Pleural dissemination of esophageal gastrointestinal stromal tumors after an eight-year interval following the primary surgery

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Abstract A 71-year-old man who had undergone surgical resection of esophageal gastrointestinal stromal tumors (GISTs) through a right posterolateral thoracotomy 8 years earlier was referred for treatment of an anterior mediastinal mass discovered on a follow-up chest radiograph in October 2007. Computed tomography findings revealed a tumor, 82 × 49 mm, with calcification, in the anterior mediastinum. When we radically resected the tumor via a median sternectomy, we found that it was actually located in the pleural cavity, and there was a small nodule near the main tumor on other pleura. Microscopically, the tumor was comprised of uniform spindle cells with fibrillary eosinophilic cytoplasm. In addition, immunostaining showed that the tumor was positive for CD117 (c-kit). The diagnosis was pleural dissemination of esophageal GISTs 8 years after primary surgery, making this the first report of pleural dissemination of esophageal GISTs after such a prolonged postsurgical interval.

Key words Mediastinal tumor · Gastrointestinal stromal tumor · Pleural dissemination

Introduction Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumor of the gastrointestinal tract. GISTs have several degrees of malignant potential. They exhibit pathobiology and clinical behavior different from those of smooth muscle and Schwann cell tumors; and they reportedly arise in all parts of the alimentary tract, as well as from the omentum, mesentery, peritoneum, and retroperitoneum. On the other hand, they rarely occur in the esophagus. Here we present the case of a 71-year-old man who had undergone surgical resection of esophageal GISTs 8 years earlier and was now exhibiting pleural dissemination of GISTs.

Case A 71-year-old man had undergone surgical resection of esophageal submucosal tumors similar to giant polyps, 120 × 55 mm, through a right posterolateral thoracotomy 8 years earlier. The tumor location at that time was chest upper esophageal submucosa with steal, 17 × 12 mm; it was not invasive to the esophageal mucosa or the surrounding organ. Enucleation with excision of surrounding esophageal muscle was performed. That operation was curative surgery, and no tumor cells existed on the surgical stump. The histology indicated that it was a gastrointestinal stromal tumor (GIST) with spindle cells and loose connective tissue with CD117 (c-kit) and CD34 part positivity and Ki-67(Mib-1 index) negativity on immunostaining. Adjuvant chemotherapy was not undertaken because he had chronic renal failure.

During October 2007, he was referred for treatment of an anterior mediastinal mass discovered on a follow-up chest radiograph (Fig. 1). The patient had no dysphagia or chest symptoms, and no tumor had been present on a radiograph obtained a year earlier. Computed tomography (CT) revealed the tumor to be 82 × 49 mm...
size, with calcification, and to be located in the anterior mediastinum, perhaps directly invading the superior vena cava (SVC) (Fig. 2). Based on the radiological findings, an anterior mediastinal tumor was suspected, and a thymoma or teratoma seemed most likely. The results of baseline laboratory testing were all within the normal range, as were the levels of the tumor markers carcino-embryonic antigen and squamous cell carcinoma antigen.

In December 2007, we radically resected the tumor via a median sternectomy. At that time, we found that the tumor was actually located in the pleural cavity, originating from the parietal pleura, and that neither the mediastinum nor the SVC had been invaded. Part of the right upper pulmonary lobe was also resected, as the tumor was firmly attached to the lung. In addition, we detected a small (5 mm) nodule near the main tumor on the pleura; that lesion was widely resected along with the parietal pleura. No other tumors were found (Fig. 3).

Grossly, the main tumor measured about 100 × 85 mm, was elastic and hard, and the cut surface was white. Microscopically, the main tumor had uniform spindle cells with fibrillary eosinophilic cytoplasm and nucleoli containing fine chromatin as well as inconspicuous nucleoli (Fig. 4a). Immunostaining revealed CD117 (c-kit) positivity, with the positive cells staining dark brown (Fig. 4b). Expression of Ki-67 (MiB-1 index) was detected in more than 30% of tumor cells, indicating that the tumor had malignant potential; S-100 protein and CD34 assays were negative. The immunohistological findings for the small nodule on the pleura were the same as for the main tumor. Based on these findings, the diagnosis for both the main tumor and the small nodule was pleural dissemination of esophageal GIST 8 years after the primary surgery.

The patient was started on imatinib as adjuvant chemotherapy. He was alive with disease 20 months after surgery.

Discussion

In 1983, Mazur and Clark challenged the longstanding concept that most mesenchymal tumors of the stomach were of smooth muscle origin and introduced the concept