Treating “asthma” with a scalpel – achalasia in a patient impersonating asthmatic condition

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In every case of asthma which remains partially controlled or even uncontrolled despite qualified treatment, experts stress the necessity to verify the diagnosis, ruling out conditions which can masquerade as asthma [1,2]. According to some reports, misdiagnosis of non-asthmatic conditions treated as uncontrolled asthma runs as high as 12-30%, which is why a degree of skepticism is recommended [3].

An 18-year-old female was referred to Allergy Department with the diagnosis of severe asthma. Her asthma was partially controlled or even uncontrolled, with signs of bronchial obturation which persisted despite intensive treatment (GINA guidelines, grade 5: daily doses of fluticasone propionate 1000mcg and salmeterol 100mcg; additionally prednisone 20-40mg/daily for 5-7 days every second or third month). Since the onset of her sickness, when she was 13, her predominant symptoms were: cough, breathlessness and wheezing that occurred predominantly at night, causing frequent nocturnal wakening. She also reported rhinorrhea during sleep. The patient’s mother termed the nocturnal symptoms as ‘noisy breathing’. During the last two years, the patient had pneumonia twice, and many times “bronchitis” treated with antibiotics. The patient denied occurrences of paroxysmal dyspnea, exercise-triggered dyspnea or one induced by other factors, specific or nonspecific. Neither did she report any other symptoms, particularly gastric ones. She
presented sensitization to house dust mites. She had no family history of atopy and asthma. She never smoked cigarettes.

Physical examination and laboratory tests results in the course of hospitalization revealed no abnormalities. Spirometry confirmed a moderate obstructive picture (FEV1, 1.92 L [59.7%]; FVC, 3.61 L [98.2%]; FEV1/FVC 53.2%). Postbronchodilator spirometry revealed lack of response to inhaled bronchodilators. Body plethysmography showed elevated airway resistance, both inspiratory and expiratory (respectively 305 i 300 % predicted value) and increased residual volume (RV 254%). Blood gasometry and FeNO (7 ppb) were normal. No eosinophils in sputum and nasal discharge were found. Blood eosinophil count was normal. Chest radiograph was unremarkable. The chest CT scan revealed a massively dilated esophagus filled with food residue, which made tracheal compression evident (figure 1). It also revealed parenchymal lung changes: distally located diffused concentration, areas of ground-glass opacity, micronodules and tree-in-bud opacities. Esophagogastroscopy showed a dilated esophagus. Esophageal high resolution manometry (HRM) revealed a hypertensive lower esophageal sphincter (LES) that did not relax on swallowing; there was no peristaltic wave in the esophageal corpus either. The resultant diagnosis was achalasia.

Already after collecting all the tests results, while completing the medical history, doctors were informed by the patient that for at least three years she had suffered from nocturnal vomiting containing undigested food; also her nasal discharge had contained food particles. The symptoms had been associated with a persistent cough.

After establishing the diagnosis of achalasia, the patient was referred to surgery, peroral endoscopic myotomy (POEM). During a control visit, 2 months following the surgery, it was found that the symptoms of cough, dyspnea, running nose and nocturnal vomiting abated.
thoroughly. All asthmatic medications were discontinued. Spirometry normalized (FEV1, 3.89L [121%]; FVC, 4.3 L [117%]; FEV1/FVC, 90%). The bronchial provocation test with methacholine performed at that time was negative.

The presented case concerns a patient with respiratory symptoms resulting from achalasia that were misdiagnosed as severe asthma. As a matter of fact, the reported symptoms were caused by a few years lasting recurrent aspiration of small amounts of gastric contents that occurred largely at night. The chest CT scans performed on the patient’s admission to the Allergy Department were characteristic of bronchiolitis and reflected chronic bronchiolocentric inflammation caused by recurrent aspiration. The clinical picture and imaging scans correspond with the diagnosis of diffuse aspiration bronchiolitis (DAB), complicated by incidents of aspiration pneumonia. The term diffuse aspiration bronchiolitis was first used by Matsuse et al as a name for a chronic inflammation of the bronchioles produced by frequent aspiration of foreign particles [4]. Although DAB is originally diagnosed in the elderly, some studies report its occurrences in young patients with clinical manifestations similar to those in the elderly [5,6,7,8]. In younger patients, the major risk factors responsible for DAB are dysphagia due to achalasia and gastro-esophageal reflux disease (GERD) with concomitant recurrent aspiration.

In the case of the patient in question, the delay in establishing the right diagnosis might have been caused by a few factors. First, apart from vomiting that was erroneously interpreted as a consequence rather than the cause of coughing, for 5 years there were no other accompanying symptoms characteristic of achalasia. Second, auscultatory phenomena were interpreted as "asthmatic wheezing" while in fact they might have resulted from pressure on the trachea and/or might have been caused by bronchiolitis that
is also responsible for obturation changes in spirometry. Third, achalasia is a rare disorder diagnosed mostly in elderly adults (mean time of diagnosis – the 6th decade); its estimated prevalence and incidence being respectively 10.82 cases per 100,000 and 1.63 cases per 100,000 [9]. Forth, the primary symptoms of achalasia are gastrointestinal ones while the respiratory symptoms are less frequent. In up to 40% of achalasia cases, pulmonary problems like cough, wheezing and recurrent aspiration can occur, but DAB is reported very rarely [10]. The correct diagnosis of bronchiolitis associated with chronic aspiration can pose serious difficulties. A diagnosis of DAB should be taken into consideration in patients with respiratory symptoms such as chronic cough, wheezing and obturation, persistent radiologic abnormalities in HRCT and with a high risk of aspiration. Considering the scope of the observed changes in the discussed patient’s respiratory system, the possible consequences of a further delay in performing a surgical treatment of achalasia could be serious. The presented case confirms the prevailing stance of asthma experts who claim that if asthma symptoms persist despite intensive pharmacological treatment, it is advisable to reanalyze the patient’s clinical history, considering the diagnosis of a different disorder that might impersonate asthma.

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**Conflicts of Interest**

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References


Figures legends

Figure 1. Massive dilatation of oesophagus and trachea compression in the chest CT scan.