Endobronchial Obstruction by an Inflammatory Myofibroblastic Tumor

Obstrução Endobrônquica por um Tumor Miofibroblástico Inflamatório

Keywords: Airway Obstruction; Bronchial Neoplasms; Bronchoscopy; Myofibroblasts
Palavras-chave: Broncoscopia; Miofibroblastos; Neoplasias Brônquicas; Obstrução das Vias Aéreas

Dear Editor,

Inflammatory myofibroblastic tumors (IMT) are rare benign neoplasms comprised of myofibroblasts associated with a polymorphic inflammatory stromal infiltrate[1]; they may occur in different organs/structures[2] and mostly affect young individuals, up to forty years old.[3]

We present a case of a 26-year-old woman who attended the emergency department with a two-day history of fever and a dry cough; she also noted mild fatigue with approximately three weeks duration. At presentation she was tachypneic and hypoxemic; pulmonary auscultation showed decreased lung sounds in the lower left hemithorax. A chest radiograph documented lower left lobe atelectasis. Her condition deteriorated rapidly during the next 24 hours, with development of severe respiratory failure; a second chest radiograph now showed total left lung atelectasis. Computerized tomography (CT) (Fig. 1A) confirmed the atelectasis,

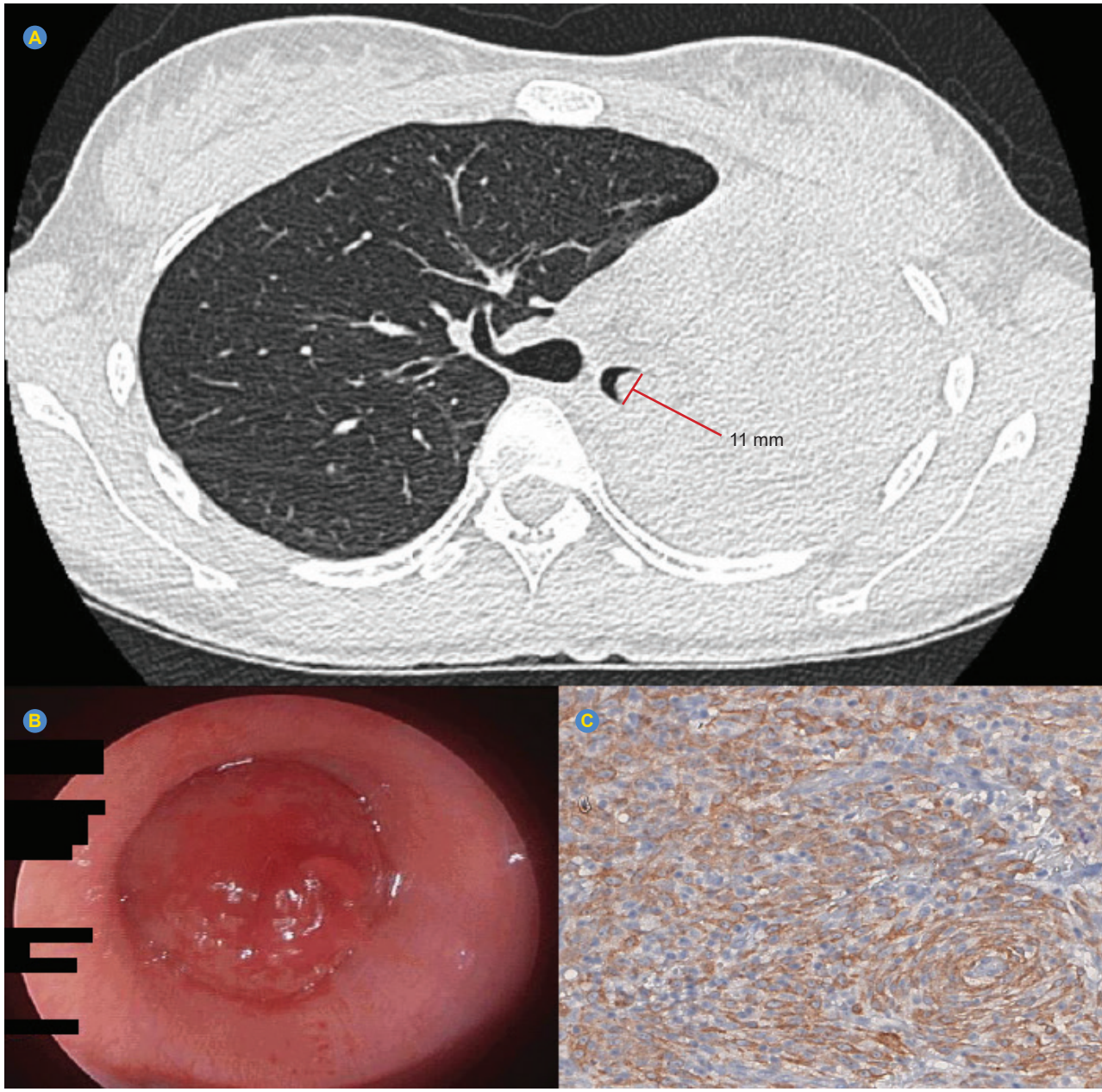

Figure 1 – Computerized tomography with total left lung atelectasis, showing endoluminal lesion (A); Endobronchial image obtained via rigid bronchoscopy, showing an obstructive, highly vascularized lesion in the left main bronchus (B); Immunohistochemical exam, with positive staining for cytoplasmatic ALK expression (C).

and an expansive lesion in the left main bronchus (LMB) was noted. Rigid bronchoscopy was performed, showing a highly vascularized, smooth tumor, causing total occlusion of the LMB (Fig. 1B). Laser photocoagulation and mechanical debulking were performed; there was severe bleeding, which was controlled with cold saline, topical adrenaline, and tranexamic acid. The tumor was totally removed, and the complete patency of the left bronchial tree was documented. Respiratory failure was completely resolved after the procedure. The patient is currently doing well and is being followed with regular bronchoscopies and CT scans. A positron emission tomography has also been performed which excluded the presence of any tumor with metabolic activity.

Microscopically, the tumor was composed of spindle to epithelioid myofibroblasts arranged in fascicular and storiform patterns, accompanied by inflammatory infiltrate composed of lymphocytes, plasmacytes, eosinophils and neutrophils. Necrosis was absent and mitotic activity was evaluated as low-to-moderate, with five mitoses in 10 high power fields. Immunohistochemical study demonstrated cytoplasmic expression of anaplastic lymphoma kinase (ALK) (Fig. 1C) and no immunoexpression for desmin and CK8/18. The histopathologic characteristics favored the diagnosis of IMT.

Inflammatory myofibroblastic tumors rarely manifest in the lung or bronchus (0% - 12%)[3] and are a rare cause of lung tumors (0.04% - 1%), which reinforces the rarity of this case. Although they are benign, they may have an aggressive course, with a risk of local progression, relapse (up to 37%)[3] and metastasis (up to 11%),[3] highlighting the importance of being aware of this entity and considering it in the differential diagnosis of rapidly growing endobronchial tumors. In this case, due to rapid patient deterioration, emergent treatment was required. Although surgical excision is considered the best treatment,[3,4] bronchoscopic intervention has been used, with comparable success to surgery and the benefit of being more readily available. In cases of relapsing or metastatic disease, which is more likely if the resection is incomplete, the role of medical treatment remains understudied.[4,5] It has been shown that overexpression of ALK (which is present in up to 60% of IMT[4]) may be a therapeutic target, with the possibility of treatment with crizotinib or ceritinib.[3] Current evidence suggests that, in patients with advanced disease who are not surgical candidates, crizotinib may be an interesting option, with response rates of up to 50% in ALK-positive patients, which favors considering its use in this subset of patients as standard treatment; further studies should be undertaken to better understand the role of ALK inhibitors, both as a complement and as an alternative to local excision.

AUTHOR CONTRIBUTIONS

BSS, MLS: Data collection, writing of the manuscript.
HNB: Critical review of the manuscript.
All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in 2013.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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