Letter to the Editor

**Idiopathic Plastic Bronchitis as an Uncommon Cause of Massive Pulmonary Atelectasis**

**Bronquitis plástica idiopática como causa infrecuente de atelectasia pulmonar masiva recurrente**

To the Editor,

Plastic bronchitis (PB) is an uncommon disease characterized by the formation of bronchial casts that partially or completely block the bronchial lumen.\(^1\)\(^,\)\(^2\) It has been described as a complication of respiratory diseases, lymphatic abnormalities, infections and, particularly, congenital heart disease surgery, especially Fontan palliation,\(^1\)\(^,\)\(^2\) and is commonly confused with bronchoaspiration.\(^2\) We report a case of idiopathic PB.

A 10-year-old boy was admitted for irritative cough, pain in left hemithorax and dyspnea with onset 3 weeks previously. Of note on examination was severe hypoventilation of the left hemithorax. Anteroposterior chest X-ray showed left pulmonary collapse (Fig. 1A). Pneumonia was suspected, so intravenous antibiotics were started, with no clinical or radiological improvement. Lung computed tomography showed atelectasis of the left upper and lower lobes due to bronchial stenosis located 2 cm from the carina. The patient’s mother was consulted again and she remembered a choking fit when the boy was eating a hamburger approximately 2 months previously. In view of suspected foreign body, fiberoptic bronchoscopy (FB) was performed revealing a foreign body in the left main bronchus. Multiple fragments (Fig. 1B and C) were extracted by rigid bronchoscopy (RB). Two weeks later, the patient’s situation worsened. PB was repeated, showing fragments of yellowish intrabronchial foreign bodies that were extracted by RB, and on pathological examination were reported to be mucoid, fibrinopurulent material. In view of this, PB was diagnosed and treatment with nebulized DNase was started. After 2 relapses of atelectasis, oral prednisolone (2 mg/kg/day), azithromycin (500 mg/day, 3 days/week) and nebulized N-acetylcysteine were added. Various complementary studies were performed to rule out diseases associated with PB: all were negative. Two years later, the patient remains asymptomatic and continues to receive azithromycin.

PB is a rare, heterogeneous lung disease.\(^1\) Documented cases are generally secondary to an underlying condition.\(^1\) The most widely accepted classification is that of Seear, based on the characteristics of the bronchial cast and the underlying disease:\(^2\) Type I (inflammatory) consists of fibrin casts, Charcot-Leyden crystals and eosinophilia, and is generally associated with allergies or inflammatory diseases;\(^2\) Type II consists of mucin casts and occurs with congenital heart disease.\(^2\) Clinical manifestations are non-specific and vary widely, including expulsion of bronchial casts, dyspnea, wheezing, fever and cough, similar to an asthma attack or aspiration of foreign body,\(^1\)\(^,\)\(^2\) as was the case in our patient. Radiological signs are non-specific, including mainly atelectasis of the involved segment and compensatory hyperinflation.\(^2\) A more specific radiological finding is an elongated opacity with undulating borders.\(^2\) Treatment is a source of controversy, and recommendations are generally based on isolated cases or series of heterogeneous cases.\(^2\) The most common treatments are azithromycin,\(^3\) nebulized N-acetylcysteine, urokinase or recombinant tissue plasminogen activator (rt-PA) and DNase, respiratory physiotherapy and corticosteroids for PB associated with asthma or infections,\(^4\) and in more severe cases, extraction of the casts by bronchoscopy.\(^1\)\(^,\)\(^2\) Prognosis tends to be good,\(^1\) except in cases with associated congenital heart disease, where mortality rates can reach 29% and life-threatening events can occur in up to 41% of patients.\(^5\) Despite its rarity, it is worth underlining the need for considering this disease in the differential diagnosis of intrabronchial foreign body, particularly in cases of recurrent atelectasis.

![Fig. 1.](image)

(A) Massive left pulmonary atelectasis. (B) Bronchial cast of up to 4 cm in length extracted by rigid bronchoscopy. (C) Other bronchial casts.

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An Uncommon Intrafissural Mass: Pleural Primitive Neuroectodermal Tumor

Una masa intracisural infrecuente: tumor neuroectodérmico primitivo pleural

To the Editor:

We report the case of a young asymptomatic patient with a chest mass that was finally diagnosed as an intrafissural primitive neuroectodermal tumor (PNET). A pulmonary mass was detected in this 27-year-old woman on an unrelated chest X-ray. All laboratory test results were normal. Computed tomography (CT) revealed an elliptic, heterogeneous mass with contrast uptake in the left hemithorax, about 12 cm in diameter with smooth borders, located within the oblique fissure of the left pleura (Fig. 1). CT-guided fine-needle aspiration biopsy cytology was inconclusive. Surgical resection revealed an intrafissural mass. Hematoxylin and eosin staining produced a surprisingly uniform pattern of solidly compacted round cells.

Immunohistochemical assays showed tumor cells that were strongly positive for MIC2 (CD99) and for vimentin, but negative for other markers, such as cytokeratin (CK), small cell lung cancer, chromogranin, CK7, CK19 and thyroid transcription factor-1. Molecular genetic studies using fluorescence in situ hybridization showed positive translocation of the long arms of chromosomes 11 and 22 [t(11;22)(q24;q12)]. These results were suggestive of PNET. No distant metastases were found using standard staging methods, including brain magnetic resonance imaging (MRI), whole body MRI, abdominal CT and bone scintigraphy. After complete surgical resection, the patient received chemotherapy. After complete surgical resection, the patient received chemotherapy. Since then, she has been monitored in the outpatient clinic and her situation remains unchanged after 2 years.

Chest PNETs, also known as Askin's tumors, probably originate in the embryonic neural crest cells. They consist of small round cells with neural differentiation, and are associated with translocation of chromosome 22. They typically appear in the soft tissues of the chest. A primary tumor located inside the lung is very rare, but pleural involvement is common. Patients are generally children or young adults, although PNET can develop at any age. It usually presents with pain in the chest wall, and can be associated with pleural effusion and dyspnea.

The most common CT finding in PNET is a mass with heterogeneous contrast uptake. Occasionally, a central area of low density with no contrast uptake seen inside the mass. The heterogeneous aspect of these large tumors is usually due to focal areas of bleeding or necrosis.

Diagnosis of PNET is made using various techniques, such as optical microscopy, histological findings of small round cells arranged in a lobular pattern with rosettes or pseudorosettes, and by immunohistochemical and ultrastructural signs of neural differentiation. Reciprocal translocation of the long arms of chromosomes 11 and 22 [t(11;22)(q24;q12)] is currently thought to be characteristic of this tumor family. Although there are no