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Achalasia with megaesophagus and tracheal compression in a young patient: A case report



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ABSTRACT

INTRODUCTION: Achalasia is one of the most common causes of dysphagia. Typical symptoms include difficulties in controlling the swallowing process, regurgitation, weight loss, and chest pain. A megaesophagus rarely causes tracheal compression with consecutive acute dyspnea or similar respiratory symptoms.

PRESENTATION OF CASE: A 23-year-old male patient presented with difficulties in swallowing, a consecutive massive weight loss over the past three years, and minor respiratory ailments. Further diagnostics revealed a megaesophagus caused by achalasia leading to a severe compression of the trachea.

A laparoscopic Heller myotomy with anterior semi-fundoplication 180° according to Dor was performed.

DISCUSSION: Acute dyspnea and similar respiratory symptoms are rarely observed in patients with achalasia, especially in young patients. Early diagnosis and timely, proper treatment are the hallmarks of restoring esophageal and tracheobronchial function and of successful prevention of severe long-lasting complications of the disease. When not treated properly, the disease may have progressed rapidly, leading to distinct respiratory symptoms such as stridor and acute dyspnea

CONCLUSION: This report emphasizes that physicians should be alert and consider airway obstruction and signs of dyspnea as severe and threatening symptoms in extensive cases of achalasia with megaesophagus. Early surgical treatment provides a therapeutic option to obviate the occurrence of acute respiratory distress and consecutive complications. In particular, difficulties in intubation prior to surgery must be considered.

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1. Introduction

Achalasia is one of the most common causes of motoric dysphagia. The reported incidence is approximately one per 100.000 individuals annually and the prevalence is ten per 100.000. Males and females are equally affected. The incidence increases with age [1]. Histological analysis revealed that achalasia is the result of degeneration of ganglion cells in the myenteric plexus of the esophageal body and the lower esophageal sphincter (LES), leading to an amotile esophageal body as well as to a non-relaxing LES. Current treatment options are symptom-oriented, as the resting and consequently residual pressure of the hypertensive non-relaxing LES are reduced. However, genetic research may further improve our understanding of etiology

and pathogenesis of achalasia and may lead to better treatment options in our tool-box in the future [2,3]. A recently published genetic association study in 1068 achalasia cases and 4242 controls implies that immune-mediated processes are involved in the pathophysiology of achalasia [4]. Current treatment possibilities are laparoscopic Heller myotomy (LHM) with partial fundoplication, per oral endoscopic myotomy (POEM), pneumatic dilation (PD), and endoscopic botulinum toxin injection (EBTI), each with its own magnitude of advantages and disadvantages [5].

While typical symptoms, such as dysphagia, regurgitation, weight loss, and chest pain are the most common complaints patients present with at first diagnosis, this report describes the rare case of a young patient suffering from achalasia and associated megaesophagus leading to massive tracheal compression in its early course of disease, a so called “bull frog neck” [6].

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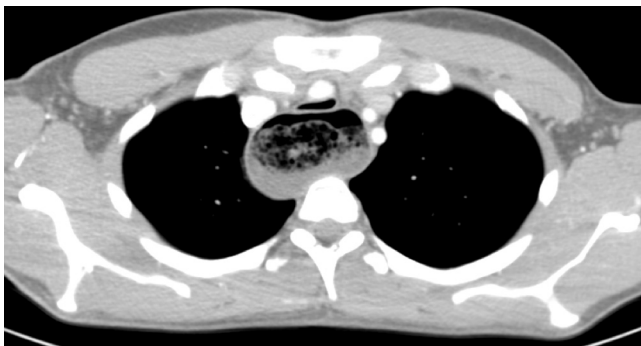


Fig. 1. CT scan of chest showing massive dilation of the esophagus (megaesophagus) and a consecutive slit-shaped narrowing of the trachea [black arrow] (residual tracheal lumen of 3 mm).

2. Case report

A 23-year-old male patient was admitted to our clinic due to progressive difficulties in swallowing and a constant weight loss of 23 kg, which had developed over the past three years. During meals, dysphagia was overcome by drinking water. Additional symptoms were retrosternal pain, regurgitations (Eckardt score of 10), and respiratory ailments [6]. While the laboratory results did not show any abnormalities, a computed tomography (CT) scan of the chest revealed a massive extension of the esophagus starting proximally of the cardia up to cervical vertebral body 7. The consequence of this megaesophagus was a transversal widening of the mediastinum up to 6.7×5.2 cm in diameter accompanied by a consecutive slit-shaped cranial narrowing of the trachea (Fig. 1). Due to the twisted passage of the esophagus, a standard manometry was not feasible. Upper endoscopy did not show any pathological alterations in the mucous membrane of the esophagus, stomach or duodenum.

Due to the age of the patient and his physical complaints we performed a laparoscopic Heller myotomy with anterior semi-fundoplication 180° according to Dor. Because of the profound tracheal stenosis, intubation was done with fiber optic equipment. Intraoperative findings revealed a distinctive thickening of the muscularis at the high-pressure zone of the LES. Surgical myotomy was performed by incision of muscle fibers with a length of 7.0 cm at the distal esophageal body and additional 2 cm at the proximal stomach. Due to the complicated intubation conditions, for safety reasons, the patient was transferred intubated and ventilated to the intensive care unit. Peri- and post-operative courses were uneventful. The patient was extubated one hour after surgery without complications. After the transfer to the patient ward, he was gradually accustomed to normal diet. He initially complained about occasional heartburn, which was successfully treated with proton-pump inhibitors.

Five months after surgery, the technical postoperative findings appeared normal. The physical examination did not reveal any pathology. Body weight remained stable and retrosternal pain and regurgitations disappeared, resulting in an Eckardt score of 1. The patient tolerated normal diet very well. Conventional post-operative esophageal manometry showed a resting pressure of 5.6 mmHg. The barium swallow test detected a residually altered morphology represented by diminished motility and disturbed contractions. In addition, the barium remained for an extended time within the esophageal body and complete emptying of the stomach was only possible by drinking copious amounts of water (Fig. 2). The performed tracheal X-ray imaging did not present a tracheal compression any longer. Overall, the patient was gratefully satisfied with the early postoperative result. Three and a half years after surgery, the structured interview demonstrated good results. Besides minor difficulties and pain in swallowing during



Fig. 2. Barium swallow performed 3 months after surgical treatment. The short-term follow up demonstrates a reduced dilation of the esophagus and some retained barium inside the esophagus.

meals, the patient presented in a very good clinical condition with an Eckardt score of 2. The initially reported symptoms of dysphagia, regurgitation, and respiratory ailments were absent.

3. Discussion

The majority (90%) of patients suffering from achalasia complain about dysphagia for solids and liquids as their primary symptom. Weight loss, regurgitations, chest pain, and heartburn are reported in 40–60% of patients. Occasionally, respiratory symptoms, such as aspiration leading to pneumonia and bronchiectasis, are associated with this disease. Only a few publications report dyspnea and stridor as well as swelling of the neck, also referred to as a “bull frog neck”, associated with advanced stages of achalasia [6,7]. However, even cases of acute total airway compression and death are mentioned in the literature [8].

In the present case, a massive weight loss of 23 kg over three years indicates the severeness of achalasia. Although the CT scan did show an extensively dilated sigmoid-shaped megaesophagus with consecutive compression of the trachea, respiration was not significantly compromised. However, intubation before the surgical intervention was accompanied with relevant difficulties. Thus, fiber optical equipment had to be used to accomplish intubation. For safety reasons, extubation was performed on the intensive care unit. It can be assumed that in older patients and possible respiratory co-morbidities, this distinct finding of a slit-shaped compressed trachea due to megaesophagus might result in serious and possibly life-threatening symptoms also in earlier courses

of the disease. Therefore, it is crucial to consider especially atypical symptoms of achalasia as warning signs and important indicators for timely and proper therapy in order to restore the configuration of megaesophagus as early as possible. The fact that achalasia is a rather rare esophageal disorder, usually diagnosed too late and the symptoms are often misdiagnosed as other foregut disturbances, such as gastroesophageal reflux, which often complicates the time to correct diagnosis for many physicians. However, we recommend that patients with suspicion of achalasia – expressing itself with typical or atypical, especially pulmonary symptoms – are referred for further diagnostic work-up (including high resolution manometry and barium swallow) to centers of excellence at even very discrete stages of foregut symptoms.

The causal connection of massively dilated esophagus in achalasia and airway obstruction could not be fully explained yet. The “pinch-valve” theory assumes that the dilated esophagus may be displaced behind the cricopharyngeal muscle folding over itself. This may lead to a one-way valve trapping the air inside the esophagus. A second theory mentions the disability of the upper esophageal sphincter to relax during the process of swallowing. Furthermore, a loss of the belch reflex, which physiologically causes a relaxation of the LES and a simultaneous upper esophageal sphincter (UES) relaxation, has been discussed as well [9,10].

Only very few cases of tracheal compression due to extensive dilation of the esophagus in achalasia are reported in the literature and therefore represent a rare feature of the disease [11–14]. The radiologic signs and clinical symptoms in these patients may vary and are often unspecific – independently of the stage of achalasia – and thus might be present at even rather early stages of the disease. In the presence of acute airway obstruction in suspected or yet undiagnosed achalasia, early endotracheal intubation with “rapid sequence induction” (RSI; aspiration prophylaxis), emergency esophageal decompression, or local anesthetic tracheostomy are recommended [9,15]. If surgery is planned electively, as in our patient, special care should be devoted to prevent potential anaesthetic complications, especially during intubation.

In the present case, respiratory symptoms were mild at initial diagnosis of achalasia and the time of elective surgery, although severe problems during intubation of the patient were evident. However, if adequate treatment had been delayed, distinct pulmonary complications would have been most likely. Restoration of proper tracheal function and states such as tracheomalacia might have been impossible. Thus, even more severe problems associated with endotracheal intubation and/or postoperative extubation, such as the need for tracheostomy, could have occurred.

4. Conclusion

This report emphasizes that early diagnosis and timely treatment of achalasia is mandatory in order to prevent severe and life-threatening complications, caused by slit-shaped tracheal compression due to megaesophagus, which might remain clinically silent as opposed to upper gastrointestinal key symptoms of achalasia. If diagnosed by radiologic imaging only, signs of advanced tracheal occlusion and/or tracheomalacia in pulmonary not comprised patients should lead to dedicated attention by anaesthesiologists during intubation for surgical interventions, such as laparoscopic myotomy.

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The authors disclose no conflicts.

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Ethical approval

Approval has been given by an ethics committee. Name of the committee: Ethical Committee of the Medical Faculty of the University of Leipzig, Germany Reference number: 043-15-09032015.

Consent

Patient has given consent. No patient details were used. Ethical committee approved studies in achalasia (see above).

Authors contribution

Study concept: Moritz Kathz, Ines Gockel, Hauke Lang. Data collection: Moritz Kathz, Daniel Foltys, Uwe Scheuermann, Mari Stempel, Stefan Niebisch, Maren Ebert, Boris Jansen-Winkel. Writing the paper: Ines Gockel.

Guarantor

Moritz Kathz, Ines Gockel.

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