A case of spindle cell primary intrapulmonary thymoma:
a rare histologic subtype

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Primary intrapulmonary thymomas are very rare. Among them, pure spindle cell primary intrapulmonary thymoma is ever rarer. A 47-year-old woman was found to have a mass located on the right side of the chest during routine preoperative tests for a goiter operation. She underwent exploratory thoracotomy in the right hemithorax, during which a well-defined mass in the right lower lobe, close to the pericardium was completely excised. Pathological evaluation revealed mixed type thymoma with predominance of spindle cells. The patient received adjuvant radiation therapy in the postoperative period. No recurrences were detected within a follow-up of two years.

Key words: Lung neoplasms/pathology/surgery; thoracotomy; thymoma/pathology/surgery.

Thymoma is an epithelial neoplasm of the thymus and usually presents as an anterior mediastinal tumor.1 Primary thymomas located outside the anterior mediastinum are rarely seen.2 Primary intrapulmonary thymomas are defined as thymomas arising in an intrapulmonary location without an associated mediastinal component and are very rare.3 A total of 23 cases have been reported in the literature since 1951.3 To the best of our knowledge, only two cases of pure spindle cell intrapulmonary thymoma have been described.4,5 We report another case of spindle cell intrapulmonary thymoma.

CASE REPORT

A 47-year-old woman was referred to our clinic by the general surgery department. A mass in the right hemithorax had been noticed on a chest roentgenogram during routine evaluation for a goiter operation. Her complaints were mainly due to a past hyperthyroid state and no respiratory symptoms were present. Her past medical history was unremarkable except for an episode of hyperthyroidism which was turned to euthyroid state by medical therapy. Physical examination was normal with the exception of a palpable thyroid gland. Computed tomography revealed a mass with a calcific wall at the right paracardiac localization, measuring 42 mm in diameter. No mediastinal or hilar adenopathies were present (Fig. 1). Pulmonary hydatid cystic disease was strongly suspected.

Exploratory thoracotomy was performed and a well-defined tumor adhering to the pericardium was noted in the right lower lobe (Fig. 2). The tumor was completely excised from the pulmonary parenchyma and the pericardium. The resected tumor was a well-circumscribed, solid, firm, and encapsulated mass. No invasion to the parenchymal tissue beyond the capsule was present.

Macroscopically, the specimen from the right lower lobe measured 6x4.7x4.5 cm. Sectioning revealed a
firm, yellow-white, encapsulated, and lobulating nodule with cystic areas. Calcific degeneration was noted in areas adjacent to the capsule. On microscopic examination, it was an encapsulated tumor which was lobulated by thin fibrous bands formed by bundles of spindle cells intermingled with mature lymphocytes (Fig. 3a). In some areas, capsule invasion was noted (Fig. 3b). Mitotic figures were rare and no necrosis was noted. Immunohistochemical analysis was performed using the B-SA (biotin-strepavidin antibody) technique and Pan CK (clone AE1/AE3), EMA (clone GP 1.4), and LCA (clone Ab-3) neomarkers. The epithelial tumor cells displayed positivity for Pan CK and EMA. The lymphocytic component stained predominantly with LCA. The tumor was benign in terms of its cytological appearance; however, it might also have a malignant course due to capsule invasion. Thus, the tumor was diagnosed as spindle cell predominating mixed type thymoma and type AB in the WHO classification. According to the Masaoka system of staging, the tumor was at stage 2 due to its microscopic invasion into the capsule.

Adjuvant radiation was administered postoperatively and the patient’s postoperative course was uneventful. There was no evidence for recurrence within two years.

DISCUSSION

Intrapulmonary thymomas are classified as either central or peripheral with respect to localization. They may develop in the hilus of the lung, in relation to the wall of a major bronchus or attached to the pericardium, or may be peripheral in the lung localized beneath the visceral pleura. In our case, the tumor was located centrally and was attached to the pericardium.

When a thymoma develops at unusual sites such as intrapulmonary or pleural regions, it may be difficult to distinguish from other epithelial neoplasms such as lung carcinoma or mesothelioma. The differential diagnosis of intrapulmonary thymoma includes a wide range of lesions. Lymphoid-predominant lesions may be confused with thoracic lymphoid lesions, while epidermoid-predominant lesions may be confused with metastasis to the lung. In the present case, pulmonary hydatid cystic disease was strongly suspected in the preoperative period.

Histologic features of thymomas present as a mixture of epithelial cells and lymphocytes in varying proportions, and classified as lymphocyte-dominant, epithelial cell-dominant, or mixture type. Immunohistochemical staining combined with gross and microscopic pattern of the tumor and patient’s clinical history is helpful in differentiating a primary pulmonary thymoma from any other intrathoracic neoplasm. In our case, the tumor was diagnosed as spindle cell predominating mixed type thymoma, with type AB according to the WHO classification.

The morphologic spectrum of this lesion encompasses variations in location, with either the epithelial or lymphocytic component being predominant in most instances. However, only two cases of intrapulmonary spindle cell thymoma have been reported.

Thymomas grow slowly and remain asymptomatic until they reach a size sufficient to cause problems associated with local growth, which include pain or bronchial obstruction. Independent of local size, the tumor has a good prognosis if it is well circumscribed, encapsulated, and completely resected. James et al. reported that, like ordinary mediastinal thymomas, the prognosis might be determined principally by the pres-
ence of invasion and, to a lesser degree, by the histological subtype. Verley and Hollmann reported that the presence of invasion and histological subtype were important prognostic features. Spindle cell tumors have a better prognosis than lymphoepithelial tumors and obviously malignant tumors. In our case, the tumor was encapsulated and no invasion to the lung tissue beyond the capsule was present. We were able to excise the tumor completely and the prognosis was favorable during the postoperative two years.

Despite limited experience with these lesions, surgical resection has been curative in most instances. As with mediastinal thymic tumors, adjuvant radiation therapy should be considered if complete resection is not possible or extension of thymic tissue beyond a circumscribed capsule is noted. Ishibashi et al. reported a case of primary intrapulmonary thymoma with nodal metastasis and concluded that systematic mediastinal lymph node dissection according to the lymph node map for primary lung cancer should be considered. Since gross appearance of the mass closely resembled pulmonary hydatid cystic disease, following excision of the mass the rest of the operation was carried out as with routine hydatid cyst excision. Intrapulmonary lesions that cause strong suspicion of a pulmonary hydatid cyst in the preoperative period and have a macroscopic appearance of a hydatid cyst at surgery can rarely be malignant lesions. During the operation of intrapulmonary lesions, definitive diagnosis should be made with frozen section if possible. In addition,

Fig. 3. Histologic appearance of the specimen demonstrating (a) predominating spindle cells intermingled with mature lymphocytes (H-E x 100), and (b) fibrous capsule invasion of the thymoma (H-E x 40).
because of the absence of a definite diagnosis, a complete mediastinal lymph node sampling may be necessary even no preoperative or intraoperative evidence for lymphadenopathy has been obtained.

In conclusion, despite their rarity, intrapulmonary thymomas should be kept in mind among possible causes of pulmonary nodules. It is advisable that frozen section examination be available and complete mediastinal lymph node sampling be made.

REFERENCES