

Extra-Gastrointestinal Stromal Tumor Presenting as an Anterior Chest Wall Mass

Junghyeon Lim, M.D.¹, Sung Woo Cho, M.D.², Hee Sung Lee, M.D.³,
Hyoungho Kim, M.D.¹, Yong Han Kim, M.D.¹, Bong Suk Park, M.D.¹

¹Department of Cardiothoracic Surgery, Hanllym University Sacred Heart Hospital, Hanllym University College of Medicine,

²Department of Cardiothoracic Surgery, Hanllym University Kangdong Sacred Heart Hospital, Hanllym University College of Medicine, ³Department of Cardiothoracic Surgery, Hanllym University Dongtan Sacred Heart Hospital, Hanllym University College of Medicine

A 71-year-old man was referred for an anterior chest wall mass. Chest computed tomography (CT) and positron emission tomography-CT suggested a malignant tumor. Surgical biopsy through a vertical subxiphoid incision revealed an extra-gastrointestinal stromal tumor (EGIST). En bloc resection of the tumor, including partial resection of the sternum, costal cartilage, pericardium, diaphragm, and peritoneum, was performed. Pathologic evaluation revealed a negative resection margin and confirmed the tumor as an EGIST. On post-operative day 17, the patient was discharged without any complications. At the 2-week follow-up, the patient was doing well and was asymptomatic.

Key words: 1. Extra-gastrointestinal stromal tumor
2. Gastrointestinal stromal tumors
3. Chest wall tumor

Case report

A 71-year-old man was referred for evaluation of a palpable mass on his anterior chest wall. He had no dysphagia or chest symptoms. Based on findings from chest computed tomography (CT), the mass was suspected to be a malignant tumor, such as a sarcoma, plasmacytoma, or metastatic cancer (Fig. 1). Positron emission tomography (PET)-CT was performed as part of a metastatic cancer work-up. PET-CT revealed that the mass was probably a plasmacytoma or sarcoma and revealed focal hypermetabolic activity in the rectum suspicious of a rectal polyp or malignant tumor. A surgical biopsy of the tumor on the anterior chest wall was performed through a sub-

xiphoid vertical incision. C-KIT (3+) and CD34 (3+) immunohistochemistry suggested that the tumor was an extra-gastrointestinal stromal tumor (EGIST). Resection of the tumor was performed through an anterior midline incision from the level of the fourth rib to 2 cm below the xiphoid process. The tumor had invaded the lower sternum, the fourth to the eighth costal cartilages on both sides, the pericardium, the diaphragm, and the peritoneum. There was no invasion of the lung or the liver. En bloc resection of the tumor, including partial resection of the sternum, costal cartilage, pericardium, diaphragm, and peritoneum, was performed (Fig. 2). To achieve a negative resection margin, the tumor was resected radically, maintaining a 3- to 5-cm margin grossly. The

Received: September 29, 2016, Revised: October 20, 2016, Accepted: October 24, 2016, Published online: August 5, 2017

Corresponding author: Sung Woo Cho, Department of Cardiothoracic Surgery, Hanllym University Kangdong Sacred Heart Hospital, 150 Seongan-ro, Gangdong-gu, Seoul 05355, Korea
(Tel) 82-2-1588-4100 (Fax) 82-2-488-0114 (E-mail) cswoo1@hallym.or.kr

© The Korean Society for Thoracic and Cardiovascular Surgery. 2017. All right reserved.

© This is an open access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

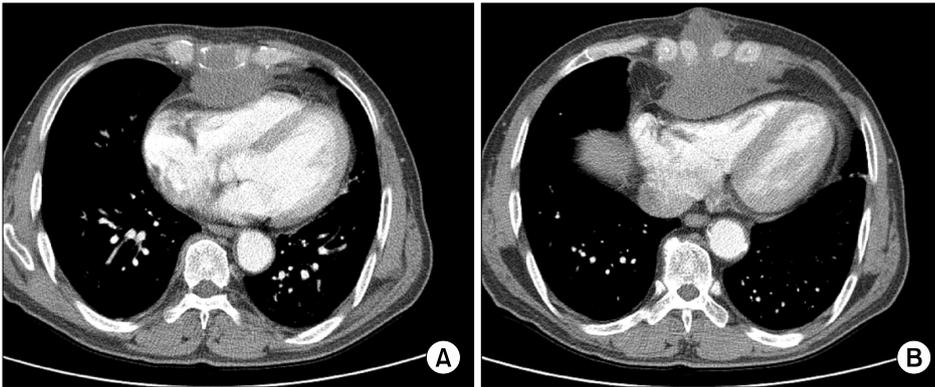


Fig. 1. (A, B) Preoperative chest computed tomography image. Computed tomography revealed an irregular mass measuring about 12 cm in the sternal body, involving anteriorly, the subcutaneous fat layer; posteriorly, the lower anterior mediastinum; and inferiorly, the diaphragm and the upper anterior abdomen.

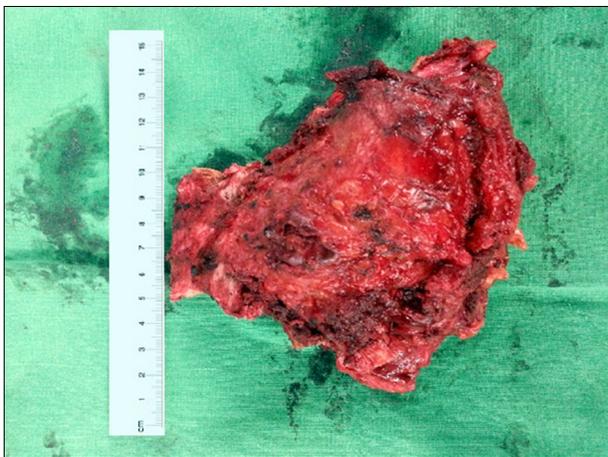


Fig. 2. Specimen after en bloc resection. En bloc resection of the tumor, including partial resection of the sternum, costal cartilage, pericardium, diaphragm, and peritoneum, was performed.

partially resected diaphragm and peritoneum were reconstructed with a bovine pericardial patch. The anterior chest wall was reconstructed with a 2-mm Gore-Tex patch. On postoperative day 2, the patient was transferred to the general ward. On postoperative day 6, his chest tube was removed. Because a rectal polyp was revealed on PET-CT imaging, a colonoscopy was performed preoperatively. A biopsy of the rectal polyp was performed during the colonoscopy, and the pathologic report indicated a well-differentiated adenocarcinoma. On postoperative day 11, the rectal polyp was resected through endoscopic submucosal dissection. Pathologic evaluation after the en bloc resection of the tumor revealed a negative resection margin, and the tumor was definitively confirmed to be an EGIST. On postoperative day 17, the patient was discharged without any complications.

At the 2-week follow-up, the patient was doing well and exhibited no symptoms. The patient was referred to an oncologist for further follow-up and adjuvant therapy. Although further surgery for rectal cancer was required, the patient refused to undergo it. The oncologist recommended follow-up visits every 3 months thereafter.

Discussion

In 1983, Mazur and Clack first described a gastric stromal tumor. The concept of gastric stromal tumor was extended to include mesenchymal tumors without demonstration of smooth muscle or Schwann cell differentiation [1]. The most common mesenchymal tumors of the gastrointestinal (GI) tract are GI stromal tumors (GISTs). GISTs primarily occur in the stomach or the small intestine, but they can be found anywhere in the GI tract. Mutation of the *c-KIT* gene, inducing an over-expression of the KIT receptor, is the main pathogenesis [2].

There are more benign GISTs (60%–80%) than there are malignant GISTs. The diagnosis of a mediastinal malignant GIST is associated with tumor necrosis, mitosis counts, and invasion [1]. The relative prevalence of benign and malignant GISTs varies by tumor location. Benign GISTs are more frequent in the stomach, whereas they are less common than malignant GISTs in the intestines. At presentation, metastatic GISTs show an extremely poor prognosis. Mitotic rate, the tumor size, and the tumor site are 3 prognostic factors. GISTs showing mitotic activities of less than 5 mitoses per high-power field (HPF) and that are smaller than 2 cm show better prognoses. In the stomach, most tumors are benign, and the mi-

otic count is generally fewer than 5 mitoses per 50 HPFs. Some GISTs do not show mitotic activity and have little possibility of metastasis. Mitotic rates greater than 5 mitoses per 50 HPFs are usually found in malignant GISTs. Although it is not always available, information about the Ki67 status of a tumor is useful for identifying its malignant potential. For a more accurate assessment of a tumor's malignant potential, a genetic evaluation can be useful [3].

EGISTs, which are GISTs outside of the GI tract, are very rare; only approximately 5% of GISTs occur outside of the GI tract. Metastasis from a primary GIST is a common form of EGIST. Common sites for EGISTs are the omentum, mesentery, and retroperitoneum. Immunohistochemical evaluations of c-KIT protein (CD117) and CD34 for the diagnosis of EGIST are valuable and reliable modalities [4].

Interstitial Cajal cells are considered to be the origin of GISTs. However, the exact origin of EGISTs remains controversial. Several studies have suggested the stomach to be the origin of omental EGISTs and the small intestine to be the origin of mesenteric EGISTs. In another study, GISTs were reported to originate from multipotent mesenchymal stem cells [5].

In the present case, results of the diagnostic work-ups, including CT and endoscopies, suggested that the anterior chest wall mass was not a metastatic lesion from another primary cancer. Other findings suggestive of an EGIST were the immunohistochemical findings, such as those for c-KIT and CD34, and the large, asymptomatic tumor. In this case, the EGIST was suspected to have originated from the peritoneum, because other tissues invaded by the tumor, including the pericardium or pleura, are very rarely reported as the origins of GISTs.

Currently, surgery is the definitive treatment for GISTs and EGISTs. Because a preoperative confirmative diagnosis of EGIST is difficult, obtaining a specimen intraoperatively for freezing is necessary. If the results of the pathological analysis of the frozen section reveal a stromal tumor, the tumor should be considered malignant. Because metastasis to lymph nodes from GISTs is not frequent (<10%), radical lymph node dissection is not necessary. Even when there is a pathologically confirmed complete negative resection margin, tumors recur frequently. In the literature, 4 of the 9 reported cases experienced re-

currence within 10 years after the first treatment. Adjuvant therapy seems to be required because of the recurrence rate. However, neither chemotherapy nor radiation has shown statistically significant benefits for treating GISTs or EGISTs [6].

When patients require resection of the chest wall, surgeons should be cautious about 3 points. To resect all of the non-viable tissue, sufficient tissue resection should occur. Second, when radical chest wall resection is needed, chest wall reconstruction should be considered to prevent flail chest. Lastly, viable healthy tissue coverage is important for protecting visceral organs and vessels, filling the pleural space, and protecting against infection.

Currently, there are many options for reconstruction. When the defect is smaller than 5 cm or the location of the defect is covered by the scapula, reconstruction using only soft tissue can be stable without the need for skeletal component reconstruction. However, huge chest wall resection and/or a collapsed lung might require chest wall reconstruction and stabilization with a prosthetic material for replacing skeletal components. The ideal prosthetic material for chest wall reconstruction needs to have the following characteristics: (1) rigidity for preventing flail chest, (2) inertness for ingrowth of tissue and preventing infection, (3) flexibility for adjustment to the appropriate shape, and (4) radiolucency for easy follow-up with a simple radiologic evaluation. When chest wall reconstruction requires a prosthetic material, Prolene mesh does not significantly differ from polytetrafluoroethylene soft-tissue patches in terms of the frequency of complications and postoperative outcomes. Typically, the surgeon's preference is an important factor in choosing a prosthetic material [7]. In the present case, complete resection with a pathologically negative margin was achieved, and the radical resection of the chest wall required chest wall reconstruction.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

References

1. Kim JM, Yoon YH, Lee KH, Kim JH. *Malignant gastro-*

Extra-Gastrointestinal Stromal Tumor Presenting as an Anterior Chest Wall Mass

- intestinal stromal tumour in the posterior mediastinum.* Interact Cardiovasc Thorac Surg 2012;14:497-9.
- Alkhatib L, Albtoush O, Bataineh N, Gharaibeh K, Matalaka I, Tokuda Y. *Extragastrointestinal stromal tumor (EGIST) in the abdominal wall: case report and literature review.* Int J Surg Case Rep 2011;2:253-5.
 - Miettinen M, El-Rifai W, H L Sobin L, Lasota J. *Evaluation of malignancy and prognosis of gastrointestinal stromal tumors: a review.* Hum Pathol 2002;33:478-83.
 - Lee JR, Anstadt MP, Khwaja S, Green LK. *Gastrointestinal stromal tumor of the posterior mediastinum.* Eur J Cardiothorac Surg 2002;22:1014-6.
 - Franzini C, Alessandri L, Pisciole I, et al. *Extra-gastrointestinal stromal tumor of the greater omentum: report of a case and review of the literature.* World J Surg Oncol 2008;6:25.
 - Zhang W, Peng Z, Xu L. *Extragastrointestinal stromal tumor arising in the rectovaginal septum: report of an unusual case with literature review.* Gynecol Oncol 2009; 113:399-401.
 - Mansour KA, Thourani VH, Losken A, et al. *Chest wall resections and reconstruction: a 25-year experience.* Ann Thorac Surg 2002;73:1720-5.