

Case Report

Isolated pulmonary chondroma: a case of incomplete Carney triad?*

RAUL LOPES RUIZ JÚNIOR¹, JÚLIO DEFAVERI², ANTONIO JOSÉ MARIA CATANEO³,
ROGÉRIO CARDOSO DA SILVA⁴, SÉRGIO MARRONE RIBEIRO⁵, CRISTIANO VENTORIM DE BARROS⁶

ABSTRACT

A 45-year-old man presented with recurrent pulmonary infection for four years, cough, bloody sputum, yellowish excretion and nonpleuritic chest pain. Tomography of the chest revealed a calcified nodule occluding the right lower lobe bronchus. A right lower and middle lobectomy was performed, and the histopathological examination of the bronchi revealed chondroma, a rare pulmonary tumor usually associated with the Carney triad (pulmonary chondroma, gastric leiomyosarcoma and extra-adrenal paraganglioma), being the less common of the three components. In the present case, the other two components of the triad were not observed. Since these components may appear years later, long-term follow-up care is necessary.

Keywords: Chondroma; Lung neoplasms/radiography; Lung neoplasms/surgery; Leiomyosarcoma; Stomach neoplasms; Paraganglioma, extra-adrenal; Case report

INTRODUCTION

Pulmonary chondroma is a rare neoplasm.⁽¹⁻⁶⁾ It is primarily seen in women and is part of the Carney triad, which consists of pulmonary chondroma, gastric leiomyosarcoma and functioning extra-adrenal paraganglioma.⁽¹⁾ In describing this triad, Carney reported four cases involving young women. He suggested that the concomitant development

of these rare tumors was not random and might constitute a syndrome. Two years later, he confirmed this hypothesis when he observed that fifteen patients under 35 years of age presented one of the three tumors of the triad and should be examined periodically in order to diagnose the other two components.⁽²⁾ Since then, it has been shown

*Study conducted at the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil.

1. Assistant Professor of Thoracic Surgery in the Department of Surgery and Orthopedics of the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

2. Adjunct Professor in the Department of Pathology of the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

3. Adjunct Professor of Thoracic Surgery in the Department of Surgery and Orthopedics of the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

4. Medical Student at the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

5. Assistant Professor of Radiodiagnostics in the Department of Tropical Diseases and Diagnostic Imaging of the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

6. Resident at the Universidade Estadual Paulista (UNESP) Julio Mesquita Filho School of Medicine in Botucatu, São Paulo, Brazil

Correspondence to: Raul Lopes Ruiz Jr. Caixa Postal 501 CEP: 18618-970 - Botucatu - SP

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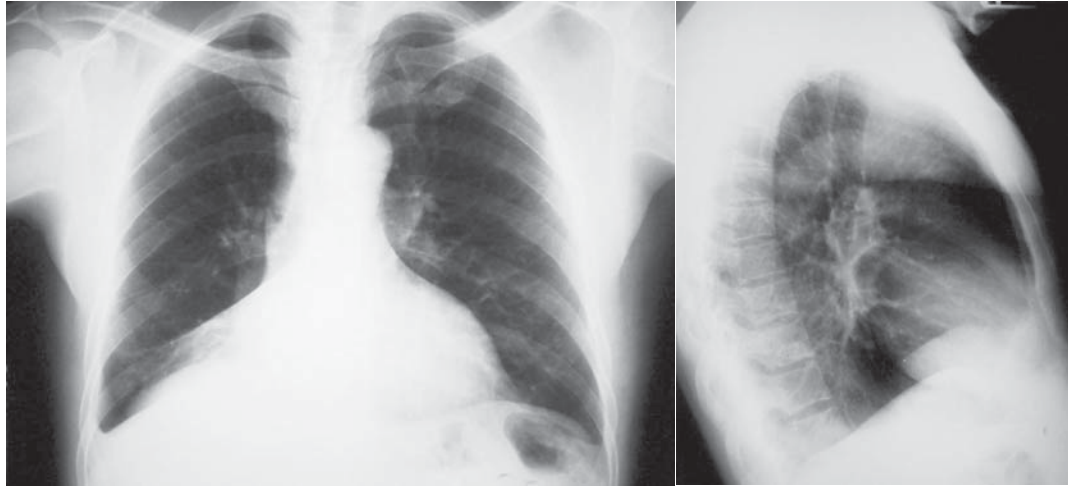


Figure 1 - Anteroposterior and lateral chest X-ray: atelectasis of the right lower lobe, characterized by a t-shaped opacity obliterating the right border of the heart, with flattening of the right hilum and elevated right diaphragmatic dome

that most patients present only two of the three components of the triad.⁽³⁾

In this study, we report the interesting case of a man presenting with pulmonary chondroma, apparently as the only component of the triad.

This study was approved by the Ethics in Research Committee of the Paulista State University School of Medicine at Botucatu.

CASE REPORT

A 45-year-old male patient, presented with cough and yellowish excretion streaked with blood. He reported continuous, piercing, moderately-intense chest pain, located in the posterior right hemithorax, which did not become more intense upon coughing or in the decubitus position. The patient had a four-year history of recurrent pulmonary infections. He stated that he did not have diabetes, arterial hypertension or cardiopathy. He had been a smoker for 30 pack-years and was a social drinker. He denied having lost weight.

Anteroposterior and lateral chest X-rays revealed signs of reduced volume in the right lower lobe and blurring of the right cardiac silhouette (Figure 1).

A computed tomography scan of the chest confirmed atelectasis and revealed a small calcified lesion in the right lower lobe bronchus (Figure 2), in addition to bronchiectasis with probable

impacted mucus in the collapsed pulmonary lobe.

Bronchoscopy revealed a vascularized lesion, totally obstructing the lumen of the right lower lobe bronchus (Figure 3A), near the middle lobe bronchus. An endobronchial biopsy was not carried out due to the risk of bleeding, since a probable diagnosis of carcinoid tumor was inferred.

Due to the clinical suspicion of malignant lesion, the patient was submitted to a bilobectomy of the lower and middle lobes. The postoperative evolution was favorable and uneventful.

The results of anatomopathological tests revealed a nodular tumor measuring 1.5 x 1.0 x 0.8 cm, totally obstructing the lumen of the bronchus and easily seen (Figure 3B). The histology revealed a typical chondroma with central calcification, bronchial mucous with epithelial metaplasia covered the luminal surface. Being aware of the possible correlation with the Carney triad, we carried out radiocontrast exams of the esophagus, stomach and duodenum. In addition, we performed endoscopy of the upper digestive tract and an abdominal ultrasound, as well as computed tomography of the chest, abdomen and neck. No other lesions were found.

Scintigraphy with metaiodobenzylguanidine (¹³¹I MIBG) was used as a test for extra-adrenal paragangliomas, and none were found.⁽⁷⁾

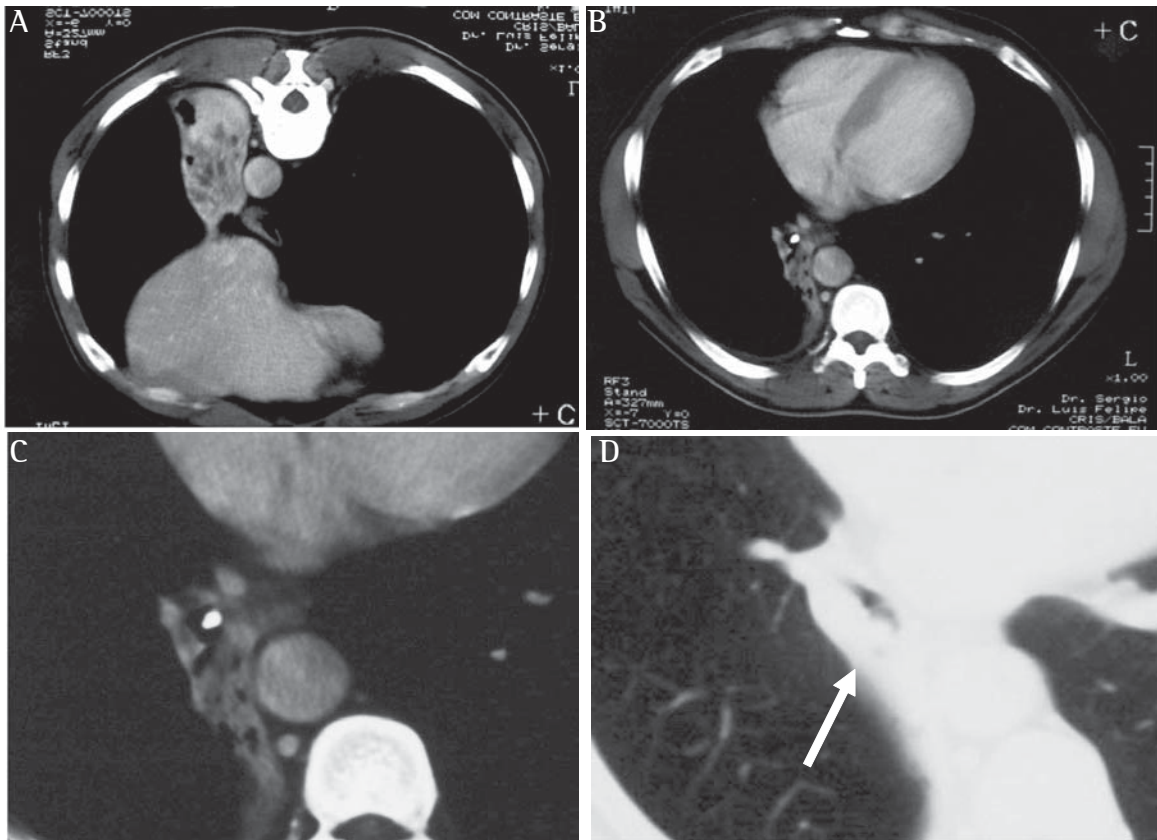


Figure 2 - Computed tomography scan of the chest: signs of reduced volume with bronchiectasis in the right lung (A and B) due to a totally calcified nodule in the lumen of the right lower lobe bronchus (C and D)

DISCUSSION

The combination of pulmonary chondroma, gastric leiomyosarcoma and extra-adrenal paraganglioma constitutes the Carney triad, which presents some peculiarities.⁽³⁾ except in metastases, tumor multicentricity among different organs/tissues is uncommon. Such multicentricity occurs among three well-defined organs/tissues: the stomach, the lung and the central nervous system. In addition, the syndrome is predominantly found in young women.

The incomplete triad comprises the pulmonary and gastric components and was the combination that Carney found to be the most frequent in his last study.⁽³⁾ He observed that 42 (53%) of the 79 patients evaluated presented this combination, and 17 (22%) presented all three components.⁽³⁾ A diagnosis of the complete triad, in which the three components are present simultaneously, is even rarer.⁽⁶⁾ A finding of a pulmonary chondroma in

isolation is rare and is almost always accompanied by one of the other two components.⁽³⁾ The case of a Brazilian woman who presented the incomplete Carney triad, consisting of gastric leiomyosarcoma and pulmonary chondroma, has been reported.⁽⁶⁾

Reports in the international literature show that the gastric component is the most frequently present. This component is characterized by early onset and local symptoms. Despite progressing slowly, the gastric component of the triad typically evolves to lymphatic and hematogenous abdominal metastases.⁽³⁾

Pulmonary chondroma is a benign slow-growth mesenchymal neoplasm that presents no symptoms or deleterious effects to the patient and tends to develop other tumors.⁽³⁾ Therefore, patients with this disease should be monitored, clinically and radiologically, over time. Pulmonary chondroma is usually diagnosed during preoperative evaluation. After the anatomopathological diagnosis is confirmed, radiological monitoring will suffice.

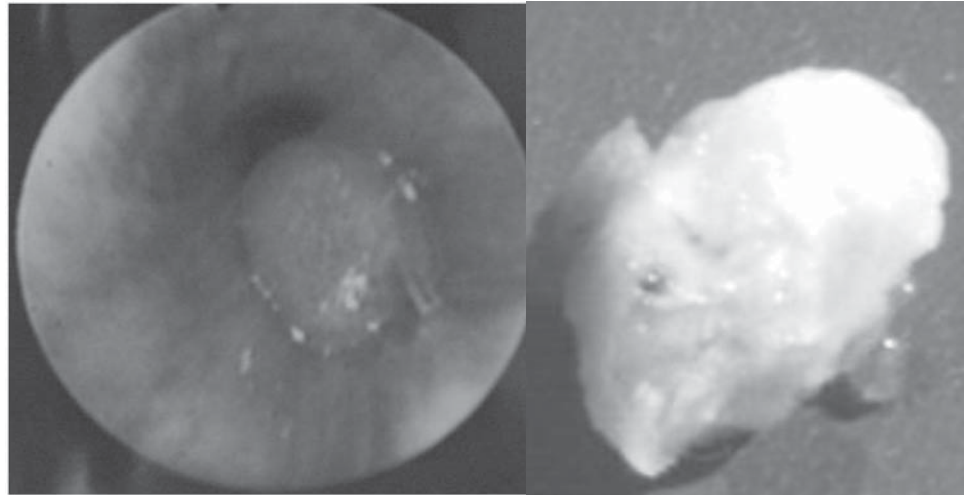


Figure 3 - Bronchoscopic view: A) Lesion totally obstructing the lumen of the right lower lobe bronchus. The lumen of the middle lobe bronchus is shown above; B) A "plug" occluding the right lower lobe bronchus

Surgical treatment is recommended in cases presenting pulmonary function impairment or other tumor-related complications.⁽³⁾

In radiographic images, pulmonary chondroma may appear as a single tumor, multiple unilateral lesions or bilateral lesions. It may affect any lobe of either lung. In half of the cases reported in the literature, there was calcification at the time of diagnosis.⁽³⁾

In view of the statistics reported by Carney,⁽³⁾ the present case is quite uncommon. The patient was male and over 35 years of age; the tumor was central and isolated; and the symptomatology was atypical. In his most recent study, Carney described only one patient over 35 years of age, and the great majority of patients were female (67 of the 79 described). All except one of the patients studied were asymptomatic.⁽³⁾ In our patient, the pulmonary component appeared as an endobronchial obstructive lesion, leading to signs and symptoms of pulmonary suppurative disease with recurrent infections.

Two of the three components of the Carney triad are malignant tumors with significant rates of metastases. In addition, since there are long intervals between the onset and the recurrence of the tumors, long-term follow-up evaluation is necessary after the first appearance of the symptoms. Even asymptomatic patients should be

examined every three years to determine whether the other components of the syndrome have appeared. Such examinations should also be carried out as soon as new symptoms appear.⁽⁸⁻⁹⁾

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