INTRODUCTION

Pulmonary hamartoma is the third most frequent cause of solitary pulmonary nodules with an overall prevalence of up to 5.7% (1), and chondromatous hamartoma is the most common type of pulmonary hamartoma (2). Masses are composed of abnormal mixtures of epithelial and mesenchymal elements, such as cartilage, fat, fibromyxoid tissue, smooth muscle, and bone (3, 4).

In spite of its relative frequency, no report has been published that details the CT findings of pulmonary hamartomas with rare pathologic patterns, such as fibrous or fibroleiomyomatous types. Here, we describe, for the first time, the unusual contrast-enhanced CT findings of pulmonary hamartoma with a predominant fibroblast component and their relations with histopathologic findings. This case report was approved by our Institutional Review Board.

CASE REPORT

A 20-year-old male was referred to our hospital for exertional dyspnea of two weeks’ duration. Clinical examination and routine laboratory test results were unremarkable, but chest radiography revealed a lobulated mass in the right lower lung field (Fig. 1). Non-enhanced CT showed a 3.4 cm lobulated mass without evidence of fatty deposits or calcification. The mass was positioned near the medium bronchus in the posterior basal segment of the right lower lobe. It had a large extrabronchial component and a small endobronchial component, that is, a 'tip of
Fig. 1. 20-year-old male with pulmonary fibroblastic hamartoma.

A. Chest radiograph demonstrating the presence of a lobulated mass in the right lower lung field.

B. Axial contrast-enhanced chest CT image with mediastinal setting (left) and oblique two-dimensional coronal reformatted image with lung setting (right) showing a 3.4 cm mass in the posterior basal segment of the right lower lobe. The mass consists of small endobronchial (arrows) and large extrabronchial components, suggestive of the ‘tip of the iceberg’ sign.

C. Dynamic contrast-enhanced CT image showing persistent enhancement with a net enhancement of 47 Hounsfield units (HU). Non-contrast CT provides no evidence of fatty deposits or calcification. The CT HU measurements are 17 HU pre-contrast, 21 HU at 0.5 minute, 32 HU at 1 minute, 41 HU at 1.5 minutes, 44 HU at 2 minutes, 51 HU at 4 minutes, 57 HU at 5 minutes, and 64 HU at 15 minutes.

D. Hematoxylin and eosin (H&E) staining showes that the tumor was composed predominantly of spindle cells, adipose tissue, and blood vessels. In addition, normal-looking pulmonary alveoli and bronchi are seen inside the tumor (× 40).

E. Spindle cells has elongated nuclei with inconspicuous nucleoli and are arranged in short fascicles in a collagenous background (H&E staining, original magnification × 200).
Unusual CT Findings of Pulmonary Hamartoma

the iceberg’ appearance, on contrast-enhanced CT (Fig. 2). Dynamic contrast-enhanced CT demonstrated a heterogeneous, persistently enhancing mass (Fig. 3). The CT Hounsfield unit (HU) measurements were: 17 HU pre-contrast, 21 HU at 0.5 minute, 32 HU at 1 minute, 41 HU at 1.5 minutes, 44 HU at 2 minutes, 51 HU at 4 minutes, and 64 HU at 15 minutes. Therefore, the net enhancement of the mass was 47 HU. Positron emission tomography and magnetic resonance imaging were not performed. At this time, the differential diagnosis was inflammatory myofibroblastic tumor or carcinoid tumor, because of the relationship between the mass and the airway and its enhancement pattern.

Percutaneous needle aspiration biopsy was not performed because the mass was relatively large and symptomatic and because this technique has a low negative predictive value (about 80%) (5). The mass was excised by right lower lobectomy via video-assisted thoracoscopic surgery. The gross pathologic specimen thereby obtained appeared as an ill-defined, white, solid endobronchial mass measuring 3.0 × 2.5 cm with exophytic growth. Hematoxylin and eosin staining revealed that the tumor was composed of mostly of spindle cells, adipose tissue, blood vessels, and alveoli (Figs. 4, 5). Immunohistochemistry showed that the spindle cells were negative for S-100 protein, desmin, and smooth muscle actin, so they were thus identified as fibroblasts. Angiomyolipoma was considered because the tumor contained spindle cells, blood vessels, and adipose tissue, but negative immunostaining for HMB45 enabled us to exclude a diagnosis of angiomyolipoma or other clear cell tumors. Accordingly, a diagnosis of pulmonary hamartoma with a predominant fibroblast component was made. Five months after surgical resection, the patient was well and the condition had not recurred.

DISCUSSION

Pulmonary hamartomas contain disorganized mixtures of various connective and epithelial tissues normally found in lung (6). Hamartomas are divided into subtypes based on their main component; chondromatous hamartoma is the most common (2). In our case, fibroblasts and fibrous stroma predominated, and thus, the mass was of the fibroblastic subtype. To the best of our knowledge, previous publication has reported the radiologic findings in cases of fibroblastic hamartoma.

Most pulmonary hamartomas are located peripherally in lungs, and only 1–19.5% are located endobronchially (2). Patients with endobronchial hamartoma can have respiratory complaints (7). Our patient had a mass containing mostly extrabronchial components with small endobronchial components, thus exhibiting the ‘tip of the iceberg’ sign on CT images, which has not been previously described for fibroblastic hamartoma.

Hamartomas that contain calcifications or fatty deposits can be diagnosed by CT in 62% of cases (8). In a report of 11 hamartomas (mean diameter, 1.5 cm; range, 1–3 cm), all masses exhibited thick capsular and septal enhancement and persistent enhancement without washout (4). However, our patient had a relatively large mass (diameter, 3.8 cm) without evidence of calcification or fatty deposits. Furthermore, although persistent enhancement was observed on dynamic CT, net enhancement was relatively high (47 HU). These abnormal findings prevented our making a diagnosis by CT.

According to a previous study, malignant nodules generally present a net enhancement of ≥ 25 HU and a washout of 5–31 HU. Also, benign nodules are characterized by one of the following: a net enhancement of < 25 HU, a net enhancement of ≥ 25 HU in combination with persistent enhancement, or a net enhancement of ≥ 25 HU in combination with a washout of > 31 HU (9). Considering that the mass showed a net enhancement of 47 HU without washout, dynamic contrast-enhanced CT could have been useful for identifying a benign nodule in our case. We considered that the fibrous stroma of the tumor may have contributed to the persistent enhancement observed in our case, and the delayed contrast enhancement might have been the result of contrast redistribution. Typically, after wash-in of a well-vascularized tumor cell zone during the early phase, contrast medium moves into fibrous stroma, which has scant blood supply, in the late phase, where it remains for a considerable time (4). Thus, it seems that the persistent enhancement pattern observed for many benign nodules is related to the amount and degree of fibrosis tissue (4).

We present the CT findings of pulmonary hamartoma with a predominantly fibroblastic component in a 20-year-old male. Awareness of its imaging features, which include persistent enhancement and ‘the tip of the iceberg sign’ on CT images, may substantially aid in the future diagnosis of this disease.
REFERENCES


비전형적 폐과오종의 역동성 전산화단층촬영 소견과 그 병리학적 연관성: 증례 보고

홍승백1·정연주1·이지원1·김영대2·안효영2·이창훈3·김아롱3·이지원1 *

저자들은 드문 병리학적 결과의 비전형적인 폐과오종을 갖는 20세 남자 환자의 역동성 조영증강 전산화단층촬영 소견을 보고하고자 한다. 전산화단층촬영에서 3.4 cm 크기의 “빙산의 일각” 소견(“tip of the iceberg” sign)을 보이는 소엽성 종괴가 우하엽 후기저분절의 중간 크기의 기관지에서 관찰되었다. 역동성 조영증강 전산화단층촬영에서 종괴의 밀도는 조영 증강 전에는 17 Hounsfield unit (이하 HU)이었고 조영증강 후 1분에 32 HU, 2분에 44 HU, 4분에 51 HU, 15분에 약 64 HU로, 지속적이며 비균질적인 조영증강을 보였다. 폐엽절제술을 시행하였고 병리 소견상 종괴는 두드러진 섬유모세포 성분을 가진 폐과오종으로 판명되었다.

부산대학교병원 의생명연구원 1영상의학과, 2흉부심혈관외과, 3병리과