Clinical case

Medical Ultrasonography
2010, Vol. 12, no. 1, 66-70

Clinical and ultrasound approach to achalasia in a child. Case report

Daniela Iacob[1], Otilia Fufezan[1], Dorin Farcău[1], Natalia Hagău[2], Constantin Ciuce[3]

[1] University of Medicine and Pharmacy „Iuliu Haţieganu", III[rd] Pediatric Clinic, Cluj-Napoca
[2] University of Medicine and Pharmacy „Iuliu Haţieganu", Department of Anaesthesia, I[rst] Surgical Clinic, Cluj-Napoca
[3] University of Medicine and Pharmacy „Iuliu Haţieganu", I[rst] Surgical Clinic, Cluj-Napoca

Abstract

Achalasia is a primary esophageal motor disorder of unknown etiology characterized by insufficient lower esophageal sphincter (LES) relaxation in response to deglution. We present the case of a child diagnosed with asthma, with no response to asthma treatment. On account of associating dysphagia, regurgitation of food and weight loss he was given an abdominal ultrasound that revealed a massive dilated and narrowing of its distal segment esophagus. Ultrasound examination proved to be a good diagnostic tool, reorienting from the initial asthma diagnosis to the correct achalasia diagnosis, confirmed through barium swallow, upper digestive endoscopy and esophageal manometry. Esocardiomiotomy with antireflux technique was performed with excellent results.

Key words: achalasia, ultrasound, child

Rezumat

Achalazia reprezintă o afecţiune motorie primitivă a esofagului de etiologie neprecizată caracterizată prin relaxarea insuficientă a sfincterului esofagian inferior în timpul deglutiţiei. Se prezintă cazul unui copil diagnosticat cu astm, fără răspuns la terapie. Asocierea disfagiei, regurgitărilor pentru alimente şi scăderii în greutate au impus efectuarea ecografiei abdominale. S-a evidenţiat un esofag dilatat, cu îngustare distală. Ecografia abdominală a permis reorientarea diagnosticului iniţial de astm bronşic spre diagnosticul corect de achalazie. Diagnosticul a fost confirmat prin esofag baritat, endoscopie digestivă superioară şi manometrie esofagiană. S-a practicat esocardiomiotomie cu procedură antireflux cu evoluţie foarte bună.

Cuvinte cheie: achalazie, ecografie, copil

Introduction

Idiopathic achalasia is a rare primary motility disorder of the esophagus of unknown etiology characterized by esophageal aperistalsis and abnormal lower esophageal sphincter (LES) relaxation in response to deglutition. The disease was first described in 1674 by Sir Thomas Williams. In 1927 Arthur Hurst first named this disease achalasia, meaning in Greek „lack of relaxation". Achalasia is a rare disease with an annual incidence of 1/100.000 and a prevalence rate of 10/100.000. It may occur at any age, affecting both genders, being usually diagnosed between 25 and 60 years of life [1,2].

In the pathogenesis of this disease a degeneration of the myenteric plexus has been reported, resulting in a lack of inhibitory neurons necessary for coordinating the LES relaxation and peristaltic contraction of the esophagus. The neurodegenerative insult is believed to be genetic, autoimmune, infectious, but a definite trigger has not been identified. The motor abnormalities result in stasis of ingested food, leading to clinical symptoms, such

Received 14.12.2009 Accepted 22.01.2010
Med Ultrason, 2010
Vol. 12, No 1, 66-70
Address for correspondence: Daniela Iacob
III[rd] Pediatric Clinic
University of Medicine and Pharmacy
„Iuliu Haţieganu"
Câmpeni street nr. 2-4
Cluj-Napoca, Romania
E-mail: iacobdaniela777@gmail.com

as dysphagia, regurgitation of food, retrosternal pain and weight loss.

The diagnosis of achalasia is suggested by clinical features and confirmed by diagnostic tests, such as upper digestive endoscopy (UDE), esophageal manometry and barium swallow.

Treatment goals in achieving long-term symptom relief are obtained through endoscopic methods and surgery. Many studies have proved that laparoscopic Heller myotomy with partial fundoplication is superior to endoscopic methods in maintaining patients long-term symptom free [3,4].

We report a case that illustrates the valuable contribution of abdominal ultrasound in a pediatric patient with achalasia.

Case Report

A 9-year-old boy presented for cough, dysphagia, vomits and weight loss. Family history and past medical history were uneventful. One year before the admission he developed a persistent cough. Diagnosed with asthma (prick test for allergens proving allergy to dust, Alternaria, and dog hair), he was treated with inhaled β_2-adrenergic receptor agonist, inhaled glucocorticoid and oral antihistaminic drug. The symptoms persisted despite this combined therapy. After 3 months he associated decreased appetite and early satiety. One month prior to the actual admission he developed vomiting, dysphagia and epigastralgia. Diagnosed with *Helicobacter pylori* gastritis, he received proton pump inhibitor and antibiotics. The worsening of his respiratory and digestive symptoms with weight loss made his admission to a pediatric unit compulsory.

On presentation his weight was 28 kg (P 25), with a weight loss of 3 kg in the last 10 months. His body mass index was 14. Afebrile, he presented a frequent cough but no rales, normal breathing sounds. He had daily vomiting with undigested food and dysphagia. His swallowing was difficult, both for solid food and fluid. He presented days with 3-4 normal stools alternating with periods of constipation (one stool at 3-4 days).

The chest x-ray revealed an enlarged mediastinum, with a convex linear opacity on the right side. No hyperinflation of lungs was found (fig1). The spirometry was not interpretable, due to frequent coughing.

Ultrasound examination of the abdomen with a SA 8000 EX machine revealed a massive dilated esophagus with a narrowing of its distal segment. The content of the dilated esophagus was heterogeneous: in the most fluid content, air and debris foci presented a to-and-fro motion (fig 2). The suprasternal view of the esophagus proved the dilation of the upper esophagus with the same heterogeneous content (fig 3).

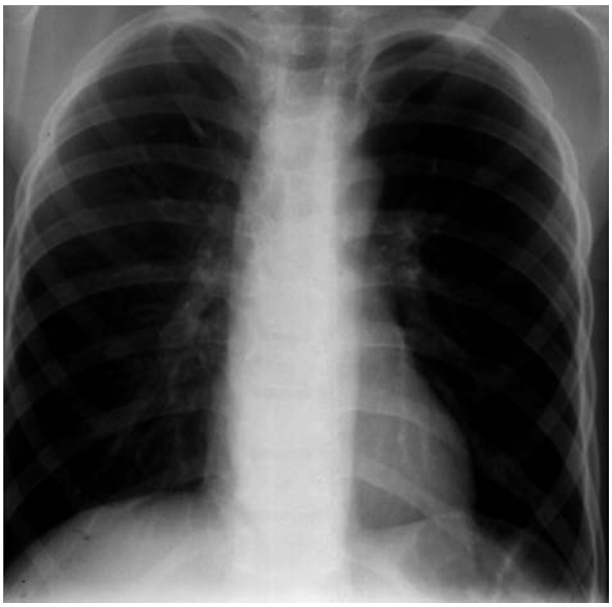


Fig 1. Chest x-ray. Enlarged mediastinum, with a convex linear opacity on the right side.

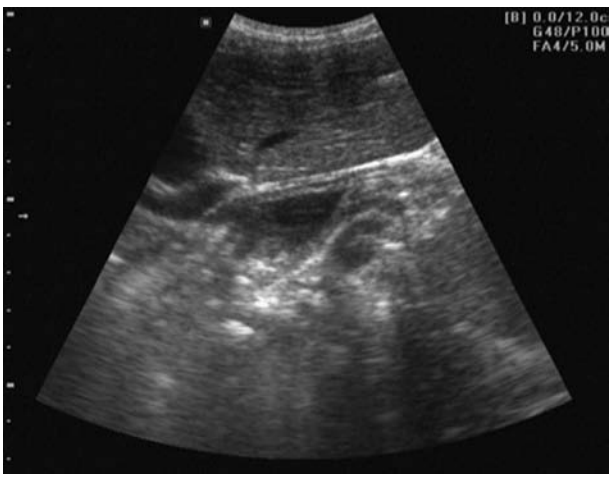


Fig 2. Abdominal ultrasound: massive dilated esophagus, narrowing in its distal area.

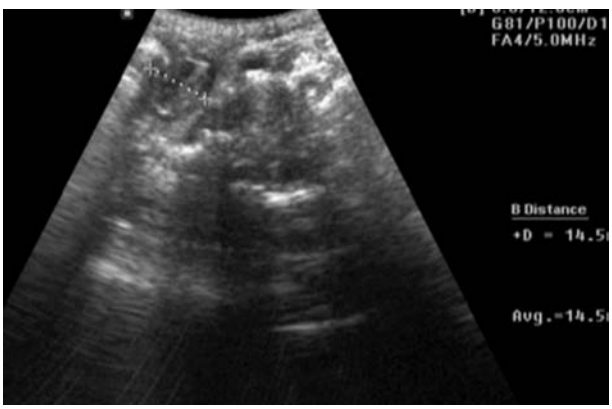


Fig 3. Ultrasound, suprasternal view: dilated upper esophagus.

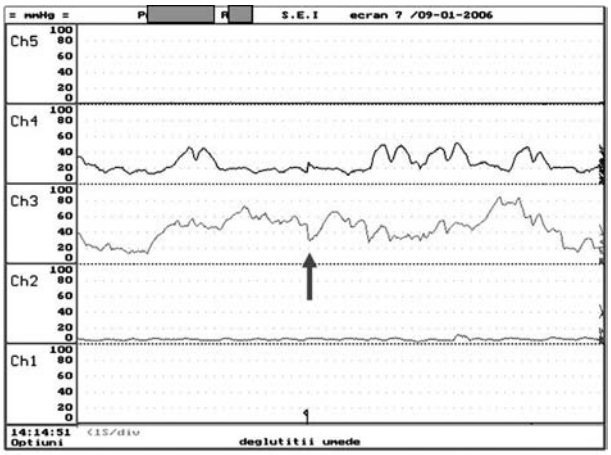


Fig 4. Barium swallow. Dilated esophagus with „bird's beak" appearance suggestive for achalasia.



Fig 5. Esophageal manometry: incomplete relaxation of lower esophageal sphincter after a wet swallowing (arrow).

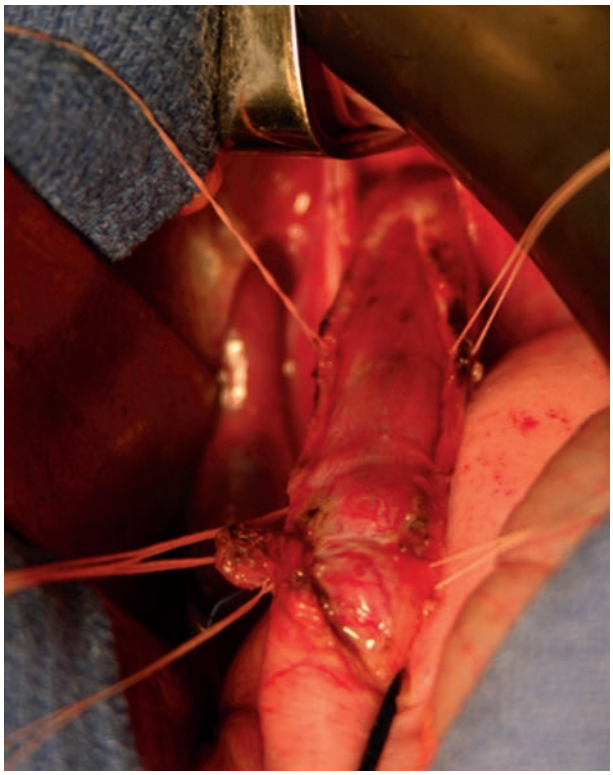


Fig 6. Surgical procedure: esocardiomiotomy.

The barium esophagogram confirmed the massive dilation of the esophageal body with a heterogeneous air-fluid level at the top of the barium column. The typical radiographic finding of smooth tapering in the distal esophagus with the typical „bird's beak" appearance was found (fig 4). The barium esophagogram documented also a delayed esophageal and gastric emptying.

The UDE revealed a dilated esophageal body, allover his length, retention of food and secretion, diffuse congestion of esophageal mucosa. No hiatal hernia was found.

The esophageal manometry measured an upper esophageal sphincter pressure of 60-100 mmHg (3-4 times greater than normal values) and a LES pressure of 40-75 mmHg (twice as normal). The esophageal peristaltic was normal. The patient presented an incomplete relaxation at wet swallow (fig 5).

The diagnosis of achalasia was established and the patient was transferred to the Surgical Department. The surgical procedure consisted of esocardiomiotomy with antireflux technique (fig 6, fig 7). The patient had a rapid recovery with no vomiting, weight gain and disappearance of asthma symptoms. He remained completely asymptomatic in the four years follow up period after the surgical treatment.

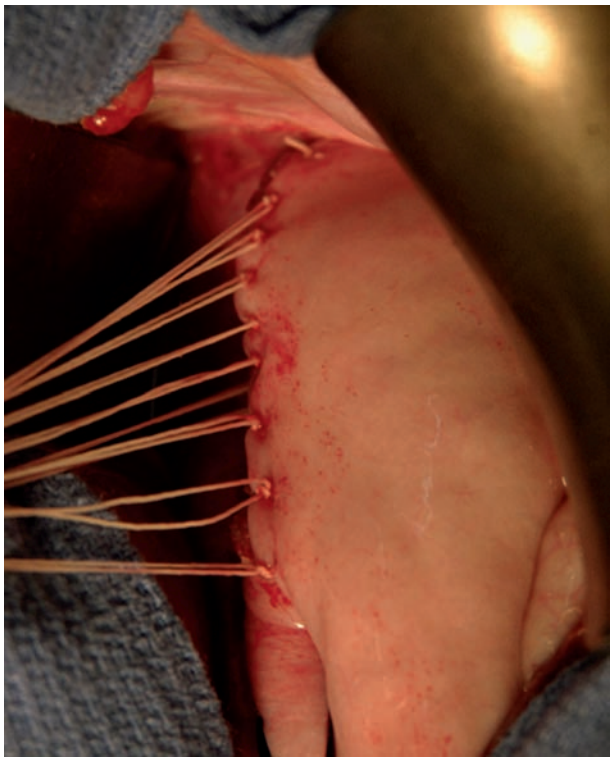


Fig 7. Surgical procedure: esocardiomiotomy with antireflux technique.

Discussion

Idiopathic achalasia is a rare primary motility disorder of the esophagus. The classical features are incomplete relaxation of a hypertensive LES and a lack of peristalsis in the tubular esophagus.

The motor abnormalities in achalasia lead to dysphagia for solids and liquids (97-100%), stasis, regurgitation (59-64%), weight loss (30-91%), chest pain (17-95%), nocturnal cough (11-46%), and heartburn [3]. Like most achalasia patients, our case developed progressively dysphagia for solids and liquids, stasis, regurgitation, vomiting and weight loss. Regurgitation of undigested, retained food occurs most commonly in the recumbent position, awaking the patient from sleep because of coughing and choking. Our patient presented a persistent cough, being diagnosed and treated for asthma, with no clinical benefit. The persistent cough was most probable due to recurrent regurgitation of food from the dilated esophagus.

Due to the similarity of symptoms, achalasia can be mistaken for more common disorders, such as gastroesophageal reflux disease, hiatal hernia, pseudoachalasia, asthma, psychosomatic disorders.

Once the clinical suspicion of achalasia arises, UDE, esophageal manometry and barium swallow are the primary tools of investigation. UDE might reveal a dilated esophagus, retention of food and secretions. In our case UDE showed all these aspects. In about 44% of patients with achalasia UDE can show normal aspect [3]. The esophagogastric junction can usually be easily traversed, despite the elevated LES pressure [3]. This pattern was also found in our patient. Pseudoachalasia, usually neoplastic, can be better differentiated through transabdominal ultrasound.

The abdominal ultrasound examination of our patient revealed a dilated esophagus, narrowing in its distal area, reorienting the investigations and diagnosis from a lung pathology to a digestive disorder.

In achalasia, the barium esophagogram (barium swallow) shows a dilation of proximal esophagus, lack of peristalsis during fluoroscopy and the typical finding of smooth tapering of the lower esophagus, resembling a „bird's beak" [1,3]. Our patient presented dilated esophagus, poor emptying with retained food and saliva and the „bird's beak" sign. Biopsy, if performed, shows hypertrophied musculature and absence of some nerve cells of the myenteric plexus.

Esophageal manometry, the gold standard diagnostic investigation in achalasia, reveals three essential features: aperistalsis of the smooth muscle area of esophagus, incomplete relaxation of the lower esophageal sphincter and elevated resting pressure of the lower esophageal sphincter [3]. In our case the esophageal manometry documented increased resting pressure of the lower, but also upper esophageal sphincter, partially relaxation of the LES during wet swallows. A new diagnostic method, high resolution manometry, might allow the prediction of treatment responses [3].

The treatment of esophageal achalasia is only palliative, aiming to decrease the pressure gradient across LES and to facilitate food bolus passage.

Patients with untreated achalasia will develop a dilated esophagus, increasing the risk for aspiration pneumonia, esophageal cancer or organ perforation. Structural or functional pulmonary abnormalities occur in about 50% of patients with achalasia, due to recurrent microaspiration or tracheal compression from a dilated esophagus. The most feared complication of achalasia is esophageal cancer [3].

Pharmacologic treatment of achalasia includes smooth muscle relaxants such as channel blockers, nitrates and phosphodiesterase inhibitors. These pharmacologic therapies have a limited value in achalasia.

Based on meta-analysis of treatment trials, studying data including remission rate, relapse rate, complications and adverse effects most authors proved that laparoscopic myotomy combined with an antireflux fundoplication is the treatment of choice, in comparison with pneumatic dilata-

tions or botulinum toxin injection [1,4,5,6,7]. Comparing efficacy of various treatments for achalasia through the effects on esophageal body function (peristalsis and clearance) using combined manometry (peristalsis) with Multichannel Intraluminal Impedance (MII) (esophageal clearance) Tatum et al describes restoration in peristalsis as well as improved bolus clearance after Heller myotomy [8].

We presented this case because ultrasound examination proved to be the early diagnostic tool, reorienting from the initial asthma diagnosis to the correct achalasia diagnosis, confirmed through barium swallow, upper digestive endoscopy and esophageal manometry. Esocardiomiotomy with antireflux technique represents an excellent treatment option.

References

1. Farrokhi F, Vaezi MF. Idiopathic (primary) achalasia. Orphanet J Rare Diss 2007; 2: 38.
2. Pohl D, Tutuian R. Achalasia: an overview of diagnosis and treatment. J Gastrointestin Liver Dis 2007; 3: 297-303.
3. Eckardt AJ, Eckardt VF. Current clinical approach to achalasia. World J Gastroenterol 2009; 15: 3969-3975.
4. Richards WO, Torquati A, Holzman MD, et al. Heller myotomy versus Heller myotomy with Dor fundoplication for achalasia: a prospective randomized double-blind clinical trial. Ann Surg 2004; 240: 405-412.
5. Campos GM, Vittinghoff E, Rabl C, et al. Endoscopic and surgical treatments for achalasia: a systematic review and meta-analysis. Ann Surg 2009; 249: 45-57.
6. Vaezi MF, Richter JE. Diagnosis and management of achalasia. American College of Gastroenterology Practice Parameter Committee. Am J Gastroenterol 1999; 94: 3406-3412.
7. Wang L, Li YM, Li L, Yu CH. A systematic review and meta-analysis of the Chinese literature for the treatment of achalasia. World J Gastroenterol 2008; 14:5 900-5906.
8. Tatum RP, Wong JA, Figueredo EJ, Martin V, Oelschlager BK. Return of esophageal function after treatment for achalasia as determined by impedance-manometry. J Gastrointest Surg 2007; 11: 1403-1409.