

Giant gastrointestinal stromal tumor of the esophagus presenting with dyspnea

Ahmet Basoglu, MD,^a Ekrem Kaya, MD,^b Burain Celik, MD,^a and Levent Yıldız, MD,^c Samsun and Bursa, Turkey

Gastrointestinal stromal tumors (GISTs) are rare neoplasms thought to arise from mesenchymal cells of the gastrointestinal tract. These tumors demonstrate a pathobiology and clinical behavior different from those of smooth muscle and Schwann cell tumors.¹ GISTs account for 0.1% to 3% of all tumors in the gastrointestinal tract. Two thirds of these tumors arise from the stomach, 25% arise from the small intestine, and 5% arise from the esophagus.²

We describe a case of giant GIST of the esophagus that compressed the right main bronchus and caused dyspnea.

Clinical Summary

A 33-year-old woman was admitted to our hospital with a 6-year history of dyspnea and 2-year history of dysphagia and back pain. She had been treated for asthma for 6 years. On admission, chest radiograph showed a widened mediastinum.

Esophagogram showed a dilated thoracic esophagus and irregular mucosa in two thirds of the thoracic esophagus. Chest computed tomography revealed massive circumferential thickening of the esophageal wall extending from the level of the thoracic inlet to the cardia of the stomach. The giant mass compressed the right main bronchus (Figure 1).

A mucosal ulceration 33 cm from the incisors and external compression were confirmed by esophagoscopy. Endoscopic ultrasonography was undertaken, which confirmed circumferential thickening of the esophageal wall 6 to 8 cm along the thoracic esophagus.

Proximal gastrectomy, subtotal esophagectomy, and esophago-gastrostomy were performed through the abdominothoracic approach. The tumor weighed 2800 g and was 27 cm long (Figure 2). The postoperative course was uncomplicated, and the patient was discharged on the 12th postoperative day. The patient has been doing well with no recurrence 3 years after the operation.

On histopathologic examination, we observed a tumoral mass in the esophageal wall between the submucosal and subserosal layer. We diagnosed this tumor as a GIST of the esophagus. The tumor was composed of spindle cells with the following features: mitotic count, 3 to 5/50 high-power field; CD117 staining, 10% to 50%/+++; CD34 staining, 10% to 50%/+++.

From the ^aOndokuz Mayıs University, Medical School, Department of Thoracic Surgery, Samsun; ^bUludag University, Medical School, Department of Surgery, Bursa; and ^cOndokuz Mayıs University, Medical School, Department of Pathology, Samsun, Turkey.

Received for publication Nov 29, 2005; accepted for publication Dec 16, 2005.

Address for reprints: Ahmet Basoglu, MD, Ondokuz Mayıs University Medical School, Department of Thoracic Surgery, 55139 Kurupelit, Samsun, Turkey (E-mail: ahmetb@omu.edu.tr).

J Thorac Cardiovasc Surg 2006;131:1198-9

0022-5223/\$32.00

Copyright © 2006 by The American Association for Thoracic Surgery

doi:10.1016/j.jtcvs.2005.12.046



Figure 1. A, Esophagogram shows large, irregular intramural mass. B and D, Computed tomography (CT) scan shows luminal dilatation and wall thickening. C, Giant mass compresses the right main bronchus.

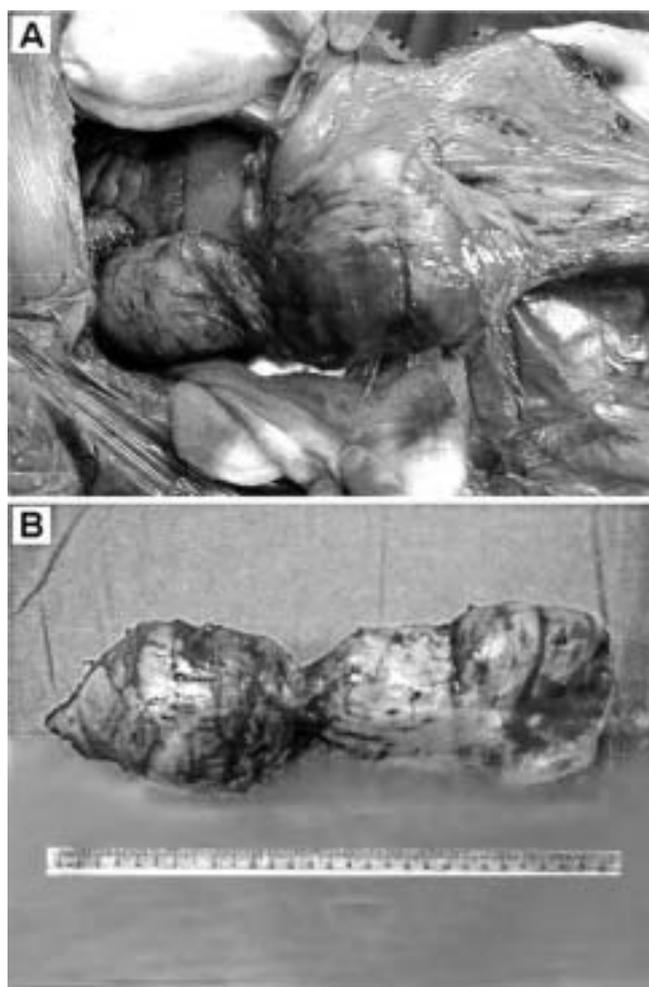


Figure 2. A, Tumor invades the esophagus and cardia. B, Resected tumor.

Discussion

GISTs were previously thought to be smooth muscle neoplasms, and most were classified as leiomyoma or leiomyosarcoma. With the advent of immunohistochemistry and electron microscopy, it became apparent that GISTs might have myogenic features, neural attributes, or characteristics of both muscle and nerve. Immunohistochemically, the tumor cells are positive for CD117 and CD34.^{2,3} The majority of GISTs occur in the subdiaphragmatic gastrointestinal tract, but a small number of cases have been reported in the esophagus.^{1,2}

Symptoms usually depend on tumor size and location. Dysphagia is the most common symptom. The other symptoms include retrosternal discomfort, dyspepsia, and vomiting.^{1,4} In our case, the tumor was too large and compressed the right main bronchus. To our knowledge, this is the first encounter with a case of giant GIST presenting with dyspnea.

GISTs occur in the older patient population, and the majority of them are benign (60%-80%). The most consistent prognostic factors are mitotic count (>5/50 high-power field) and tumor size (>5 cm). These tumors seem to be resistant to chemoradiation. En bloc surgical excision is the treatment of choice of these tumors.^{1,2,5}

It is likely that our patient has a rare case of giant esophageal tumor caused by dyspnea. Because of the lack of any effective alternative therapies, surgical resection should be considered for all patients with GISTs.

References

1. Lee JR, Anstadt MP, Khwaja S, Green LK. Gastrointestinal stromal tumor of the posterior mediastinum. *Eur J Cardiothorac Surg.* 2002;22:1014-6.
2. DeMatteo RP, Lewis JJ, Leung D, Mudan SS, Woodruff JM, Brennan MF. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg.* 2000;231:51-8.
3. Fletcher CD, Berman JJ, Corless C, Gorstein F, Lasota J, Longley BJ, et al. Diagnosis of gastrointestinal stromal tumors: a consensus approach. *Hum Pathol.* 2000;33:459-65.
4. Graadt van Roggen JF, van Velthuysen ML, Hogendoorn PC. The histopathological differential diagnosis of gastrointestinal stromal tumors. *J Clin Pathol.* 2001;54:96-102.
5. Miettinen M, Lasota J. Gastrointestinal stromal tumors—definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch.* 2001;438:1-12.