Clinics and Research in Hepatology and Gastroenterology (2019) 43, e87—e89

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CASE REPORT

Life-threatening acute airway obstruction induced by unsuspected achalasia

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Available online 5 April 2019

KEYWORDS
Achalasia;
Megaesophagus;
Airway obstruction

Achalasia is characterized by the inability of the lower esophageal sphincter to relax in a setting of absent peristalsis. It is a relatively rare affection with an annually reported incidence of approximately 1/100,000 worldwide [1]. Megaesophagus is a sigmoid deformity of the thorax esophagus and an exceptional complication of longstanding achalasia [2]. We present here the case of an acute airway obstruction resulting from megaesophagus with tracheal compression in a patient with unsuspected lower esophageal sphincter (LES) achalasia.

A 49-year-old woman was admitted to the emergency department for acute respiratory failure that occurred immediately after the ingestion of a meal. Acute respiratory failure related to inhaled food was therefore suspected. The patient's medical history was marked by controlled schizophrenia requiring neuroleptic treatment, but showed no history of pulmonary disease or signs of esophageal disease. The patient had never experienced dysphagia and had maintained normal body weight for years preceding acute respiratory failure. Chest auscultation revealed inspiratory and expiratory wheezing with decreased breath sounds bilaterally. The rest of the physical examination was unremarkable. At admission, blood oxygen saturation was 82%, which was corrected using high flow oxygen therapy. Blood samples and electrocardiogram were normal. Nasofibroscopy revealed no pharyngeal or laryngeal abnormalities but noted a stasis of saliva on the upper esophageal sphincter. Chest radiography showed an enlarged mediastinum with pronounced dilatation of the esophagus, filled with food residue, without lung field abnormalities (Fig. 1A). CT (Fig. 1B, 1C) revealed massive distension of the esophagus over a tight esophagogastric junction, with passive compression of the posterior tracheal wall. A tracheal cross-sectional area of 16.4 mm2 was measured on cross-sectional imaging (normal value: 250 mm^2; Fig. 1D), [3]. First-step management included insertion of an aspiration nasogastric probe. At distance, gastroscopy found enlargement of the esophagus without any structural or organic abnormalities, suggesting an esophageal dysmotility disorder. Apersistaltism of the esophagus body was shown using high

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https://doi.org/10.1016/j.clinre.2019.03.003

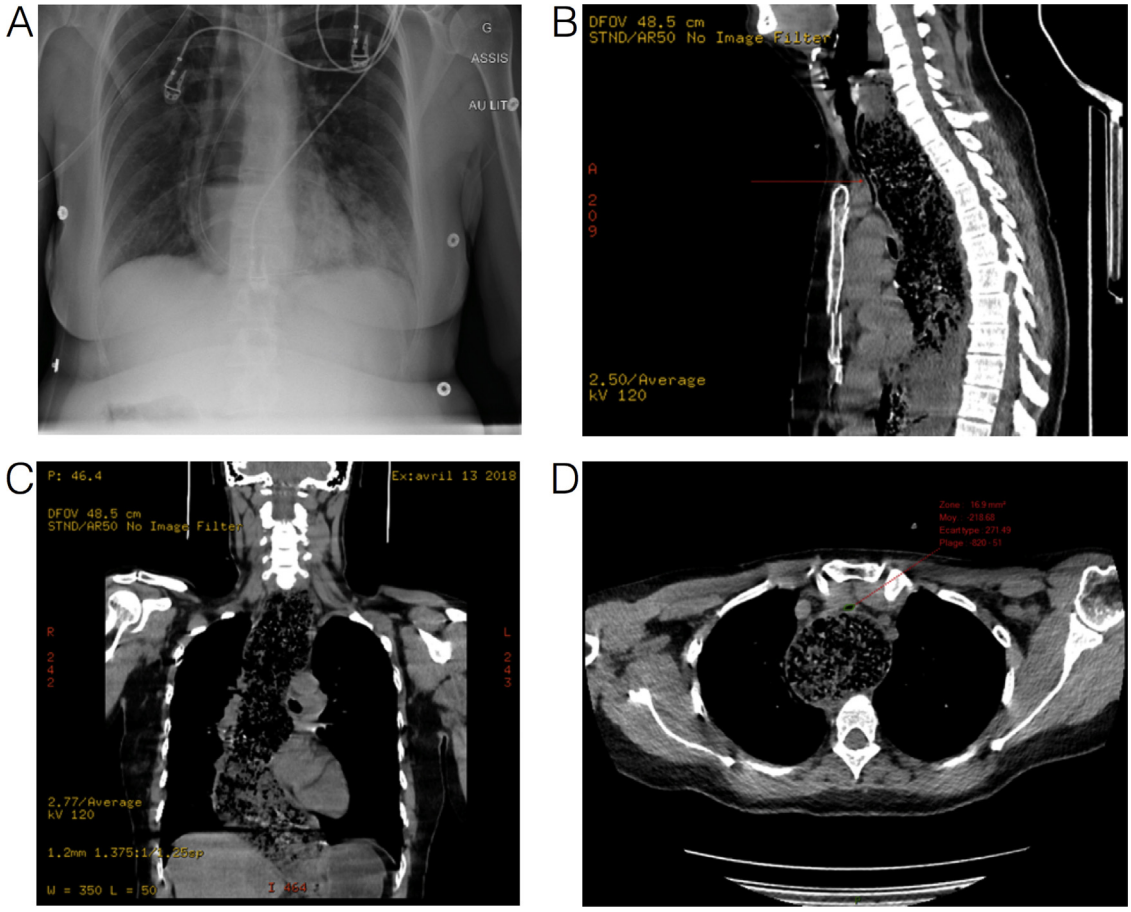

Figure 1 Anteroposterior supine chest X-ray (A) and sagital (B), coronal (C) and axial (D) CT scan showing a massive enlargement of the esophagus extended to the lower esophageal sphincter with tracheal compression (red arrow) at the level of manubrium sterna. At this level, trachea cross sectional area was measured at 16.4 mm^2 (green; normal value of 250 mm^2).

resolution esophageal manometry (Manoscan, Medtronic, Minneapolis, MN, USA) with a median IRP of 9.5 mmHg. Consequently, LES-impaired distensibility was confirmed by esophageal endoFLIP (Functional Lumen Imaging Probe) system (CROSPON, Gallway, Ireland) with a decreased LES distensibility of 2.9 mm^2/mmHg at 50 mL of inflation for normal values between 2.9 and 15 mm^2/mmHg. A final diagnosis of type 1-like LES achalasia was made according to Chicago criteria [4] in complement with LES measurements using EndoFLIP system [5]. The patient had a Per-Oral Endoscopic Myotomy (POEM) 40 cm to 50 cm from the dental arch. At 2 months, the patient's symptoms had not relapsed, with a meal intake of 1750 kcal per day. At 3 months, follow-up gastroscopy was performed with no difficulty traversing the LES. Moreover, the patient returned to regular physical activity by practicing yoga 3 times a week without any respiratory symptom relapse. The patient was still free of symptoms at 7 months of follow-up.

The most common symptoms of achalasia are dysphagia, regurgitation and weight loss, but may be inconsistent, resulting in delayed or missed diagnosis [1]. Respiratory symptoms are also commonly reported by patients with LES achalasia. Indeed, 40% of patients reported the occurrence of at least one respiratory symptom daily like cough and regurgitations [6]. However, these symptoms are due to impaired esophageal clearance of liquid and food, resulting in aspiration, rather than a compressed airway tract.

Neoplasm, inflammatory diseases, extrinsic compression by lymph node or substernal goiter, tracheomalacia, traumatism and strictures that can occur after intubation or surgery, are usual mechanisms of airway obstruction. Tracheal compression due to megaesophagus remains exceptional, with only a few case reports found in the literature [7—16]. In most of these case reports, tracheal compression occurred after the long-term follow-up of previously diagnosed LES achalasia, in aged patients with severe dysphagia. Only two published cases have reported achalasia revealed by acute airway obstruction. The first case presented a patient with a history of chronic respiratory symptoms and weight loss due to the unknown disease in the previous years before acute airway obstruction [11]. The second case described a patient with acute airway obstruction in postoperative context with a history of hiatus hernia and a previous history of digestive symptoms [12]. The originality of the present case resides in the young age of the patient and the absence of digestive or respiratory symptoms in the years before diagnosis. However, the patient had a history of schizophrenia which could have minimized her symptoms, even if the disease was well controlled with treatments.

Rapid decompression of the esophagus using aspiration nasogastric probe is a necessary key in the management of these patients. Follow-up esophageal motility examination is based on the performance of both gastroscopy with mucosal multistage biopsies and esophageal high-resolution manometry [17]. In our case, esophageal manometry revealed acontractility of the esophagus body without abnormality in LES relaxation. In a previous study, systematic evaluation of LES using Endo-FLIP in patients with dysphagia showed that a diagnosis of achalasia could be made when the integrated relaxation pressure was less than 15 mm/Hg [18,19].

There is no curative treatment for achalasia, but several non-invasive surgical treatments are available [2,20]. POEM was performed in our case because of its documented efficacy of 92—97% at one-year follow-up [20,21], instead of pneumatic dilatation balloon with risk of re-intervention and psychiatric relapse. Post-treatment reflux affected 40% of patients among studies [21] but did not occur in our patient. Moreover, pneumatic dilatation and Heller Myotomy have shown worse outcomes than POEM in cases of megaesophagus.

In conclusion, tracheal compression due to megaesophagus resulting from achalasia is an exceptional but potential cause of lethal upper airway obstruction. Here we report a rare case of LES achalasia revealed by acute airway obstruction.

Funding

The authors have no funding to declare.

Disclosure of interest

The authors declare that they have no competing interest.

Contributions

CD collected data and wrote the manuscript, CM, MN, MC and GG contributed to the revision of the manuscript and editing.

Acknowledgments

The authors are grateful to Nikki Sabourin-Gibbs, Rouen University Hospital, for editing the manuscript.

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