Association between Gastroesophageal Reflux and Endobronchial Carcinoid: A Case Report

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Abstract

Children with neurological disorders may suffer from gastroesophageal reflux disease (GERD). Typical symptoms are vomiting, regurgitation and hematemesis. Patients present with respiratory symptoms only in cases with swallowing disorders causing chronic airway aspiration. We report the case of a patient affected by chromosome 8 p deletion syndrome with mental retardation, referred to our unit for suspected GERD. Chest X-ray, performed at admission for coexisting respiratory complaints, showed left lower lobe pneumonia; esophageal pH monitoring and upper endoscopy were normal for GERD. To rule out chronic airway aspiration, gastroesophageal 99 mTc scintigraphy with lung scan 18 to 24 h after a test meal and video fluoroscopy swallowing study were performed, both negative. Two months later, a second episode of left lower lobe pneumonia occurred. A chest CT scan was performed and showed an endobronchial mass; the biopsy taken during the broncoscopy was not conclusive. Surgical excision resulted in a diagnosis of pulmonary carcinoid. Bronchial carcinoids, although rare, should be taken into consideration as a potential cause of recurrent pneumonia even in the presence of demonstrated GERD where severe respiratory infections only occur with coexisting chronic pulmonary aspiration, even in neurologically impaired people.

Key words
Bronchial carcinoid · GERD · mental retardation · recurrent pneumonia

Introduction

Gastroesophageal reflux disease (GERD) is common in children with neurological disorders and mental retardation due to esophageal motility dysfunction and incompetence of the lower esophageal sphincter, which leads to reflux of gastric content into the esophagus causing esophagitis and GERD [1,2]. Vomiting, regurgitation and hematemesis suggest GERD. Conversely, respiratory symptoms occur only in association with demonstrated chronic airway aspiration [2–4]. We report the case of a patient with mental retardation who developed a bronchial carcinoid, detected during the diagnostic workup for severe GERD and recurrent pneumonia.

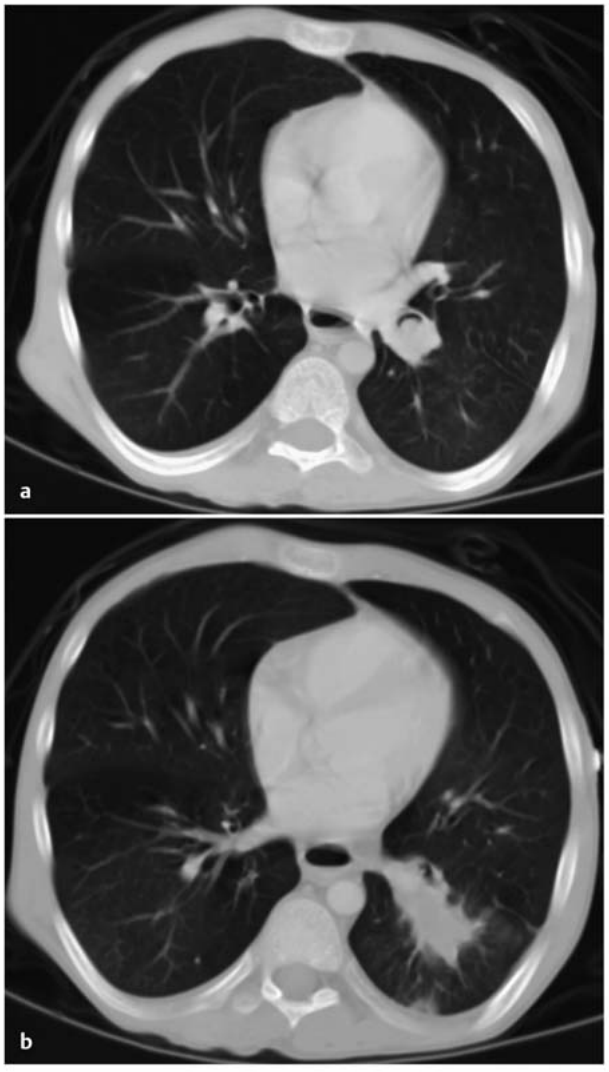

Fig. 1 a and **b** CT scan showing an intrabronchial left inferior hilar mass (**a**), located in the left lower lobe bronchus, associated with obstruction of the distal airways (**b**) and pleural thickening (posterobasal segment of the left lower lobe).

Case Description

A 13-year-old boy affected by chromosome 8 p deletion syndrome, with mental retardation and feeding difficulty, was referred to our unit for suspected GERD, because of repeated hematemesis, regurgitation and vomiting. Ten months previously he had had pneumonia. Clinical examination at admission showed impaired nutritional status (weight < 3 SD; height < 2 SD; BMI: 15), pale skin and mild tachypnea. Hemoglobin, plasmatic iron, and sodium were below and C-reactive protein above the reference ranges for age. Chest X-ray showed left lower lobe pneumonia that required antibiotic therapy. GERD was then confirmed by positive 24-hour esophageal pH monitoring time (pH < 4 of the 25%) and by upper endoscopy which showed hiatal hernia and severe esophagitis. Accordingly, we started treatment with pro-

ton pump inhibitors. To exclude chronic airway aspiration as suggested by the two episodes of pneumonia, we completed the diagnostic workup by performing gastroesophageal 99 mTc scintigraphy with lung scan 18 to 24 h after a test meal and video fluoroscopy swallowing study, both negative. Sweat-chloride test, immunological assessment and purified protein derivative (PPD)-tuberculin test were also negative.

Two weeks after admission the boy demonstrated recovery from pneumonia on a second chest X-ray, together with weight gain (1.5 kg) and normalization of blood tests. Thus he was discharged with a diagnosis of GERD and pneumonia and the recommendation to continue proton pump inhibitor treatment for at least 3 months.

Two months later he was again admitted for coughing, wheezing, tachypnea and hemoptysis. A further left lower lobe opacification was observed on chest X-ray. Therefore a chest computed tomography (CT) scan was performed and showed an endobronchial round mass of 13 × 10 mm in the left basal lobe, obstructing the left lower bronchus (**Fig. 1**).

Fiber bronchoscopy confirmed the presence of a solid mass obstructing the left lower bronchus with pink granulation tissue. Histological examination showed inflammatory infiltration, squamous metaplasia and an absence of lipid-laden macrophages but failed to provide a specific diagnosis because the specimen was very small and bleeding. The patient underwent complete surgical resection of the mass, with left lower lobectomy. Intraoperative histological examination of the lower wall of the bronchus and the peribronchial lymph nodes excluded any neoplastic involvement. Biopsy specimens showed a neoplasm composed of cords of tumor cells with uniform round nuclei, low mitotic activity and no necrosis (**Fig. 2a**). Immunohistochemical staining for chromogranin was positive (**Fig. 2b**). Based on the pathological findings, the diagnosis was a bronchial carcinoid with a typical histology of pT1, pN0, pMx, stage I.

Two months after surgery, ^{111}In-DTPA-pentetreotide scintigraphy (octreoscan) was performed which ruled out local recurrence and distant metastases. Two years have elapsed since surgery; the patient remains in a good clinical condition and octreoscan and blood chromogranin, scheduled every 6 months, have remained negative.

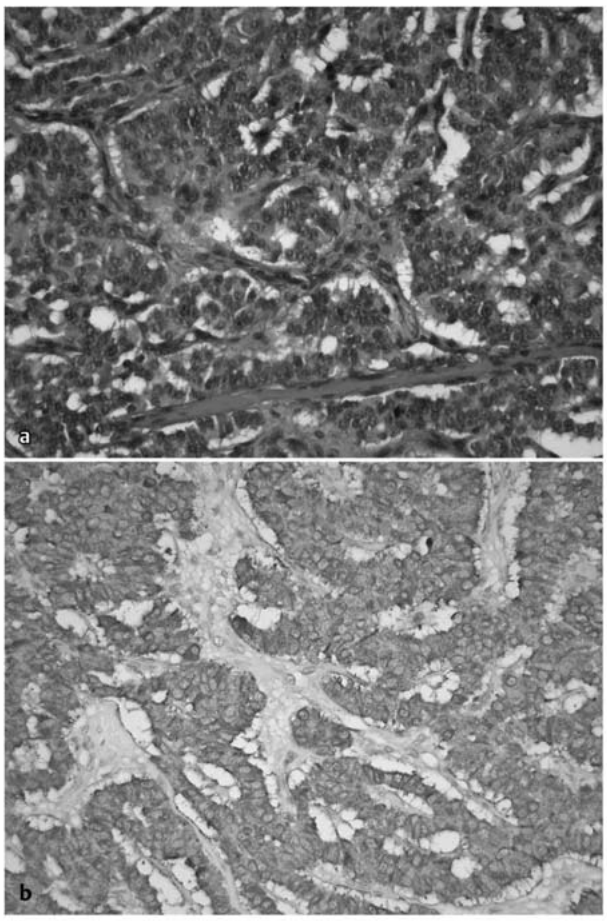

Fig. 2a and **b** **a** Trabecular and nodular pattern of neoplastic proliferation. Round-shaped cells have a uniform size and no nuclear atypia (20 × H & E stain). **b** Neoplastic cells were positive for chromogranin immunostaining in a negative stromal background (20× DAB chromogen).

Discussion

The particular interest of this case is that GERD, in a patient with mental retardation and feeding difficulty, delayed the diagnosis of a bronchial carcinoid. Indeed, we considered the recurrent pneumonia as a complication of severe GERD, although gastroesophageal 99 mTc scintigraphy with lung scan and video fluoroscopy ruled out chronic airway aspiration. Asthma and recurrent upper airway symptoms should be considered as possible complications of GERD only after excluding anatomical causes, foreign bodies, cystic fibrosis or immunodeficiency [5]. In patients with and without neurological deficits, severe respiratory infections, such as recurrent pneumonia, are also not usually associated with GERD [2,3], unless occurring due to oropharyngeal incoordination and chronic airway aspiration [6].

Bronchial carcinoid has a low incidence in children (1 to 1.42 children per million aged younger than 15 years); the presenting complaints are wheezing, hemoptysis, and pneumonia in 40–50% of patients [7]. Although bronchial carcinoid is often not malignant, its treatment is based on complete surgical resection; therefore early diagnosis increases the likelihood of definitive management before the tumor expands [7,8].

In conclusion, our experience leads us to make two recommendations: 1) Even in patients with neurological impairment, GERD should be considered as a cause of recurrent pneumonia only if it coexists with chronic respiratory aspiration, otherwise it is necessary to search for other causes; and 2) Among the potential causes of recurrent pneumonia, bronchial carcinoid, although rare, should be taken into consideration because early diagnosis, which potentially reduces the extent of surgical resection, results in a more favorable prognosis.

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