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CASE REPORT

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An invasive primary thymoma: a rare cause of intrapulmonary mass

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Summary

Background: Primary intrapulmonary thymomas are extremely rare tumors and could be mistaken for a variety of benign and malignant epithelial or mesenchymal lesions.

Case report: A case of a primary invasive intrapulmonary thymoma manifested with massive hemoptysis is presented. CT demonstrated a mass with specks of calcification, located within the upper lobe of the right lung. The lesion showed heterogeneous enhancement with areas of cystic and necrotic degeneration and hemorrhagic foci. CT differential diagnosis included the possibility of invasive thymoma, which was confirmed by surgery and histopathology.

Conclusions: Though a rare entity, intrapulmonary thymoma, because of its potentially invasive and malignant nature, should be included in the radiological differential diagnosis of intrapulmonary masses. This is important, since it can determine further management.

Key words: Thymus • ectopic • pulmonary mass • CT

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Background

Primary intrapulmonary thymomas (PIT) are extremely rare tumors at this location, which could be mistaken for a variety of benign and malignant epithelial or mesenchymal lesions [1]. Because of the potentially invasive nature of the tumor, its recognition is important for further management.

We present a case of a primary invasive intrapulmonary thymoma, which manifested clinically with massive hemoptysis.

The presentation of this case is justified by its extreme rarity, difficulty in establishing radiological and histological diagnosis and the fact that all cases known to us were published in non-radiological literature.

Case Report

A 58-year-old man was admitted with severe right-sided chest pain and hemoptysis. Blood pressure, pulse rate and temperature were normal. The physical examination revealed decreased air entry in the right lung with dull percussion.

Chest X-ray showed a large ill-defined opacity in the right hemithorax, obliterating the right cardiac border with no shift of the mediastinum and right-sided pleural effusion (Fig. 1).

Un-enhanced CT scans demonstrated a large lobulated mass with specks of calcification, partially surrounded by fat, located within the upper lobe of the right lung and abutting the superior vena cava, upper lobe vessels

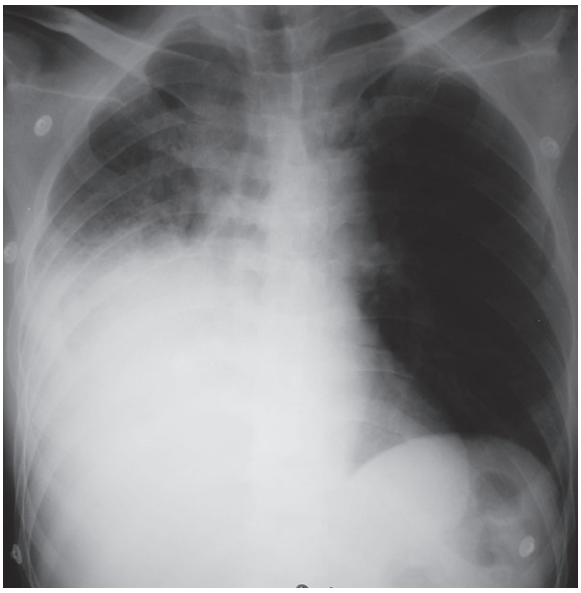

Figure 1. Portable chest X-ray: a large ill-defined opacity in the right hemithorax obliterating the right cardiac border. Right-sided pleural effusion.

and the right atrium (Fig. 2 A). Contrast-enhanced scans showed heterogeneous enhancement of the lesion with a few small low attenuation areas indicating cystic and necrotic degeneration (Fig. 2 B). Pathological vessels and hemorrhagic foci were seen within the mass (Fig. 2 C). The adjacent lung showed patchy alveolar opacification. There was also a mild right-sided pleural effusion. There was no evidence of hilar and mediastinal lymphadenopathy. In addition to other lung tumors, a possibility of invasive thymoma was included in differential CT diagnosis.

Bronchoscopy revealed extensive bleeding and clot formation in the upper lobe bronchus.

The patient refused an open biopsy and operation and he was discharged. However, another episode of massive hemoptysis with hemoglobin drop occurred and the patient was re-hospitalized. Urgent thoracotomy revealed that the right pleural cavity was full of blood and clots but without pleural metastases. A large grayish fleshy tumor occupying the antero- inferior part of the upper lobe of the right lung was found. The tumor contained a large hematoma, which had ruptured into the pleural cavity. Classic upper lobectomy was carried out.

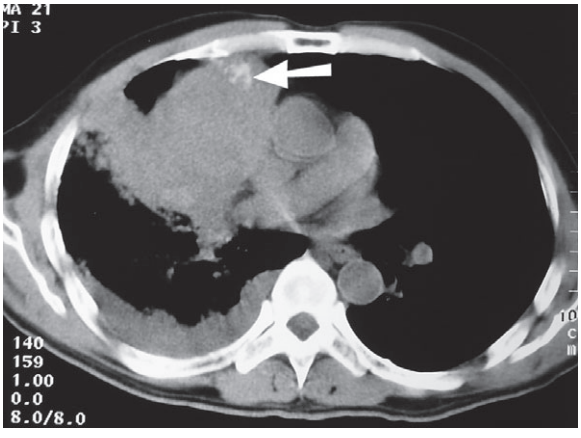

Figure 2A. Un-enhanced CT: a large mass of soft tissue density with specks of calcification (arrow), located within the upper lobe of the right lung. Right-sided pleural effusion.

Histopathological examination revealed a neoplasm growing in lobules, divided by fibrous bands. The cells were mostly epithelial with background of a few lymphocytes. A panel of immunohistochemical stains showed that the tumor cells were positive for pancytokeratin and focally for EMA and CD56 but negative for TTF-1, chromogranin and synaptophysin. The light microscopic morphology and immunophenotype were found to be consistent with those of intrapulmonary thymoma.

The post-operative course was uneventful.

Discussion

PIT are very rare tumors that may arise in the lung parenchyma without associated mediastinal involvement [2]. A total of 23 cases have been reported to date; all of them in non-radiological literature [1, 2].

There have been attempts to classify PIT into two groups: central and peripheral type.

The central tumors arise from the hilum of the lung or are occasionally attached to the pericardium [1, 2]. Peripheral tumors are usually found in the right lung [1]. Males and females are equally affected within wide range of age, from 19 to 79 years. Most of the cases were asymptomatic and incidentally detected on radiographic examination performed for unrelated causes. The symptomatic cases presented with fatigue, dyspnea, hemoptysis, chest pain or cramping [1]. The size of PIT in these various studies ranged from 1.5 to 12 cm [1].

A few hypotheses tried to explain the ectopic location of thymomas on the basis of embryology [1, 3]. The thymus is

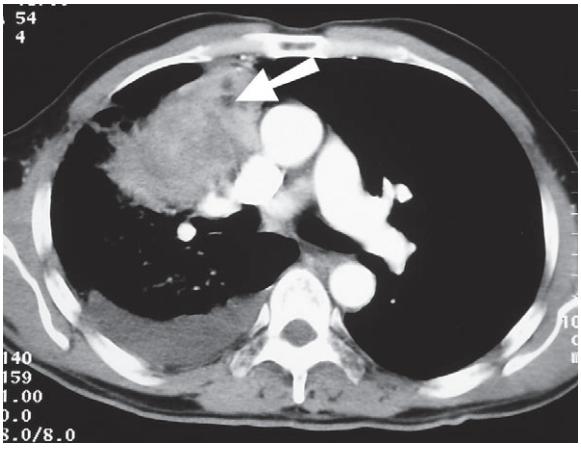

Figure 2B. Contrast-enhanced CT: heterogeneous enhancement of the mass with a few small low attenuation foci indicating cystic and necrotic degeneration (arrow). The mass is abutting the superior vena cava and upper lobe vessels.

derived from an outpouching of the 3rd and to minor extent the 4th pharyngeal pouch that appears late in the 6th week of gestation. From each side of the pouch, thymus grows caudally as a solid diverticulum into the mesoderm anterior to the aortic arch [4].

The site of the remnant tissue can be observed anywhere along the embryological descent path from the mandibular area down to the mediastinum. It can be adjacent to the thyroid tissue [4, 5], parotid gland [6], near the carotid bifurcation [7], the superior, middle mediastinum and rarely within the posterior mediastinum and the lung [1, 2, 8, 9].

A widely accepted explanation is that neoplastic transformation of the ectopic thymus tissue occurs as a secondary phenomenon to the aberrant descent of thymic tissue from the third or fourth branchial arch [10, 11]. However, this hypothesis fails to rationalize the presence of intrapulmonary and posterior mediastinal thymomas. An alternative explanation is that a solitary metastasis from an undetected small primary tumor in the mediastinum has occurred [3]. Another hypothetic possibility is a derivation from an uncommitted stem cell that differentiates along a specific pathway [1].

In our case report, we present a symptomatic invasive PIT manifested with massive hemoptysis that led to urgent surgical intervention. Despite the fact that in this case radiological examination did not provide final diagnosis, the CT features were characteristic enough to suggest the possibility of thymoma. Furthermore, the surgery and macroscopic examinations confirmed that the tumor had originated from the lung. Similarly to other reported cases, the histopathological diagnosis was extremely difficult and

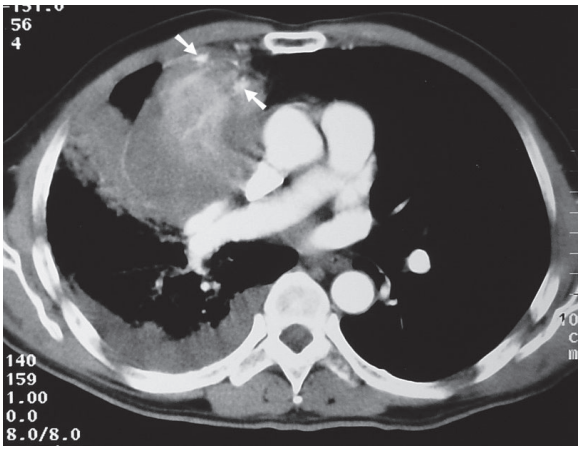

Figure 2C. Contrast-enhanced CT: Pathological vessels and contrasting hemorrhagic foci (small arrows) are seen within the mass.

included a range of possibilities, e.g. endocrine carcinoma, atypical peripheral carcinoid, sclerosing hemangioma and intrapulmonary thymoma. The latter possibility was finally confirmed by the use of a specific immunohistochemistry panel.

Despite the fact of extremely rare intrapulmonary location of thymoma and in view of its potentially invasive and malignant nature, the possibility of thymoma should be included in the radiological differential diagnosis of an intrapulmonary mass, especially when the tumor shows features suggestive of thymoma. These include partial or complete covering of the tumor by fat, cystic, necrotic or hemorrhagic components and calcifications [12, 13].

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