Sclerosing thymoma
A rare case report and brief review of literature
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Abstract
Rationale: Sclerosing thymoma is an extremely rare mediastinal neoplasm; it was recognized for the first time in 1994 and to date only 15 cases have been reported.

Patient concerns: The present study report a case of a 65-year-old man who was incidentally found to have an anterior mediastinal nodule, without clinical symptoms including fever, chest pain, and myasthenia gravis.

Diagnoses: The chest computed tomography (CT) revealed the nodule was 4.9 × 4.2 × 3.0 cm in size. And the microscopic and immunohistochemical findings indicated that the final diagnosis was sclerosing thymoma.

Interventions: The anterior mediastinal nodule was completely removed.

Outcomes: No evidence of recurrence or complication was found in the second year after surgery.

Lessons: The biologic behavior of the rare sclerosing thymoma is still largely mysterious; it is utmost important to classify the sclerosing thymoma from other mediastinal tumors. Its prognosis is favorable and thymectomy is currently the mainstay of treatment.

Abbreviations: CD1a = CD1a molecule, CT = computed tomography, Ki–67 = Antigen Ki-67, LCA = protein tyrosine phosphatase, receptor type C, P53 = p53 tumor suppressor homolog, P63 = hypothetical protein, PLAP = Phospholipase A2 activator protein, TDT = tonoplast dicarboxylate transporter, TTF-1 = transcription termination factor 1, VIM = vimentin, WHO = World Health Organization.

Keywords: mediastinum, sclerosing thymoma, thymoma

1. Introduction
The incidence of thymoma is 1.5 cases per million and thymoma is the most common primary neoplasm of the anterior mediastinum, consisted of 47% to 50% of anterior mediastinal tumors.[1–3] In 2004 the World Health Organization (WHO) classified thymoma into 5 subtypes: A, AB, B1, B2, and B3.[4] Even though the majority of thymomas express conventional morphologic and histologic characteristics, some rare variants with unusual features such as microscopic thymoma, thymolipoma, lipofibroadenoma, thymofibrolipoma, and sclerosing thymoma were reported.[5–10] Sclerosing thymoma was first reported by Kuo in 1994,[9] and to our knowledge, only 15 cases have been reported in literatures till now. Herein, we report a case of sclerosing thymoma in a 65-year-old man and review the different data from the literature.

2. Case presentation
The study was approved by the Institutional Ethics Committees of Tianjin Chest Hospital and conducted in accordance with the ethical guidelines of the Declaration of Helsinki. A written informed consent was obtained from the patient for publication of this case report and any accompanying images.

A 65-year-old man was admitted to Department of Thoracic Surgery, Tianjin Chest Hospital (Tianjin, China) on December 28, 2015 due to anterior mediastinal nodule incidentally found on chest computed tomography (CT) taken 3 months ago because of trauma. He had no fever, no shortness of breath and chest pain, no weight lose, no myasthenia gravis, and laboratory tests revealed no abnormality. No therapy was taken before admitted. A well-circumscribed anterior mediastinal nodule, measuring 49 × 42 × 30 mm was identified in CT (Fig. 1). The patient underwent complete thymectomy and discharged on January 7, 2016. At the second year postsurgery follow-up appointment, the patient was alive with no evidence of recurrence or complication.

The surgically resected specimen was well circumscribed, and part of the capsule was invaded. The cut section was homogeneously tan-white, with some cystic lesion. In some areas, cellular proliferation was active. Adipose tissues, imperfect degradation of thymus tissues and tumor components were visible. Immunohistochemistry results were presented as follows: positivity for Keratin (broad-spectrum), P63 (+) and Ki-67 (+20%), negative for TTF-1, P53, PLAP, TDT, LCA, CD1a, and VIM. The diagnosis was confirmed by Masson trichrome stain.

3. Summary of the literature
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There were 6 female and 9 male with the age ranging from 10 to 73 years old. The largest diameter of the lesions was 2 to 10 cm. 53.33% (8/15) of the patients were asymptomatic and discovered incidentally when taken routine chest X-ray or CT scans for unrelated reasons, 7 cases presented mild symptoms such as shortness of breath, chest pain or muscle weakness, and difficulty in talking. Myasthenia gravis was seen in only 3 cases (20%). Although preoperative needle biopsy was performed in 6 of the 15 cases, correct diagnosis was not obtained. Thymectomy was taken in all patients, and the postoperative was well. Follow-up information was obtained in 11 of those patients, 4 patients were alive and well for 1 to 6 years after surgical resection of the thymoma, 6 patients died of unrelated diseases, including congestive heart failure, pulmonary edema, and renal insufficiency.

4. Discussion

Sclerosing thymoma is a subtype of thymoma, and accounts for <1% of thymoma.[11] The preoperative clinical symptoms of sclerosing thymoma are varied.[10] Due to the limited number of the cases, the biologic behavior of the rare sclerosing thymoma is still largely mysterious. Over half of the patients were admitted with shortness of breath, chest pain ascribe to mass effect. Other patients may present with complicated autoimmune disease such as myasthenia gravis. In addition, some were found by CT or X-ray incidentally because the patients were asymptomatic. The lesions were usually well circumscribed and encapsulated in CT scan. The cut surface of those tumors was light tan and solid, with no hemorrhage or necrosis, calcifications, and a fat plane often could be found. Histologically, an extensive area of hyalinized and sclerotic fibrocollagen, which constituted about 85% of the tumor, was common in sclerosing thymoma. However, those factors are not unique characteristics of sclerosing thymoma; caution may be warranted before an inappropriate diagnosis of other neoplastic as well as nonneoplastic lesions, such as solitary fibrous tumor,[12] sclerosing mediastinitis, lymphoma with sclerosis, and solitary fibrous tumor of the mediastinum[13] that may occur in the anterior mediastinum, is made. Sclerosing thymoma is a variant of thymoma characterized by abundant hyalinized fibrocollagenous stroma and chronic inflammatory cells, such as plasma cells and lymphocytes,[14] whereas solitary fibrous tumor was characterized by typically patternless or storiform spindle-shaped cell morphology (minimal nuclear pleomorphism and rare or absent mitoses), and electron microscopical examination reveals features of both fibroblasts and mesothelial cells. Spindle cells could not be found in sclerosing mediastinitis. In the tumor tissue morphology of lymphoma with sclerosis, cancer cells were consisted of a lot of lymphoid cells, and without thymoma bipolar cells.

At present the most effective treatment for sclerosing thymomas is complete surgical removal of the tumor. The clinical symptoms of those patients reported were improved after the resection of the mass. No evidence of recurrence or metastasis was reported.

5. Conclusion

In conclusion, this is the 6th study of sclerosing thymoma and the 16th reported case. Its prognosis is favorable and thymectomy is currently the mainstay of treatment. Owing to the varied symptoms, it is utmost importance to classify the sclerosing thymoma from other mediastinal tumors.

| Author     | Sex   | Age, y | Size, cm | Clinical symptom                      | Myasthenia gravis | Outcome               |
|------------|-------|--------|----------|--------------------------------------|-------------------|-----------------------|
| Kuo[9]     | Female| 39     | 3        | Palpitation and dyspnea, muscle weakness and difficulty in talking | Yes               | Alive and well, 4 y   |
| Kuo[9]     | Female| 23     | 2.5      | None                                 | Yes               | Alive and well, 2 y   |
| Kim et al[15] | Male  | 47     | 2        | None                                 | None              | Alive and well, 1 y   |
| Moran et al[10] | Female| 34     | 5        | None                                 | None              | Died, congestive heart failure |
| Moran et al[10] | Male  | 58     | 6        | None                                 | None              | Died, congestive heart failure |
| Moran et al[10] | Male  | 44     | 5        | None                                 | None              | Lost to follow-up     |
| Moran et al[10] | Male  | 56     | 10       | None                                 | None              | Lost to follow-up     |
| Moran et al[10] | Female| 62     | 8        | None                                 | None              | Alive and well, 6 y   |
| Moran et al[10] | Female| 37     | 6        | Shortness of breath, chest pain       | None              | Died, pulmonary edema |
| Moran et al[10] | Male  | 69     | 7        | Shortness of breath, chest pain       | None              | Died, renal dysfunction |
| Moran et al[10] | Male  | 59     | 6        | Shortness of breath, chest pain       | None              | Died, congestive heart failure |
| Moran et al[10] | Female| 27     | 5        | None                                 | Yes               | Died, unknown         |
| Moran et al[10] | Male  | 73     | 10       | Shortness of breath, chest pain       | None              | Died, unknown         |
| Tajima et al[16] | Male  | 62     | 3.1      | None                                 | None              | —                     |
| Yabo et al[17] | Male  | 10     | 7        | Chest pain                           | None              | —                     |
Author contributions
Data curation: Xin Li.
Writing – original draft: Xin Li.
Writing – review & editing: Meng Wang, Daqiang Sun.
Conceptualization: Daqiang Sun.

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