

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 importance 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 were 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

Two cases of sclerosing thymoma were first reported by Kuo in 1994, and till now only 15 cases were described in the literature

Editor: N/A.

The authors have no conflicts of interest to disclose.

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Medicine (2018) 97:16(e0520)

Received: 24 January 2018 / Accepted: 30 March 2018

<http://dx.doi.org/10.1097/MD.00000000000010520>



Figure 1. Computed tomography scan. A well-circumscribed anterior mediastinal nodule, measuring 49 × 42 × 30 mm was identified.

(Table 1). 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.

Table 1
Summary of case reports of sclerosing thymoma.

Author	Sex	Age, y	Size, cm	Clinical symptom	Myasthenia gravis	Outcome
Kuo ^[9]	Female	39	3	Palpitation and dyspnea,	Yes	Alive and well, 4 y
Kuo ^[9]	Female	23	2.5	muscle weakness and difficulty in talking	Yes	Alive and well, 2 y
Kim et al ^[15]	Male	47	2	None	None	—
Moran et al ^[10]	Female	34	5	None	None	Alive and well, 1 y
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

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Conceptualization: Daqiang Sun.

References

- [1] Ruffini E, Filosso PL, Mossetti C, et al. Thymoma: inter-relationships among World Health Organization histology, Masaoka staging and myasthenia gravis and their independent prognostic significance: a single-centre experience. *Eur J Cardiothorac Surg* 2011;40:146–53.
- [2] Liu GB, Qu YJ, Liao MY, et al. Relationship between computed tomography manifestations of thymic epithelial tumors and the WHO pathological classification. *Asian Pac J Cancer Prev* 2012;13:5581–5.
- [3] Marom EM, Miletto MA, Moran CA, et al. Computed tomography findings predicting invasiveness of thymoma. *J Thorac Oncol* 2011;6:1274–81.
- [4] Travis WD, Brambilla E, Müller-Hermelink HK. Thymomas. WHO Classification of Tumours: Pathology and Genetics of Tumours of the Lung, Pleura, Thymus and Heart. Lyon, France: IARC Press; 2004;146–71.
- [5] Cheuk W, Tsang WY, Chan JK. Microthymoma: definition of the entity and distinction from nodular hyperplasia of the thymic epithelium (so-called microscopic thymoma). *Am J Surg Pathol* 2005;29:415–9.
- [6] Moran CA, Rosado-de-Christenson M, Suster S. Thymolipoma: clinicopathologic review of 33 cases. *Mod Pathol* 1995;8:741–4.
- [7] Qu G, Yu G, Zhang Q, et al. Lipofibroadenoma of the thymus: a case report. *Diagn Pathol* 2013;8:117.
- [8] Tobias JR, Cullen JM. Thymofibrolipoma in a labrador retriever. *Vet Pathol* 2014;51:816–9.
- [9] Kuo T. Sclerosing thymoma—a possible phenomenon of regression. *Histopathology* 1994;25:289–91.
- [10] Moran CA, Suster S. “Ancient” (sclerosing) thymomas: a clinicopathologic study of 10 cases. *Am J Clin Pathol* 2004;121:867–71.
- [11] Moran CA, Suster S. On the histologic heterogeneity of thymic epithelial neoplasms. Impact of sampling in subtyping and classification of thymomas. *Am J Clin Pathol* 2000;114:760–6.
- [12] Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. *Cancer* 1981;47:2678–89.
- [13] Witkin GB, Rosai J. Solitary fibrous tumor of the mediastinum. A report of 14 cases. *Am J Surg Pathol* 1989;13:547–57.
- [14] Müller-Hermelink HK, Engel P, Kuo TT, et al. Tumours of the thymus. In: Travis WD, Brambilla E, Müller-Hermelink HK, et al., editors. *Pathology and Genetics of Tumours of the Lung, Pleura, Thymus and Heart*. Lyon, France: IARC Press; 2004;146–71.
- [15] Kim YH, Ishii G, Naito Y, et al. A resected case of sclerosing thymoma. *Nihon Kokyuki Gakkai Zasshi* 2006;44:420–3.
- [16] Tajima S, Koda K. A case report of sclerosing thymoma of the anterior mediastinum: an exceedingly rare morphology. *Int J Clin Exp Pathol* 2015;8:4233–77.
- [17] Wang YB, Pan TC, Wei X, et al. Sclerosing thymoma: a case report and literature review [in Chinese]. *Chin J Mis Diagn* 2008;8:8579–81.