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Lithoptysis in a Patient With Primary Ciliary Dyskinesia

To the Editor: We read with interest the report by García Pachón et al[1] of a case of idiopathic chronic lithoptysis. As a complement to their observations, we describe the case of a patient diagnosed with primary ciliary dyskinesia who presented an episode of lithoptysis while being followed by our department. Dyskinetic cilia syndrome or primary ciliary dyskinesia is a recessive autosomal illness that is characterized in its full manifestation by chronic rhinitis, sinusitis, otitis, recurring bronchitis, bronchiectasis, male sterility, and corneal and olfactory abnormalities. These patients suffer from impaired mucociliary clearance, which is associated with the need for ongoing respiratory physiotherapy exercises. Broncholithiasis is an unusual entity defined by the presence of hilar calcifications or calcified peribronchial lymph nodes that erode the bronchial wall and can enter the bronchial lumen, giving rise to clinical and radiologic abnormalities.[2] Infection by *Mycobacterium tuberculosis* is currently the main cause of broncholithiasis, followed by infection by *Histoplasma capsulatum*.[3] One sign of broncholithiasis is lithoptysis, which consists of expectoration of one or more broncholiths, an event that can resolve the clinical picture in some cases.

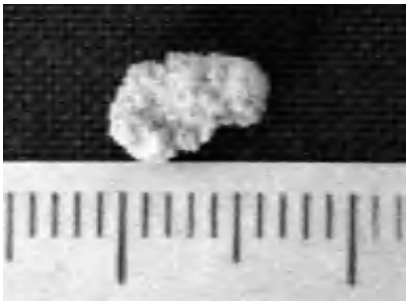

Figure 1. Photograph of the broncholith.

A 34-year-old man diagnosed with primary ciliary dyskinesia developed a cough and spontaneously expectorated a stone (Figure 1), with no other associated manifestations. The patient could not recall any similar previous episodes. His medical history included an idiopathic pleural effusion in childhood, repeated episodes of bronchial hyperresponsiveness in the context of respiratory infections, bilateral bronchiectasis, and pulmonary tuberculosis diagnosed in 1994. The patient was therefore prescribed treatment for 6 months. The family history included a sister who had also been diagnosed with primary ciliary dyskinesia. The pneumologist's examination showed only overall reduced vesicular murmur, with prolonged expiration and some isolated rhonchi. The hemogram, coagulation tests, and biochemistry—including bone metabolism, immunoglobulin study, proteinogram, α_1-antitrypsin, and thyroid function—were normal. Hepatitis and human immunodeficiency virus serologies were negative and both urine analyses and kidney function tests were normal. Spirometry at rest showed a forced expiratory volume in the first second of 39.1% of predicted and a forced vital capacity of 70% of predicted; the ratio between the 2 was 40.98. Computed tomography (CT) of the paranasal sinuses revealed hypogenesis of the frontal and maxillary sinuses and mucosal thickening. A high-resolution CT scan of the chest revealed correctly placed mediastinal structures, with no adenomegaly in the hilar and mediastinal regions; small apical fibrotic areas, bilateral cylindrical bronchiectasis, and slight apical pleural thickening were disclosed. The CT scan also revealed 2 calculi of 3 mm and 5 mm, respectively (Figure 2), inside the right bronchial tree in the upper and middle lobes. Bronchoscopy revealed no broncholiths in the bronchial lumen or abnormalities in the mucosa that might indicate underlying lithiasis. The expectorated broncholith had a hard, irregular, whitish coralliform surface upon macroscopic inspection. Mineralogical analysis showed the stone to be 70% calcium oxalate; the remainder consisted of calcium phosphate (hydroxyapatite). In light of these analyses and because the patient did not present symptomatic broncholithiasis at the time, we adopted a conservative treatment approach.

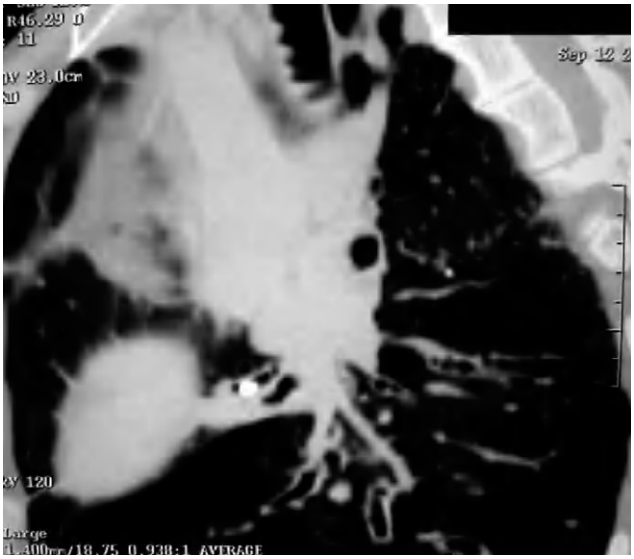

Figure 2. Computed tomography scan of the chest showing 2 calculi of 3 mm and 5 mm.

Symptomatic broncholithiasis is uncommon. Symptoms develop when these calcifications penetrate the bronchial lumen and give rise to various symptoms, such as persistent cough, hemoptysis, recurrent pneumonia, fistulas between the bronchial wall and the adjacent mediastinal structures, and lithoptysis.[2,4] In a Mayo Clinic series of cases from 1954 to 1994, lithoptysis was present in only 15 of 95 patients with broncholithiasis, making it an uncommon entity.[5]

With regard to the pathogenic mechanism involved in our case, we may suppose that pulmonary tuberculosis triggered the broncholithiasis; we should remember that the patient's sister, while also diagnosed with broncholithiasis and primary ciliary dyskinesia, had no record of suffering from pulmonary tuberculosis. A search of the MEDLINE database (1965-2005) returned no references for patients with a diagnosis of broncholithiasis associated with primary ciliary dyskinesia. Because primary ciliary dyskinesia is an entity where episodes of respiratory tract infection can be frequent, and organic material such as bacteria remains in a context of impaired mucociliary clearance and the presence of bronchiectasis, we hypothesize that these mechanisms may help to create heterogeneous nucleating sites that can form broncholiths, as noted by García Pachón et al[1] in their case report.

There is no consensus regarding treatment options, which range from mere observation to bronchoscopic broncholithectomy and, finally, surgery.[2] In our case, we opted for a wait-and-see approach given the absence of symptoms or complications.

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