

# Multiple Pulmonary Chondromas in a Young Female Patient

## *A Component of Carney Triad*

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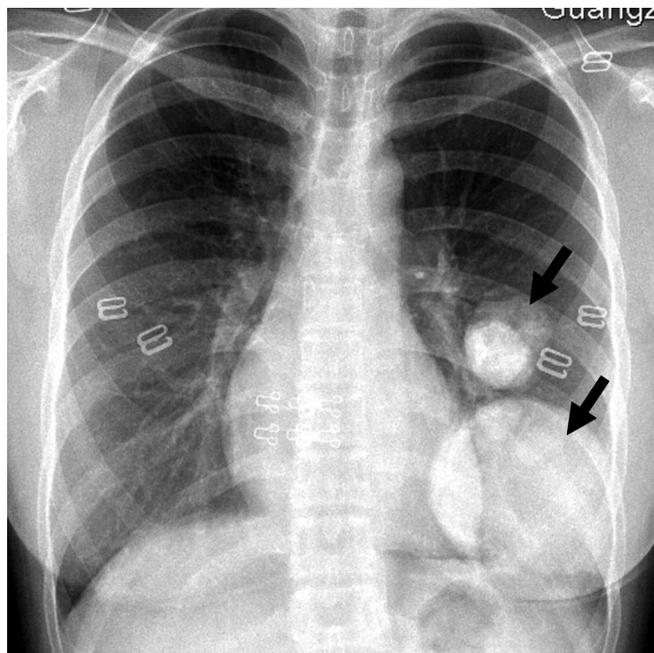
**Key Words:** Pulmonary chondroma, Pulmonary neoplasm, Carney triad.

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Chondromas of the lung are rare benign tumors, and fewer than 100 cases with multiple chondromas have been reported since Bateson<sup>1</sup> reported the first case. In 1977, J. Aidan Carney from the Mayo Clinic described pulmonary chondroma associated with gastrointestinal stromal tumors (GISTs) and extraadrenal paraganglioma as a syndrome, which was subsequently called the Carney triad.<sup>2</sup> Sporadic case reports and a recent analysis demonstrated that the chondromas in the Carney triad usually occur in young women and are multiple, small, peripheral and not endobronchial, and commonly calcified.<sup>2,3</sup> As most of the patients with Carney triad are asymptomatic and the pulmonary neoplasm(s) are usually found as incidental lesions on chest radiography, the diagnosis of Carney triad should be considered when multiple pulmonary chondromas are identified. Here, we report a case of Carney triad that initially presented with multiple lesions of the left lung.

### CASE REPORT

A 16-year-old girl was admitted to our hospital because three pulmonary lesions were detected on a chest radiograph as part of a medical examination for college entrance (Figure 1). The patient appeared well and was asymptomatic. The physical and laboratory examinations at the time of presentation were unremarkable. Computerized tomography of the chest revealed a mass in the left lower lobe and two nodules



**FIGURE 1.** The chest radiograph showed three round calcified tumors (arrows) in the left lung.

in the lingula in the left upper lung. The mass in the lower lobe was  $7.2 \times 6.5$  cm in diameter. The mass and nodules were well circumscribed, smooth, and inhomogeneous in density with peripheral calcification (Figure 2). The radiologic interpretation was multiple benign tumors of the left lung, suspicious of hamartomas or sclerosing hemangiomas. A thoracotomy was performed to obtain a definitive diagnosis and as curative treatment, and the three lesions were enucleated completely, while conserving the surrounding lung tissue. The gross resected specimens were multilobulated and consisted of elastic hard, small to large yellowish nodules. Microscopically, the tumors were composed mainly of cartilage with partial calcification and were separated from the adjacent pulmonary parenchyma by a fibrous pseudocapsule. These findings confirmed the diagnosis of multiple pulmonary chondromas.

The pathology revealed chondromas rather than hamartomas and prompted review of the literature where we found

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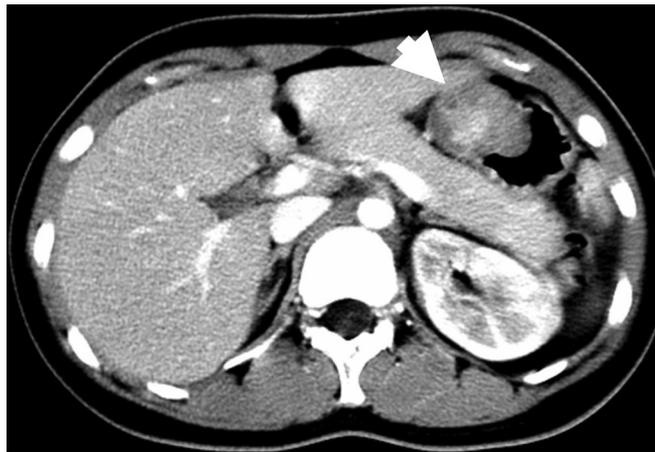
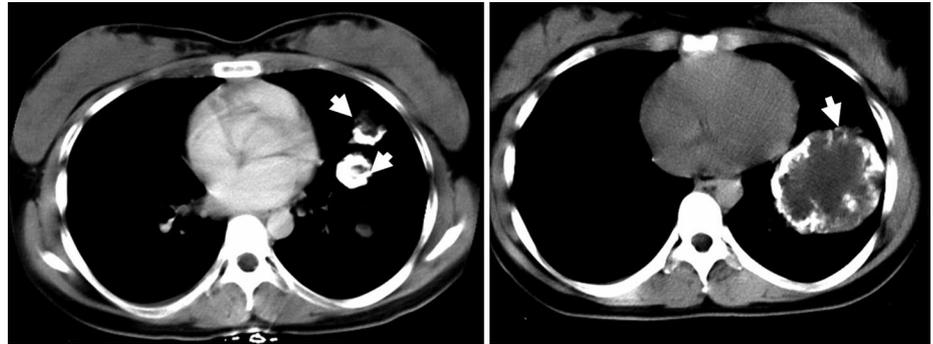
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**FIGURE 2.** Computed tomography of the chest revealed two nodules in the lingula segment of the left upper lobe (left) and a mass in the left lower lobe (right).



**FIGURE 3.** Computed tomography of the abdomen showed an inhomogeneous lesion between the left liver lobe and stomach.

that pulmonary chondromas are part of the Carney triad. Subsequent investigations were performed, including hormonal and alimentary tract examinations. Computerized tomography of the abdomen showed an inhomogeneous lesion between the left liver lobe and stomach measuring approximately  $4.4 \times 3.2$  cm in diameter (Figure 3). Gastroscopy revealed a submucosal lesion in the lesser gastric curvature. No evidence of an extraadrenal paraganglioma was detected despite an examination of urinary catecholamines and  $^{123}\text{I}$ -metaiodobenzylguanidine scanning. Two weeks after the thoracotomy, the patient underwent a subtotal distal gastrectomy. The histologic analysis confirmed the GIST, and an incomplete Carney triad was diagnosed. The patient's postoperative course was uneventful, and she received no adjuvant therapy.

## DISCUSSION

The Carney triad is a syndrome of unknown etiology that occurs predominantly in young females. The diagnosis of Carney's triad requires at least two of three components: GISTs, extraadrenal paraganglioma, and pulmonary chondroma. A recent analysis has indicated that the neoplasms comprising this disorder can appear at any age and in any sequence. Most patients had two tumors at presentation, and the most frequent combination was GISTs and pulmonary chondroma.<sup>2</sup> Because most of the patients were asymptomatic

and the pulmonary neoplasm(s) were usually identified incidentally or during an evaluation for metastatic disease after surgery for a gastric GIST,<sup>3</sup> such patients, especially young female patients, should be investigated carefully when a GIST or pulmonary chondroma is confirmed to avoid missing the other components of this rare syndrome.

The differential diagnosis of multiple benign appearing lesions includes chondromas, hamartomas, and sclerosing hemangiomas. Most patients with pulmonary hamartomas are men whose mean age at diagnosis is in the fifth decade of life. Pathologically, the chondromas in the Carney triad are composed mostly of cartilage, in contrast to pulmonary hamartomas, which have higher proportions of fibromyxoid stroma, fat, smooth muscle, and entrapped epithelium.<sup>2</sup> Pulmonary sclerosing hemangioma is a rare lung neoplasm usually presenting as a peripheral nodule, predominantly in asymptomatic middle-aged women. Histologically, a mixture of solid, sclerotic, papillary, and hemorrhagic components is observed in typical cases, and polygonal round cells with pale cytoplasm and cuboidal surface cells covering papillary structures can be found in each component.<sup>4</sup> Consequently, the pulmonary chondroma in the Carney triad differs from hamartoma and sclerosing hemangioma of the lung both clinically and pathologically.

Here, we report a case of incomplete Carney triad in a young woman, with the initial appearance of multiple pulmonary chondromas. We emphasize that the Carney syndrome must be considered and a careful investigation of other organs, especially the alimentary and nervous systems, must be conducted when multiple lesions of the lung are observed in young females. Furthermore, patients with incomplete Carney triad should be kept under close long-term follow-up because they presumably remain at risk for its full expression.

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