Clinical Case Report

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Obstructive bronchitis and recurrent pneumonia in esophageal achalasia in a child

A CARE compliant case report

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Abstract

Rationale: Esophageal achalasia is characterized by impaired relaxation of the cardia and dilation of the intrathoracic part of the esophagus. We describe the late presentation of achalasia in an 11-year old girl.

Patient concerns: She suffered from recurrent pneumonia, obstructive bronchitis, and problems with swallowing solid food. Her family noted a wet pillow in the morning.

Diagnoses: This case report describes the typical symptoms of achalasia in children in order to facilitate earlier diagnosis of this rare disease. Our patient was admitted to a pediatric hospital for treatment of severe pneumonia, low-grade fever, and pancreatitis. A computed tomography (CT) scan of the thorax showed massive dilation of the esophagus and infiltration and partial atelectasis of the right lung. An upper gastrointestinal contrast study confirmed massive dilation of the esophagus and stenosis at the level of the cardia.

Interventions: We performed laparoscopic Heller myotomy combined with Dor fundoplication. Bronchoscopic lavages were conducted in the pre- and postoperative period to relief obstruction of bronchi by purulent mucus secretions.

Outcomes: A further upper gastrointestinal contrast study demonstrated patency of the cardia and fast propulsive movement of contrast agent into the stomach. At follow up 2 months after the operation, the girl had gained 3 kg of body weight, and her respiratory, gastrointestinal, and swallowing symptoms had subsided. At follow-up 12 months after the operation, no recurrent symptoms of achalasia were recorded.

Lessons: Late presentation of achalasia in children can mimic respiratory and gastrointestinal diseases. Laparoscopic Heller myotomy combined with Dor fundoplication is feasible and advisable in children suffering from achalasia of the cardia.

Abbreviations: CT = computed tomography, EPD = endoscopic pneumatic dilatation, LHM = laparoscopic Heller myotomy, POEM = peroral endoscopic myotomy.

Keywords: aspiration, child, esophageal achalasia, laparoscopic Heller myotomy, obstructive bronchitis, pneumonia

1. Introduction

Esophageal achalasia is a rare disease in children, characterized by impaired relaxation of the cardia and subsequent dilation of the intrathoracic part of the esophagus. The overall incidence of achalasia is 1.6 per 100,000 individuals.[1] In the pediatric age group, the incidence is only 0.11 per 100,000 children. Less than 5% of cases occur in patients below the age of 15 years.[2–4] Because of the rareness of this congenital disease and lack of experience of treatment in children, diagnosis may be delayed for a significant period of time. We report the case of an adolescent girl who suffered from the symptom of "wet pillow" and severe pulmonary infection. The aim of this case study is to describe the typical symptoms and successful treatment of late-presenting achalasia to make physicians aware of this rare and curable gastrointestinal disorder.

Ethical approval was not necessary for this case study. We obtained written informed consent from the parents of the girl.

2. Case presentation

The 11-year old girl was hospitalized for 3.5 weeks for the treatment of recurrent right-sided focal pneumonia, reactive pancreatitis, and swallowing problems. Anamnesis revealed that she had suffered from obstructive bronchitis for several years occurring especially at night, low-grade fever, and recurrent headache. She complained of problems when swallowing solid food and drank large amounts of water during her meals to support swallowing the ingested food. The family observed that her pillow was wet when she got up in the morning. Prior to this hospitalization, she was treated in several outpatient clinics for symptoms of recurrent viral bronchitis.

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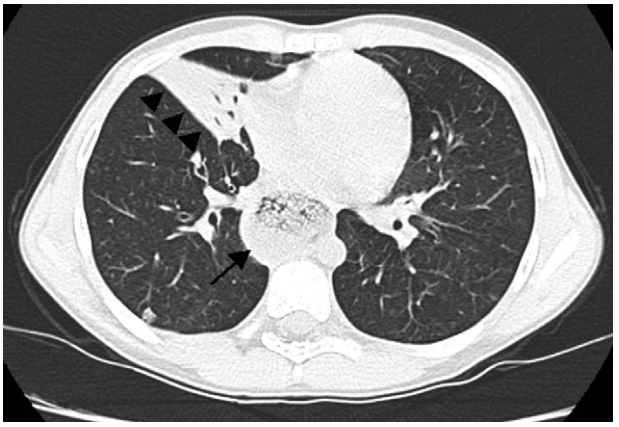

Figure 1. Multislice computed tomography (CT) of the chest. The middle lobe of the right lung is reduced in volume, caused by subtotal atelectasis. Bronchi appear obstructed (arrow heads). The esophagus appears massively dilated. There is hypodense content in the lumen of the esophagus (arrow). CT = computed tomography.

Despite antibiotic treatment and supportive care, her respiratory and swallowing problems did not improve. Therefore, a computed tomography (CT) scan of her chest was obtained which revealed right-sided pneumonia. Aspiration pneumonia was suspected. The CT scan indicated significant dilation of the intrathoracic esophagus (Fig. 1). The right main bronchus appeared obstructed and an endobronchial foreign body was suspected. The child was transferred to the Department of Paediatric Surgery at MONIKI University Hospital, Moscow, Russia, for further treatment.

At admission at MONIKI University Hospital, the girl's body weight was 32 kg (body weight at 25th percentile for age). She appeared minimally malnourished and her respiration rate was increased to 28 breaths/min (normal respiration rate in school children: 18 to 25 breaths per minute).[5] Body core temperature at admission was 36.8°C. She presented with symptoms of severe obstructive bronchitis, and coughing produced small amounts of sputum. On auscultation, wheezing was noted on the right side. Her pulse rate was 95/min, and her blood pressure was 110/80 mm Hg. Auscultation of the heart was normal. Her abdomen appeared nontender, and we noted no signs of peritonitis.

3. Investigations

In the upper gastrointestinal contrast study, the intrathoracic esophagus appeared massively dilated, and we noted a cone-shaped narrowing to 3 mm for a distance of 2.5 cm at the distal end of the esophagus. Dynamic image intensifier imaging revealed massive hypoperistalsis of the esophageal wall. Reliable propulsion of contrast agent into the stomach was not observed (Fig. 2). Based on the typical radiographic study findings,[6] we established the diagnosis of achalasia of the esophagus. At gastroscopy, we found a large amount of residual food retained in the esophagus. Insertion of the endoscope into the stomach was easily achieved and the stomach appeared empty. We performed bronchoscopy and noted purulent mucus accumulation within the lumen of the right main bronchus. The right upper lobe bronchus appeared normal. However, the middle lobe and lower lobe bronchi of the right lung were dilated, and the segment bronchi were also enlarged and tightly packed with purulent mucus. The mucosal lining of bronchi appeared inflamed. On the left side, there was moderate dilation of the lower lobe bronchus and segmental bronchi, with inflamed mucosa and large amounts of purulent mucus within bronchi. Bronchoscopic diagnosis was bilateral purulent endobronchitis and beginning formation of bronchiectasis.

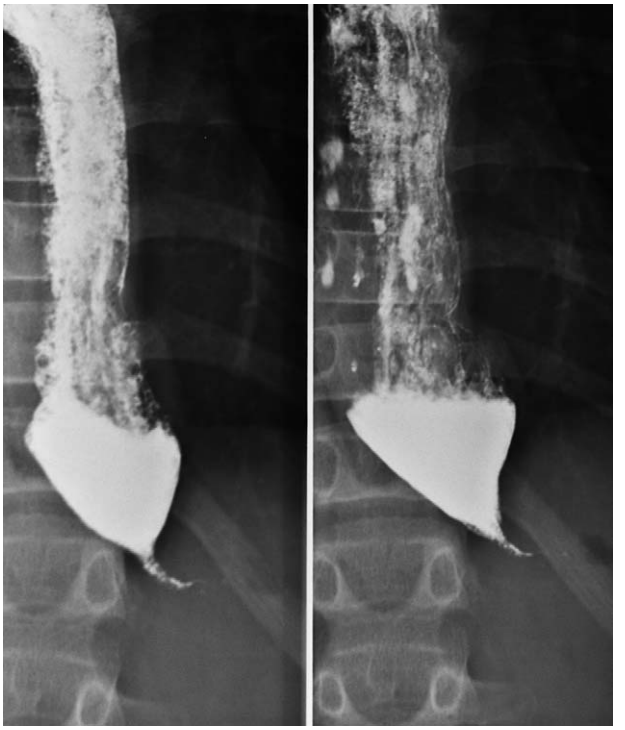

Figure 2. Contrast study of the esophagus. Note the significant dilation of the intrathoracic esophagus. The passage of contrast agent is blocked at the level of the cardia. Note the characteristic constriction of the cardia in the form of a "beak" or "the tip of the carrot."

4. Differential diagnosis

Differential diagnoses of achalasia of the esophagus complicated by aspiration pneumonia comprised gastroesophageal reflux disease, congenital stricture of the esophagus, acquired stricture of the esophagus, esophageal membrane, esophageal diverticulum, esophageal foreign body, tumors of the lower esophageal sphincter, and anorexia.

5. Treatment

Given the high risk of exacerbation of pneumonia in the postoperative period, we treated the child with antibiotics and performed bronchoscopic lavage to reduce the amount of purulent mucus within the bronchi before the operation. After improvement of the bronchopulmonary symptoms, we opted for a laparoscopic Heller myotomy (LHM) combined with Dor fundoplication, maintaining the option to switch from the laparoscopic approach to an open approach in case of intraoperative deterioration of pulmonary function. In accordance with the recommendation by Chuah et al,[6] we opted for LHM which represents a safe and highly efficient treatment of achalasia in children. LHM was most effective in relieving the symptoms of achalasia at the expense of increased gastroesophageal reflux.[6] Therefore, we decided to add Dor fundoplication

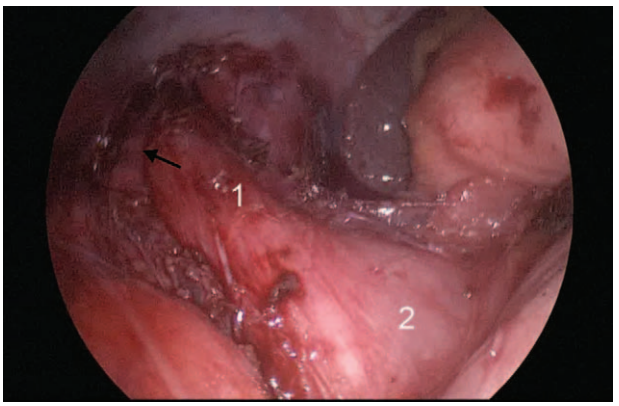

Figure 3. Photograph obtained after mobilization of the esophagus and cardia. 1—esophagus, 2— stomach; anterior branch of the vagus nerve retracted laterally (arrow).

to LHM. After the preoperative preparation, we performed a laparoscopic Heller myotomy combined with Dor fundoplication.

5.1. Surgical technique

Under general anesthesia, we created a small infraumbilical laparotomy for insertion of a 5 mm trocar and insufflated carboperitoneum to 12 mm Hg. Under laparoscopic visualization, we inserted two 5 mm trocars in the midclavicular line in the right and left epigastrium. In the midline of the epigastrium, halfway between xiphoid process and umbilicus, we inserted a 5 mm retractor to facilitate elevation of the left lobe of the liver. We obtained good visualization of the cardia. We dissected the peritoneum overlying the cardia. By blunt and sharp dissection, we created the myotomy of the cardia on the lesser curvature of the stomach and distal esophagus to a distance of 5 cm above the hiatus of the diaphragm. We identified the anterior branch of the vagus nerve and retracted it laterally (Fig. 3). The operation was completed by the creation of Dor fundoplication (Fig. 4). We placed a silicone drain in the subhepatic space and closed the incisions with interrupted sutures. There was only minimal intraoperative blood loss. The operation lasted 160 minutes.

On the 1st day after the operation, the girl was transferred from the intensive care unit to the pediatric surgical ward. The postoperative course was complicated by acute pancreatitis (maximum alpha-amylase in serum: 811 U/L; normal value: 20–112 U/L) and occurrence of a pleural effusion at the left side. Therefore, we performed drainage of the left pleural cavity and evacuated 200 mL of serous fluid. Due to the massive purulent endobronchitis, we undertook 2 bronchoscopic interventions in the postoperative period to reduce the amount of endobronchial mucus within the bronchi. The need for analgesics persisted for 3 days, and we administered antibiotic therapy for 1 week. The girl also received a short course of synthetic somatostatin analogue to treat pancreatitis. We started enteral nutrition 2 days after operation, and there were no more signs of dysphagia. We removed the drains 2 days after the operation. Serum alpha (α)-amylase values normalized 1 week after the operation. The girl was discharged from hospital 14 days after the operation.

6. Outcome and follow-up

At the follow-up visit 2 months after the operation, the girl did not complain of any symptoms. Serum alpha (α) amylase level was within normal limits, and the girl did not complain about abdominal pain and tolerated feedings well, thus clinically excluding pancreatitis. The family noted a significant improvement in health. There was no more coughing at night, dysphagia, or headache. Auscultation of breathing sounds over all parts of the lungs was normal, the rales had stopped. The child and her family reported that she showed no more signs of obstructive

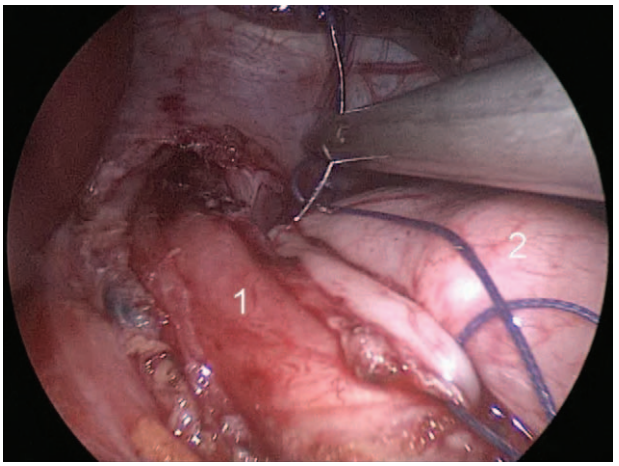

Figure 4. Photograph showing laparoscopic myotomy according to Heller combined with Dor fundoplication. 1—area of myotomy of the cardia, 2—fundus of the stomach.

Figure 5. Contrast study of esophagus with barium sulphate obtained 2 months after surgery. The lumen of the esophagus is significantly increased, and there is no obstruction of the esophageal lumen.

Figure 6. Appearance of the child 2 months after surgery.

bronchitis or pneumonia after the operation. Even headache was no longer present. The girl had gained 3 kg in body weight, and the weight percentile for age increased from 25th to 50th percentile. An upper gastrointestinal contrast study demonstrated significant improvement of the opening width of the cardia and no impairment of the propulsion of contrast agent from the distal esophagus into the stomach (Fig. 5). Laboratory tests did not reveal any signs of inflammation. Figure 6 shows the abdomen of the child 2 months after the operation. At the follow-up visit 1 year after the operation, the clinical investigation did not reveal any bronchopulmonary symptoms, and the child did not complain of any swallowing problems. The family reported that no signs of bronchopulmonary infection and abdominal pain had occurred in the interval since the last follow-up.

7. Discussion

Achalasia of the esophagus in children is a rare disease. In our patient, parents noted a wet pillow in the morning and other typical clinical symptoms of achalasia, such as obstructive bronchitis, recurrent pneumonia, and loss of body weight.[7] It must be kept in mind that similar to Hirschsprung disease, the deficient neurons in achalasia cannot be cured. The aim of treatment is to improve symptoms, to facilitate adequate emptying of the distal esophagus, and to avoid progressive dilation of the proximal part of the esophagus.[8]

Currently, no generally accepted guidelines for the treatment of achalasia of the esophagus in children are available. According to the literature, the operation most frequently conducted is myotomy of the cardia according to Heller, supplemented by one of the variants of fundoplication.[9–13]

However, LHM has evolved as the preferred treatment of esophageal achalasia in children and adults.[6,13] In a review of the literature in patients under 18 years of age, Pandian et al[13] noted that the pediatric LHM experience comprises only retrospective studies suggesting that LHM is safe and effective.[13] Unfortunately, long-term studies of achalasia in children are extremely rare.

Conservative methods of treatment in adult patients are not applicable in children (e.g., administration of calcium-channel blockers) or have only a temporary effect. Endoscopic interventions, that is, balloon dilation of the cardia or injection of botulinum toxin (Botox), can result in scarring of the distal esophagus and occurrence of complications.[9,13] In children, endoscopic pneumatic dilatation (EPD) has been applied for many years with varying rates of success. EPD may result in recurrences of esophageal obstruction necessitating repeated EPD.[14,15] In children suffering from esophageal achalasia, previous EPD and Botox injections are considered risk factors for perforation when performing LHM.[16,17] Mucosal perforation detected during LHM should be repaired immediately using absorbable sutures. In contrast, it seems not very reliable to discern between a full thickness tear and a partial tear of the esophageal wall during EPD and therefore, esophageal perforations may remain undetected, resulting in severe mediastinitis. Young age at first symptoms and classic type of esophageal achalasia are prognostic parameters for the need of repeated treatment when applying EPD.[18] The overall recurrence rate after EPD is 33%.[18]

The necessity to add partial fundoplication to LHM in children is discussed controversially. The rate of gastroesophageal reflux after LHM appears low.[16,19] Anterior partial fundoplication according to Dor is the preferred type of fundoplication in children undergoing LHM.[13,17,20–23] We opted for laparoscopic myotomy according to Heller combined with Dor fundoplication, which helped to eliminate the girl's symptoms of dysphagia and aspiration pneumonia and improved her quality of life.

The innovative treatment approach of peroral endoscopic myotomy (POEM) for achalasia is performed by experienced endoscopists and represents a new, scarless treatment option for esophageal achalasia.[24] Comparing the outcome between a group of children treated by POEM with a group of children treated by LHM, Caldaro et al[20] noted shorter operative time, longer distance of myotomy, and earlier tolerance of oral feeding in the first group.

We describe a rare disease in a child aged 11-years with a long history of symptoms caused by late presentation of achalasia of the esophagus. Achalasia was not diagnosed at the outpatient stage of treatment, which led to a delay in establishing the correct diagnosis and complications, such as obstructive bronchitis, recurrent (aspiration) pneumonia, recurrent vomiting, loss of body weight, pancreatitis, and recurrent headache.

In hindsight, Botox injection and (repeated) EPD would have been an alternative option in our patient, who presented in a poor pulmonal and physical condition initially. However, after treatment of bronchopulmonary symptoms, LHM was carried out without significant complications and with a good short-term outcome. Whether the addition of Dor fundoplication to LHM helped to achieve the good outcome in our patient remains

uncertain. Minimally invasive laparoscopic operation combined with bronchoscopic interventions helped to reduce surgical trauma and facilitated shortening of the postoperative rehabilitation period in this child. The cosmetic result of the laparoscopic intervention was better than that after open procedures.

8. Conclusion

Esophageal achalasia is a rare disease in children. Delayed diagnosis of achalasia can cause recurrent aspiration pneumonia, bronchiectasis, poor weight gain, pancreatitis, and headache. Laparoscopic myotomy of the cardia according to Heller, supplemented by one of the variants of fundoplication, is feasible and advisable in children. Laparoscopic techniques reduce surgical trauma, shorten the postoperative period of rehabilitation, and improve the cosmetic outcome.

Author contributions

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Writing – original draft: Alexandr Viktorovic Sigarchev.
Writing – review & editing: Johannes Michael Mayr, Alexandr Evgen'evich Mashkov, Andrei Viktorovich Bobylev.

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