A 76-year-old woman with hypertension presented with a 3-day history of progressive dyspnea. Physical examination revealed decreased breathing sounds in her right chest. Her routine blood investigations were unremarkable; cytology evaluation of the pleural fluid revealed no malignant cell. Chest radiography demonstrated right massive pleural effusion and a mass lesion disclosed over right heart border after adequate drainage and pigtail insertion. Computed tomography (CT) demonstrated a 11.4 x 8.4 x 13 cm well-defined heterogeneous mass with cystic and nodular areas encasing the right heart border with no mediastinal lymphadenopathy (Figure 1a). No metastatic lesion was detected on CT of abdomen.

CT-guided needle biopsy demonstrated low-grade leiomyosarcoma. Immunohistochemically, the tumor cells showed strong diffusion positive for smooth muscle actin, and negative for CD117 and S100 (Figure 1b).

Soft tissue sarcomas comprise 0.7% of adult malignancies and 2% of all tumors that occur in the mediastinum which most frequently involved the uterus, gastrointestinal tract, retroperitoneum and the vascular system. Mediastinal leiomyosarcomas (LMS) are rare neoplasms of smooth muscle cells that account for about 1.4% of all soft tissue sarcomas and for approximately 11% of primary mediastinal sarcomas. Anterior mediastinal, low-grade LMS, pedunculated from the soft tissue, are extremely rare. Histological typing is very important in determining the type and extent of therapy. Despite an aggressive surgical approach, as well as the use of radiation therapy and/or chemotherapy, the local recurrence rate and distant metastasis rates are high. Mediastinal LMS has a dismal prognosis despite multimodality treatment.

Photographs and text from: C.-Y. Liao, Internal Medicine Department, Kaohsiung Armed Forces General Hospital, Zhongzheng 1st Rd., Lingya Dist., Kaohsiung City, Taiwan; S.-T. Chien, Pathology Department, Kaohsiung Armed Forces General Hospital, Zhongzheng 1st Rd., Lingya Dist., Kaohsiung City, Taiwan; S.-F. Huan, Internal Medicine Department, Kaohsiung Armed Forces General Hospital, Zhongzheng 1st Rd., Lingya Dist., Kaohsiung City, Taiwan; S.-F. Huan, Internal Medicine Department, Kaohsiung Armed Forces General Hospital, Zhongzheng 1st Rd., Lingya Dist., Kaohsiung City, Taiwan.
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