomy was performed and confirmed the origin of the lesion from the posterior wall of the gastric antrum (Fig 3). There were no signs of invasion of adjacent organs or hepatic metastasis. Resection of the tumor and partial distal gastrectomy were performed, and 15 lymph nodes were resected.

The surgical specimen revealed a nodular, well-circumscribed tumor in the gastric antrum that was 10.5 cm in its largest dimension. On the cut surface, the tumor was solid, white-gray in color, and partially cystic with areas of hemorrhage (Fig 4).

The tumor was centered in the muscular layer of the gastric wall and was biphasic with well-demarcated, epithelial, and predominantly mesenchymal components. The epithelial component was arranged in nests, cords, and tubules that contained luminal eosinophilic secretory material. The mesenchymal component comprised short bundles of spindle cells, foci of multinucleated cells, and variable myxoid or collagenous stroma (Fig 5). Atypia of neoplastic cells was mild, and the mitotic index was fewer than five mitoses per 50 high-power fields. There was no evidence of lymphovascular or perineural tumor invasion or lymph node metastasis.

At immunohistochemical analysis, the epithelial component stained strongly and diffusely for pancytokeratin (cytokeratin markers AE1–AE3 and CAM 5.2), CD56, and, focally, CD10. The mesenchymal component expressed vimentin, CD10, and, focally, CD56. The tumor cells did not express markers that are usually present in gastrointestinal stromal (ie, c-KIT and DOG-1), solitary fibrous (ie, CD34), nerve sheath (ie, S100 protein), or mesothelial (ie, calretinin) tumors. Neuroendocrine markers (eg, chromogranin A and synaptophysin) and smooth muscle markers (eg, smooth muscle actin and desmin) were also not expressed.

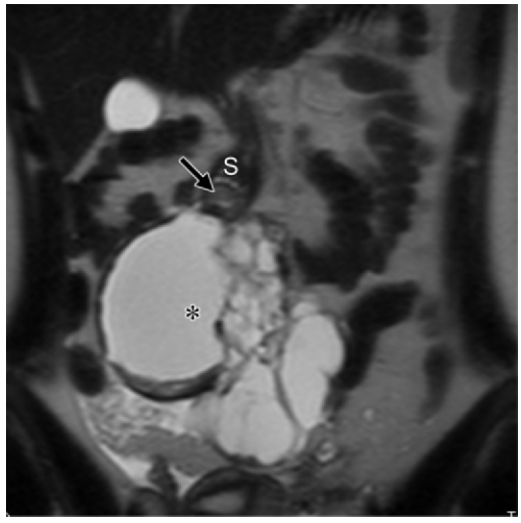

Figure 2. Coronal T2-weighted MR image shows the multiloculated cystic mass (*) arising (arrow) from the gastric antrum *(S)*.

Discussion

Gastroblastoma is a recently described tumor of the stomach, with only five reported cases to date. It is a biphasic neoplasm with mixed epithelial and mesenchymal components in various proportions, with minimal cytologic atypia and indolent clinical behavior (1–3).

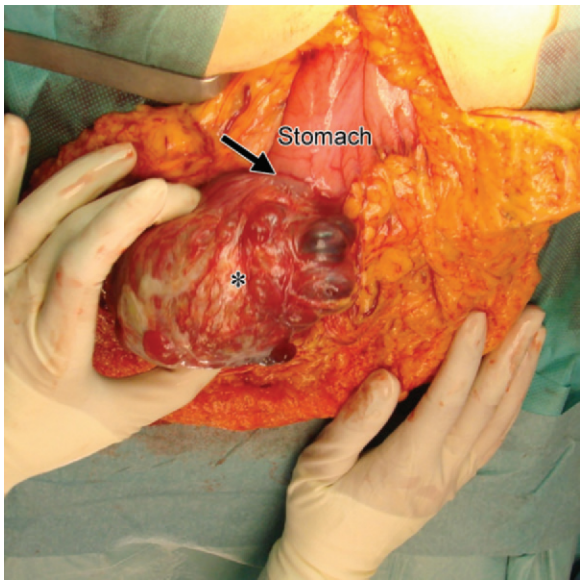

Figure 3. Intraoperative photograph obtained after median laparotomy of the superior abdomen confirms that the well-defined lobulated mass (*) protrudes (black arrow) from the gastric antrum.

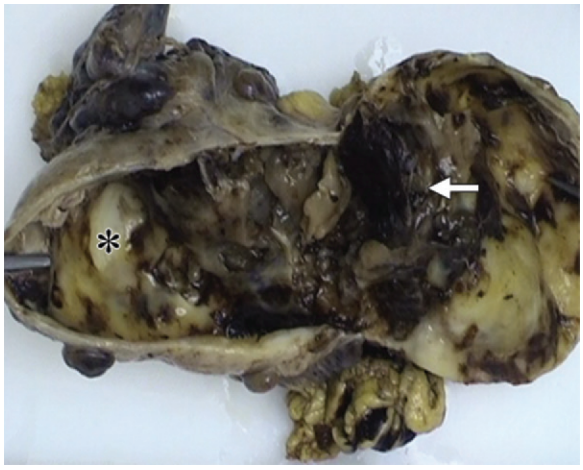

Figure 4. Photograph of the gross specimen shows the cut surface of the mass, which is solid (*) and hemorrhagic (arrow) with cystic degeneration.

Reported cases occurred in both sexes, with ages ranging from 9 to 30 years (mean age, 23 years). Clinically, patients presented with nonspecific symptoms such as vague abdominal pain, constipation, and fatigue (1–3). Tumors originated either from the wall of the antrum or the greater curvature of the stomach and were relatively large by the time of diagnosis, ranging from 3.5 cm to 15 cm in the largest dimension (mean size, 7.9 cm) (1–3).

In two cases, the lesion was seen at CT as a partially circumscribed heterogeneous mass with solid and cystic components. One was centered in the gastric wall, whereas the other manifested as a partially exophytic mass (2,3). To the best of our knowledge, imaging findings of the remaining three tumors were not described, even though one was seen as an exophytic mass on the greater curvature of the stomach at gross pathologic analysis (1).

In our case, both CT and MR imaging depicted a complex cystic mass with some foci of calcification. The differential diagnosis included tumors with an origin in the gastric wall, such as gastrointestinal stromal tumor (GIST) and gastric teratoma. Cystic mesothelima and mesenteric lymphangioma were also considered.

GISTs are the most common mesenchymal tumors of the gastrointestinal tract and most frequently occur in the stomach. They tend to grow exophytically and are usually large at the time of manifestation (4). At imaging, they commonly appear as a well-defined heterogeneous mass in which necrosis and cystic changes may occur. Features that indicated a diagnosis other than GIST were the young age of the patient and the presence of calcifications, which are not typically seen in GISTs before specific chemotherapy is performed (4).

Gastric teratoma is a rare tumor that may manifest as an exophytic lobulated heterogeneous mass with variable cystic and solid components and calcifications (5,6). However,

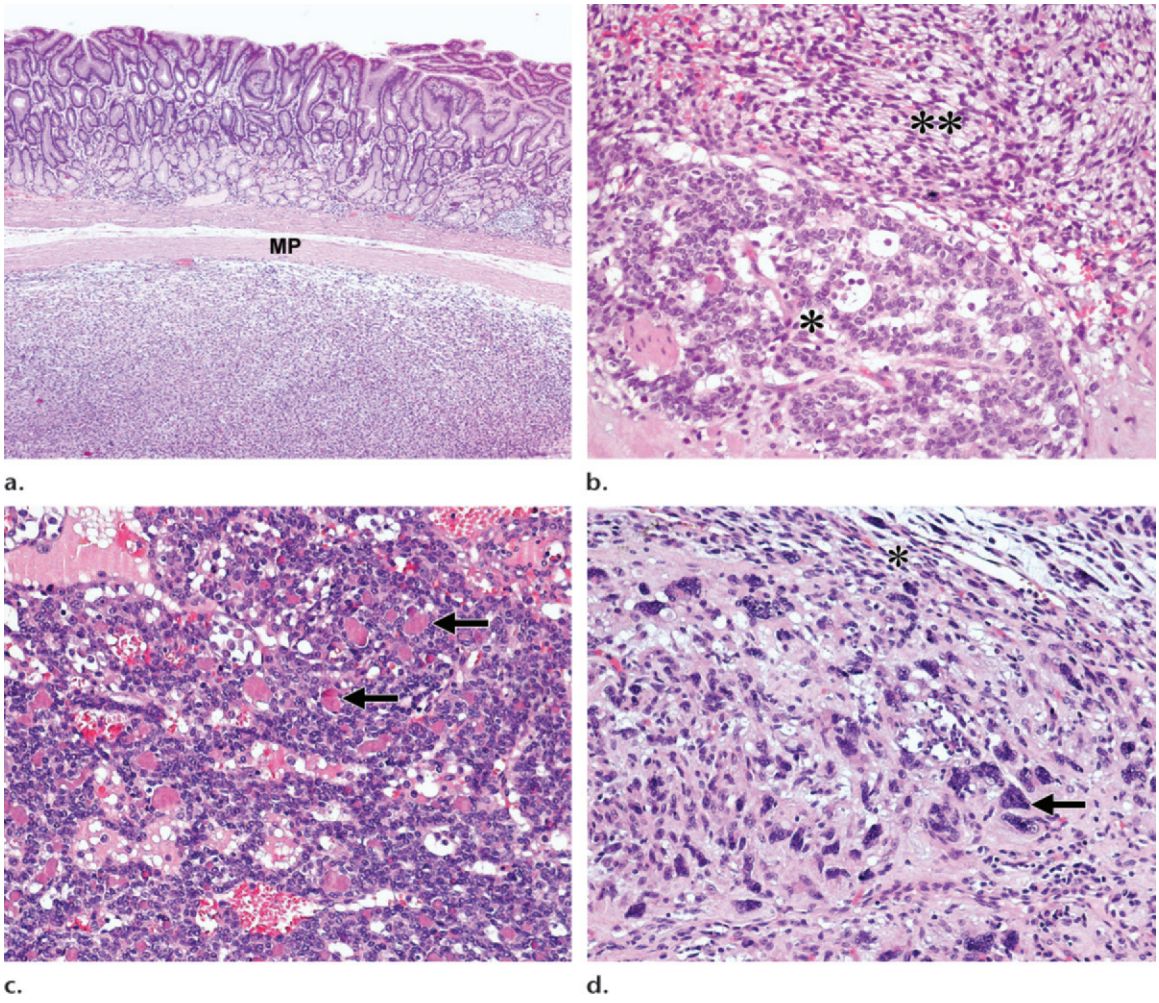

Figure 5. **(a)** Photomicrograph (hematoxylin-eosin [H-E] stain; original magnification, ×40) shows that the tumor is centered in the muscularis propria *(MP)* and is well circumscribed. **(b)** Photomicrograph (H-E stain; original magnification, ×200) shows that the tumor is biphasic with a sharply defined demarcation between the epithelial (*) and mesenchymal (**) components. **(c)** Photomicrograph (H-E stain; original magnification, ×200) shows the epithelial component, which comprises nests and tubules (arrows) with eosinophilic luminal material. **(d)** Photomicrograph (H-E stain; original magnification, ×200) shows the mesenchymal component, which is composed of short bundles of spindle cells (*) and scattered multinucleated giant cells (arrow) in a myxoid or collagenous stroma.

gastric teratomas usually occur during infancy or childhood. In addition, the absence of fat did not support this diagnosis (5).

The multilocular form of cystic mesothelioma may have imaging findings similar to those seen in our case and typically occurs in young to middle-aged women with no history of asbestos exposure (7,8). However, multilocular mesothelioma characteristically originates in the pelvis (rather than the abdominal cavity, as in our case) (7,8). Cystic lymphangioma usually occurs in young patients and may be seen in the mesentery as a complex cystic mass with scattered calcifications (7,9). Because of its exophytic growth, it was also included in the differential diagnosis.

In our case, the lesion was centered in the muscular layer of the gastric wall, which explains its exophytic growth. In addition, the complex cystic appearance that was seen at imaging was a result of cystic degeneration of the tumor.

Gastroblastoma should be included in the differential diagnosis along with other biphasic gastric tumors such as biphasic synovial sarcoma, teratoma, and carcinosarcoma (1,2). Its characteristic histologic features include demarcation between epithelial and mesenchymal components; mild atypia; a low mitotic index; and variable immunohistochemical staining with CD56 and CD10, a finding that helps confirm the diagnosis (1,2).

In the previously reported cases, all patients underwent surgical resection of the mass and partial or total gastrectomy. In four cases, the lesion was confined to the stomach, and patients remained disease free during follow-up periods that ranged from 9 months to 14 years after sur-

gery (1,2). However, in one recent reported case of gastroblastoma, regional nodal metastasis and clinical metastases to the liver and pelvis were seen at the time of diagnosis, providing evidence of the malignant potential of these tumors (3). To date, our patient has remained disease free for 20 months after tumor resection. Given the limited number of reported cases, the prognosis associated with gastroblastoma is uncertain. According to Miettinen et al (1), these tumors should at least be considered of low-grade malignancy.

In conclusion, we report one case of gastroblastoma that originated in the muscular layer of the gastric antrum and demonstrated exophytic growth through the gastric wall. At imaging, it was seen as a complex mainly cystic mass, a result of extensive cystic degeneration. Imaging findings of gatroblastomas vary according to their proportions of epithelial and mesenchymal components and the involvement of the gastric wall structures, which varies. Although they are extremely uncommon, we believe that they should be considered in the unlikely event of a focal gastric mass that manifests at a young age, especially if it is large, exophytic, and heterogeneous (both cystic and solid).

References

1. Miettinen M, Dow N, Lasota J, Sobin LH. A distinctive novel epitheliomesenchymal biphasic tumor of the stomach in young adults ("gastroblastoma"): a series of 3 cases. Am J Surg Pathol 2009; 33(9):1370–1377.
2. Shin DH, Lee JH, Kang HJ, et al. Novel epitheliomesenchymal biphasic stomach tumour (gastroblastoma) in a 9-year-old: morphological, ultrastructural and immunohistochemical findings. J Clin Pathol 2010;63(3):270–274.
3. Wey EA, Britton AJ, Sferra JJ, Kasunic T, Pepe LR, Appelman HD. Gastroblastoma in a 28-year-old man with nodal metastasis: proof of the malignant potential. Arch Pathol Lab Med 2012;136 (8):961–964.
4. Sandrasegaran K, Rajesh A, Rydberg J, Rushing DA, Akisik FM, Henley JD. Gastrointestinal stromal tumors: clinical, radiologic, and pathologic features. AJR Am J Roentgenol 2005;184(3):803–811.
5. Singh M, Rattan KN, Kadian YS, Hasija N. Gastric teratoma: a rare neoplasm. J Neonat Surg 2012;1 (2):28.
6. Gore MD, Fernbach SK. Case 52: gastric teratoma. Radiology 2002;225(2):497–499.
7. Park JY, Kim KW, Kwon HJ, et al. Peritoneal mesotheliomas: clinicopathologic features, CT findings, and differential diagnosis. AJR Am J Roentgenol 2008;191(3):814–825.
8. Weiss SW, Tavassoli FA. Multicystic mesothelioma: an analysis of pathologic findings and biologic behavior in 37 cases. Am J Surg Pathol 1988;12(10): 737–746.
9. Levy AD, Cantisani V, Miettinen M. Abdominal lymphangiomas: imaging features with pathologic correlation. AJR Am J Roentgenol 2004;182 (6):1485–1491.