A 56-YEAR-OLD previously healthy woman came for treatment after an episode of hematemesis. She denied a history of weight loss, excessive alcohol ingestion, difficulty swallowing, symptoms of gastric outlet obstruction, previous hematemesis, esophagitis, gastritis, gastric or duodenal ulcer, or a diagnosis of Helicobacter pylori infection. Physical examination demonstrated a seemingly healthy middle-aged woman with mild epigastric tenderness but no other abnormal findings. She was treated in an urgent fashion with resuscitation using a crystalloid solution, insertion of a nasogastric tube, and was to receive nothing by mouth. Laboratory tests were also performed expeditiously. The patient’s hemoglobin level was 7.45 mmol/L (12.0 g/dL), platelet count was 220.0 × 10^9/L, and international normalized ratio and partial thromboplastin time were normal. Early upper gastrointestinal tract endoscopy results revealed a fundal mass with an overlying ulcer. No active bleeding was present. Biopsy results were normal. Upper gastrointestinal x-ray films demonstrated a 4-cm circular mass outlined by contrast enhancement in the fundus of the stomach (Figure 1). A computed tomographic scan of the abdomen showed a gastric fundal mass adjacent to the diaphragm and spleen.

What Is the Most Likely Diagnosis in This Patient?

A. Gastric lymphoma
B. Giant type V gastric ulcer
C. Gastric stromal tumor
D. Linitis plastica
Based on the patient's history, physical examination, and the mass in the gastric wall, a presumptive diagnosis of a gastric stromal tumor was made. On exploratory laparotomy, a 4-cm mass was found in the posterior wall of the fundus. A partial gastric resection with a 2-cm rim of normal gastric wall was performed using a stapling device. Examination of the specimen demonstrated a 3-cm ulceration over the mass (Figure 2). Intraoperative pathological consultation described a 4 x 3-cm benign leiomyoma surrounded by a 2-cm rim of normal gastric wall. The patient's postoperative course was uneventful and she has continued to do well.

Reviews of gastric stromal tumors have credited Giovanni Battista Morgagni with the first description of a gastric leiomyoma in 1762. Despite numerous publications that have discussed leiomyomas, leiomyosarcomas, and leiomyoblastomas since that time, these diagnoses account for fewer than 2% to 3% of surgical resections performed for gastric neoplasms. Gastric leiomyomas smaller than 1 cm, however, were found in 46% of stomachs carefully sectioned at autopsy in 50 patients aged 19 to 82 years in one older study. While children can be affected, the onset of symptoms in most patients occurs in the fifth or sixth decade of life.

Gastric stromal tumors cause epigastric pain and/or upper gastrointestinal hemorrhage. When a leiomyoma or other stromal tumor in the gastric wall grows to a size of 3 to 4 cm, ulceration of the overlying mucosa occurs. In some patients, epigastric pain resembling that of a gastric ulcer results. Chronic blood loss is a more common presentation, although hematemesis has been reported. With leiomyosarcomas or leiomyoblastomas, hemorrhage has been reported to occur in 50% of patients. Finally, asymptomatic gastric stromal tumors may be discovered on radiographs, during endoscopic evaluations of the stomach, or at an exploratory laparotomy for another disease process.

A rounded mass with smooth edges and reasonably normal overlying mucosa is the characteristic appearance on a barium study (Figure 1). Endoscopic biopsy findings may be deceptive as in this patient unless ulceration is complete and the neoplasm is exposed. Also, histologic differentiation between a benign or malignant gastric stromal tumor based on the endoscopic biopsy results may not be possible. On an abdominal computed tomographic scan, larger size (>5 cm), lobulated contour, heterogeneous enhancement, ulceration, and exophytic growth pattern are all statistically significant predictors of a malignant gastrointestinal stromal tumor. On endoscopic ultrasound, irregular extraluminal margins, cystic spaces, and malignant lymph nodes had a positive predictive value of 100% for borderline or malignant gastrointestinal stromal cell tumors in one French study.

There have been continuing difficulties in differentiating benign from low-grade malignant gastric stromal neoplasms for many years. For presumably benign lesions, enucleation at operation is contraindicated. Resection of the gastric wall with a 2- to 3-cm cuff of normal tissue is appropriate. A larger exophytic lesion may require a formal gastrectomy with similar margins and resection of adherent omentum, lymph nodes, or adjacent organs. While this patient underwent surgery in the prelaparoscopic era, laparoscopic resections are now routine for presumably benign lesions.

In recent years, acronyms such as GIST (gastrointestinal stromal tumor) and STUMP (smooth muscle tumor of uncertain malignant potential) have been used by pathologists when describing tumors formerly known as leiomyomas, leiomyosarcomas, and leiomyoblastomas. This reflects the problem of correlating histologic appearance with clinical outcome and continues to prompt newer approaches to differentiation.

Benign leiomyomas are cured by resection of the entire tumor with a rim of normal gastric wall. Patients with gastrointestinal leiomyosarcomas undergoing curative resection have a 5-year actuarial survival rate of 54%. This figure is deceptive in light of the 18-month disease-free survival for high-grade lesions vs the 8-year disease-free survival rate of 80% for patients with low-grade lesions reported in one series. When only gastric leiomyosarcomas are evaluated, the survival figures are similar.

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REFERENCES


Submissions

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