INTRODUCTION

Thymic neuroendocrine carcinoma (NEC), also known as thymic carcinoid tumors, are uncommon thymic neoplasms with neuroendocrine differentiation. Among primary thymic malignancies, NEC is the least common, accounting for less than 5% of thymic tumors (1). Thymic NEC tends to be locally invasive towards adjacent structures, such as mediastinal fat, lung, pericardium, and great vessels (2). Metastatic spread can occur by either hematogenous or lymphatic routes, and mediastinal lymph node metastasis is found in about 30 percent of patients at presentation (3). Although thymic NEC is typically identified as a large, usually invasive, anterior mediastinal mass with hemorrhage and necrosis (4, 5), it is extremely rare for it to present rupture. Herein, we report a case of the thymic NEC that was initially presented as a potentially fatal hemorrhage by capsular rupture.

CASE REPORT

A 70-year-old female, with no underlying condition except for spinal stenosis, presented at an emergency room because of acute chest discomfort for a day, which on admission, was aggravated with deep inspiration and progressed to chest pain. Initial laboratory studies revealed mild leukocytosis (10.6 × 10³/µL), decreased hemoglobin concentration (10.4 g/dL), and elevated D-dimer (2.83 µg/mL). Cardiac markers and coagulation studies were unremarkable. The blood pressure was 140/90 mm Hg and the pulse was 112/min. The initial chest radiograph showed mediastinal widening with right cardiac border bulging (Fig. 1A). A retrospective review of the patient’s serial chest radiographs, performed for routine health checkups one, two, and four years ago, revealed a gradual progression of right cardiac border bulging. However, an abrupt size increment was identified in a recent checkup.

A triphasic chest CT scan was performed for further work-
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up. A very large right anterior mediastinal mass, abutting the right anterior chest wall and right cardiac border measuring up to 11 cm, was identified on the non-enhanced CT scan. The tumor consisted of a large high-density portion and a small low density portion, suggesting hemorrhage and necrosis, respectively. The high-density portion showed higher attenuation than the combined peritumoral hemorrhage (hemothorax and hemomediastinum). There was no enhancement of the tumor in the pulmonary arterial phase (Fig. 1B); however, there was a mild and heterogeneous enhancement in the delayed phase at the bottom portion of it. The enhanced portion was adjacent to the low-density area of the tumor and was irregular in shape (Fig. 1C). Although the hemothorax and hemomediastinum hindered evaluation, CT yielded no evidence of pleural, pericardial, focal lung lesions, and pulmonary embolism. The bilateral adrenal glands were unremarkable. There was no evidence of metastasis.

As the initial impression of clinician was spontaneous mediastinal hematoma including aortic rupture, the patient was admitted to the intensive care unit for continuous monitoring. The amount of right hemothorax gradually increased on the follow-up chest radiograph and CT that was performed the next day. The follow-up hemoglobin concentration fell to 6.2 g/dL. A partial, heterogeneous enhancement of the tumor was still identified only in the delayed phase CT scan. As progressive intrathoracic hemorrhage was suspected clinically and radiologically, urgent surgery was performed.

After a posterolateral incision through the 5th intercostal space

Fig. 1. A 70-year-old woman with ruptured thymic neuroendocrine carcinoma (NEC).
A. Initial chest radiograph reveals abrupt increment of right cardiac border bulging with mediastinal widening, compared with previous radiograph (not shown).
B. Pulmonary arterial phase of initial triphasic chest CT. There is a large heterogeneous anterior mediastinal mass associated with hemomediastinum (arrow) and bilateral hemothorax. Note that there is no demonstrable enhancement within the tumor.
C. In the delayed phase of the initial triphasic chest CT, an irregular enhancement adjacent to the low-density portion of the tumor is noted.
D. The cut surface of the tumor shows diffuse hemorrhagic necrosis with foci of viable soft tissue adjacent to the rupture site (thick arrow).
E. Histopathological examination for the viable portion of the tumor (hematoxylin and eosin, × 400) shows cells with rounded nuclei and salt-and-pepper chromatin, which are compatible with NEC. A few mitoses are also identified (thin arrow).
and draining 3000 mL of hemothorax, the mass attached to the right upper mediastinum was detected with bleeding through the ruptured capsule. The possibility of a malignant thymic mass was raised during an intraoperative frozen section biopsy and the thymic tissue was maximally resected.

The gross specimen revealed a red-tan mass with a ruptured and ragged capsule (Fig. 1D). The cut surface showed diffuse hemorrhagic necrosis with foci of viable soft tissue adjacent to the ruptured capsule. Most of the hemorrhage was considered to be recent, as there was no hemosiderin-laden macrophage, as seen on the magnification view. Diffuse expression of chromogranin and CD56 was identified by means of an immunohistochemical stain. The final histopathologic diagnosis was well-differentiated thymic NEC, atypical carcinoid type (Fig. 1E).

The patient presented uneventful postoperative recovery and was discharged two weeks after the surgery. Adjuvant chemotherapy is planned.

**DISCUSSION**

In a study of 342 cases of mediastinal or thymic NECs, Soga et al. (6) report that symptomatic cases account for 63% of the total amount of cases and the cases with tumors over 50 mm account for 84%. Based on these data, Katsura et al. (7) suggested that most of the symptoms of thymic NECs (e.g., dyspnea, cough, chest pain, and hypertension) are caused by mass effect. Although several reports are available about other mediastinal tumors, such as teratomas, initially presented with tumor rupture, it is extremely uncommon for thymic NECs to initially present symptoms caused by rupture. To the best of our knowledge, this is the third report of a ruptured thymic NEC, following the two previous cases in the Japanese literature and the first report in the English literature.

The case reported by Katsura et al. (7) includes two CT scans performed with a 10-day interval that revealed a tumor size decrement from 4 cm to 2 cm and the disappearance of the hemothorax and hemomediastinum. In that case, the enhancement pattern of the tumor changed from poor enhancement, probably because of intratumoral hemorrhage, to diffuse heterogeneous enhancement. This may be explained by the small tumor size. The hemorrhage stopped for some reason, perhaps the blood clot might have sealed off the small rupture site, and hence the hemothorax and hemomediastinum may have been absorbed. The heterogeneous enhancement because of internal hemorrhage and necrosis is known as a typical CT finding of thymic NECs (4, 5), thus the presence of the heterogeneous enhancement on the later scan in the case reported by Katsura et al. (7) may be because of the viable tumor portion. By contrast, in this case the amount of hemothorax and hemomediastinum increased within two days and active bleeding through the ruptured capsule was identified at the operating field. The size of the tumor in the current case was much larger and the capsular rupture could probably have not been sealed off. The enhancement of the tumor in this case was partial, mild, and limited in the delayed phase. Most of the tumor did not show enhancement. The viable portion of the surgical specimen was focal and most of the tumor showed hemorrhagic necrosis. Hence, the enhancement of the tumor in this case is more likely to be intratumoral active venous bleeding, rather than the enhancement of the viable portion. However, considering the report about the ruptured thymoma by Santo-prete et al. (8), extratumoral veins, such as subcapsular veins or veins from thinned overlying mediastinal pleura, should be considered as alternative bleeding sites, as the study in this case was inconclusive with regard to the origin of the hemorrhage.

The cause of the spontaneous rupture of a mediastinal tumor still remains unclear. Ellison et al. (9) suggested several mechanisms of spontaneous bleeding from a mediastinal tumor, including rapid lesion growth, transient increase of intrathoracic pressure, abrupt rise in blood pressure, and altered hemostasis. Choi et al. (10) suggested that the tendency of rupture for mediastinal teratoma can be because of tumor enlargement and consequent ischemia and necrosis. Katsura et al. (7) observed internal cystic change of thymic NEC which was caused by hemorrhage and necrosis. Those authors also assumed that it weakens the pseudocapsule of the tumor and that the subsequent rupture occurs with tumor size increment (7). In the case of this study, the serial chest radiographs suggest a gradual tumor size increment for a few years. Intratumoral hemorrhage and necrosis might have progressed. We assume that the hemorrhage, which cannot be stopped, occurred at a certain moment. Although a marked difference was observed for a year interval, as compared with serial plain radiographs, the tumor enlargement may have been more abrupt, because there was no hemosiderin-laden macrophage on the magnification view. As pre-
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SUMMARY

This case describes a rare instance of thymic neuroendocrine carcinoma (NEC) with spontaneous rupture, leading to hemothorax and hemomediastinum. The tumor was accompanied by heterogeneous enhancement, internal necrosis, and hematoma. CT imaging was crucial in identifying this unusual presentation of NEC.

REFERENCES