"Stray" achalasia: From gastroenterologist to pulmonologist and back

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SUMMARY

Introduction Achalasia is a rare esophageal disorder which, due to frequent presence of both respiratory and gastrointestinal symptoms, can initially be referred either to pulmonologist or gastroenterologist.

Case Outline A 50-year-old patient was initially referred to gastroenterologist with the following symptoms: nausea, vomiting, occasional hiccups, weight loss, chest pain, dysphonia, and dry cough. After chest X-ray, the patient was referred to pulmonologist with differential diagnosis for pulmonary infiltration and thoracic aortic aneurysm. Pulmonologist interpreted chest X-ray as showing paratracheal mediastinal enlargement with air-fluid levels, thus suspecting achalasia. Computed tomography scan of the thorax with per os contrast showed extremely dilated esophagus with food stasis. The patient was then referred to thoracic surgeon, who ordered additional diagnostics (esophageal passage with contrast, esophagomanometry, esophagogastroduodenoscopy), and finally performed Heller myotomy. Postoperatively there were no complications, and the patient was symptom free during the follow-up.

Conclusion Although achalasia can also result in respiratory symptoms, fastidious anamnesis and accurate radiological interpretation are essential for the correct diagnosis.

Keywords: achalasia; chest pain; dysphonia

INTRODUCTION

Achalasia or "cardiospasm" is an esophageal disorder, characterized by incomplete or absent relaxation of lower esophageal sphincter (LES) and the loss of esophageal peristalsis [1]. Consequently, LES does not relax after food intake – it stays contracted, which prevents normal food passage. Food stasis then leads to esophageal dilatation. This disorder is more common in male patients, with peak incidence between 30 and 60 years of age. The symptoms are primarily gastrointestinal: difficulty swallowing both solids and liquids, food regurgitation and weight loss. However, chest pain, dysphonia and chronic cough are also frequent symptoms. Differential diagnosis includes gastroesophageal reflux disease [2], pseudoachalasia due to malignant infiltration of myenteric plexus [3] (adenocarcinoma of gastroesophageal junction, pancreatic carcinoma, lung carcinoma), or secondary achalasia due to previous surgical interventions on esophagus [4]. Achalasia can also be diagnosed as part of Chagas disease (*Trypanosoma cruzi* infection) [5].

Patients with achalasia are usually diagnosed by gastroenterologists, although there are cases in which respiratory symptoms such as chronic cough or recurring pneumonias dominate, urging pulmonologists to consider this condition.

CASE REPORT

A 50-year-old patient was initially referred to a gastroenterologist due to chronic complaints: difficulty swallowing, nausea, vomitus, occasional hiccups, and weight loss, followed by more recent symptoms – chest pain, dysphonia, and cough. However, the patient focused on more recent symptoms, those being chest pain, cough and dysphonia, neglecting gastrointestinal symptoms, which turned out to be present for almost ten years. Gastroenterologist ordered chest X-ray, and referred the patient to a pulmonologist suspecting pulmonary infiltration or thoracic aorta aneurysm. The pulmonologist interpreted the chest radiogram as paratracheal enlargement of mediastinum with two air-fluid levels and marked absence of a gastric air bubble, thus suspecting achalasia (Figure 1). The lateral chest radiography showed dilated esophagus with air-fluid level and compression on trachea, as well as air-fluid levels in esophagus, which had a glass-hour shape (Figure 2). Thoracic surgeon was consulted, and the patient underwent additional diagnostic methods. Computerized tomography (CT) of the thorax with per os contrast showed marked esophageal dilatation (10 cm long), along with intraluminal content interpreted as food stasis. Lung parenchyma and mediastinal lymph nodes were normal (Figure 3). Laboratory findings revealed nothing more

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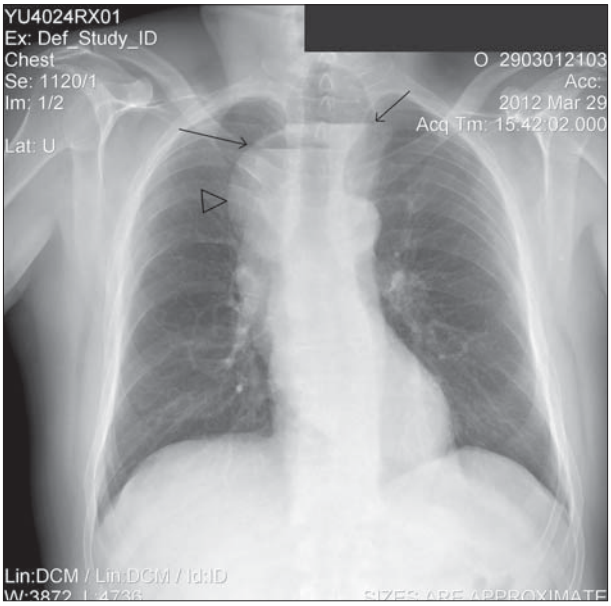

Figure 1. Chest radiography with widened upper mediastinum (arrowhead), two air-fluid levels (arrows), and absent gastric air bubble

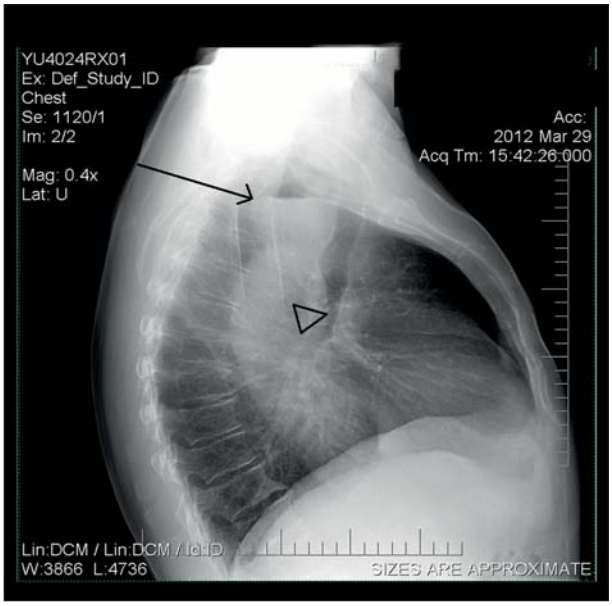

Figure 2. The lateral chest radiography showed dilated esophagus with air-fluid level (arrow), and compression on trachea (arrowhead)

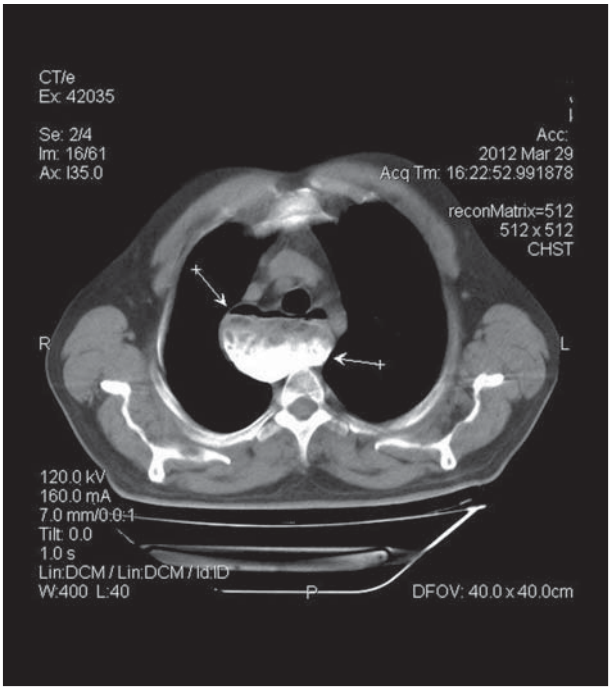

Figure 3. Axial CT shows dilated esophagus (arrow). Per os taken contrast and food remains make up hydro-aeric level (arrow)

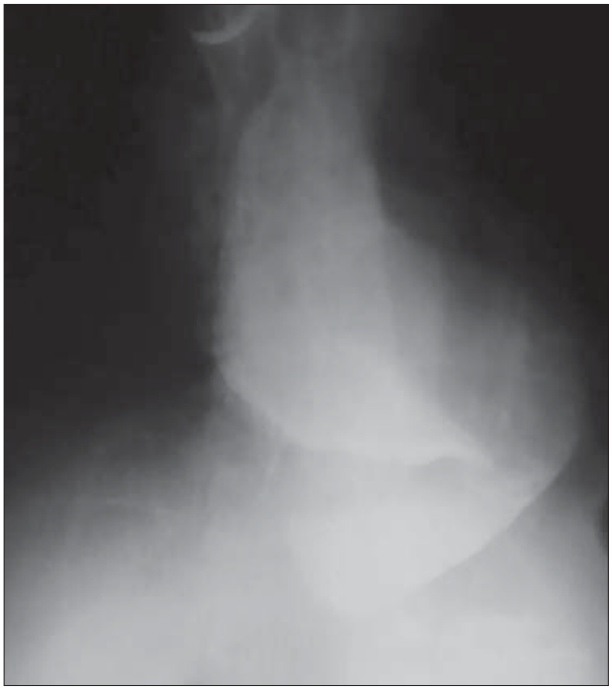

Figure 4. X-ray of esophageal passage preoperatively

than slightly elevated alanine transaminase and gamma-GT. Electrocardiography showed left anterior hemiblock, sinus rhythm with 70 beats/min, with no signs of myocardial ischemia. Echocardiography showed only mild diastolic dysfunction of the left ventricle, ejection fraction was estimated at 65%. X-ray of the esophageal passage showed normal swallowing act, and contrast agent propagated across the dilated, tortuous esophagus, delineating food remnants. No esophageal contractions were noted, and the contrast agent propagated further to stomach, whose tonus was normal. Esophagus had a "sock-like" appearance, characteristic of achalasia (Figure 4).

Esophagogastroduodenoscopy was performed – the endoscopist described dilated esophagus containing food particles, and the signs of chronic esophagitis, as well as the lack of esophageal peristalsis. Given the spasm of cardia, transition to stomach was forced, upon which diffuse edema and erythema of gastric mucosa were seen. Esophageal manometry was also performed (Figure 5), complicated by the fact that manometric catheter could not be placed through LES. All contractions noted in the esophageal body were aperistaltic, simultaneous and of decreased amplitude, which corresponds to typical findings in achalasia. The patient underwent surgery – Heller

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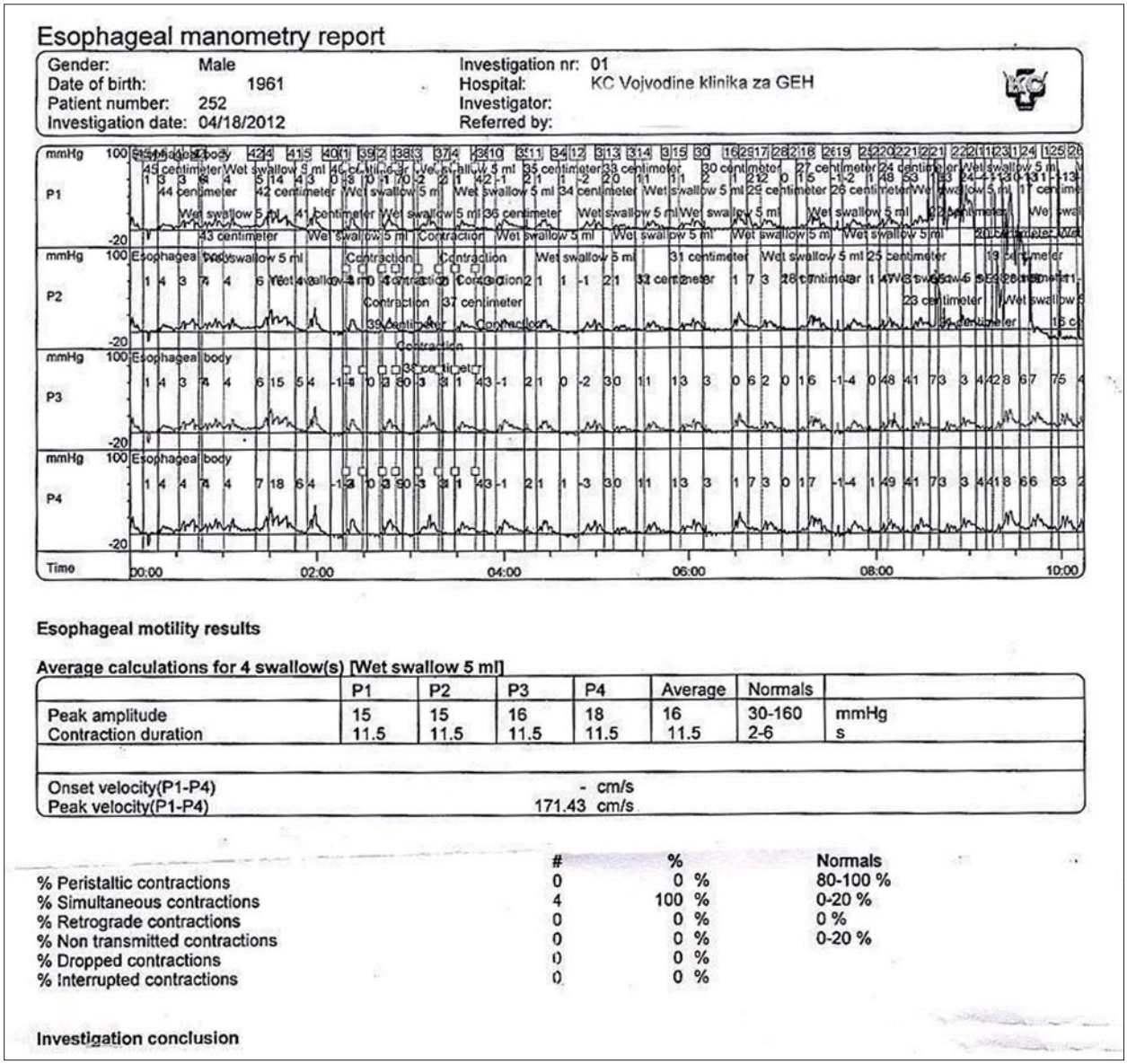

Esophageal manometry report

Gender: Male
Date of birth: 1961
Patient number: 252
Investigation date: 04/18/2012
Investigation nr: 01
Hospital: KC Vojvodine klinika za GEH
Investigator:
Referred by:

Esophageal motility results

Average calculations for 4 swallow(s) [Wet swallow 5 ml]

	P1	P2	P3	P4	Average	Normals	
Peak amplitude	15	15	16	18	16	30-160	mmHg
Contraction duration	11.5	11.5	11.5	11.5	11.5	2-6	s

Onset velocity(P1-P4)	- cm/s
Peak velocity(P1-P4)	171.43 cm/s

	#	%	Normals
% Peristaltic contractions	0	0 %	80-100 %
% Simultaneous contractions	4	100 %	0-20 %
% Retrograde contractions	0	0 %	0 %
% Non transmitted contractions	0	0 %	0-20 %
% Dropped contractions	0	0 %	
% Interrupted contractions	0	0 %	

Investigation conclusion

Figure 5. Esophagomanometric findings

cardioesophagomyotomy with Toupet fundoplication was performed from the beginning of dilatation on distal portion of esophagus up to 2 cm across cardia, the total length being 8 cm. Postoperatively there were no complications and the patient was discharged from the hospital.

A follow-up X-ray examination with barium showed unobstructed passage of the contrast agent to the stomach, without reflux in the Trendelenburg position. The patient is currently symptom free, and he has gained 5 kg of body mass. He refused follow-up esophagogastroduodenoscopy and esophageal manometry.

DISCUSSION

Achalasia is a rare esophageal disorder with different prevalence (in the United States of America it is 10.8 per 100,000 inhabitants) [6]. Predominant clinical features are gastrointestinal, such as dysphagia, food regurgitation, and weight loss. However, these patients may also have respiratory symptoms: dyspnea, chest pain, dysphonia, and recurring aspiration pneumonias are also a possibility. According to the study by Sinan et al. [7] which included 110 adult patients with achalasia, most frequent respiratory symptoms were cough (37%), aspiration (31%), and dysphonia (21%). In children with achalasia, cough is a common symptom [8].

Diagnosis of achalasia is based on the detailed anamnesis, clinical features, and additional diagnostic procedures. These are the following: chest radiogram typically showing dilated esophagus and absence of gastric air bubble, which is insufficient for the final diagnosis, leading to further diagnostic methods such as gastroduodenal radiography with contrast agent, esophageal manometry and endoscopy. Also, a valuable diagnostic tool is a thoracic CT scan with per os contrast. Typical finding is a constricted LES, along with esophageal dilatation above the sphincter. Esophagomanometric measures indicate changes in esophageal wall pressures. During this test, a thin probe with numerous sensors is placed through the patient's

nose or mouth in order to measure pressure changes in the esophageal wall. This test is used to confirm the diagnosis of achalasia, the typical finding being elevated LES pressure and inability to relax this sphincter during the act of swallowing, as well as the lack of peristalsis in distal esophageal segments [9]. The main advantage of endoscopy is the ability to confirm potential alternative diagnoses which may lead to the clinical findings similar to those of achalasia. These would primarily include esophageal carcinoma, esophageal ulcers, and bacterial or fungal infections. Also, biopsies taken during endoscopy may lead to pathohistological confirmation of the listed diagnoses.

Current guidelines of the American Gastroenterologists Association state that treatment modality in patients with achalasia should be determined with regard to sex, age, advantages of the particular therapeutic option but also the experience of the doctor in charge [9]. According to these guidelines, the first step in the treatment of achalasia should be either balloon dilatation or surgery. Balloon dilatation implicates balloon inflation at the level of esophageal and gastric junction with resulting rupture of muscle fibers, with preservation of esophageal mucosa. Success rate for this procedure is 80%, according to the study by Doder et al. [10] and esophageal perforation is noted in 5% of patients, which requires urgent surgical intervention [11]. Surgical method of choice is Heller's myotomy with partial fundoplication. Main complication of this procedure is gastroesophageal reflux [12]. Another treatment modality is endoscopic injection of botulin toxins (Botox) in the muscle fibers of LES [13].

Medicamentous therapy implies the use of calcium channel blockers and sublingual nitrates (isosorbide dinitrate or nitroglycerin), which have a temporary effect on LES pressure. These drugs are efficient in up to 10% of patients with mild or intermediate form of achalasia, but their therapeutic effect is short-term [14, 15]. Also, phosphodiesterase-5-inhibitor is used for similar purposes [16]. Dietetic regimen includes frequent small meals, thermally processed, followed by appropriate fluid intake. It is important to advise patients not to lie down after meals. Patients should avoid food which potentiates reflux (alcohol, caffeine, chocolate, ketchup). Peroral endoscopic myotomy is a more recent treatment modality [17]. Our case report pertains to the patient with achalasia which was not initially recognized by gastroenterologist. Literature cites several cases where achalasia was diagnosed by pulmonologists, due to dominant respiratory symptoms, such as chronic cough. Case report by Kwon et al. [18] describes a patient whose leading symptom was chronic cough, with occasional dysphagia and gastric reflux during the cough. Chest radiogram showed pneumonic infiltration in the right lung. Esophagogastroduodenoscopy was performed, which visualized esophageal mucosal ulcers and chronic gastritis. The patient was treated with levofloxacin and proton pump inhibitors. This therapy resulted in radiological resolution of previously described pneumonic infiltration, but chronic cough persisted, which led to thoracic CT, which showed dilatation of esophagus, that was filled with liquid content, which was interpreted as achalasia, along with new bilateral pneumonic infiltrations (aspiration pneumonia).

Esophagogastroduodenoscopy is not the diagnostic method of choice in these patients, since achalasia cannot be distinguished from gastroesophageal reflux disease [18]. Thoracic CT and esophagography are diagnostic methods which allow better visualization of morphologic changes on esophagus, which was the case in our patient. Since the patient was referred to a pulmonologist after a chest radiogram, thoracic CT was initially performed in order to guide further diagnostics.

Since the American Gastroenterologists Association states that myotomy or pneumatic dilatation may both be used, depending both on the individual characteristics of each patient and the doctors' experience, our patient underwent Heller's myotomy. Research performed on 73 patients with this surgical intervention showed excellent results in 89% of patients, and the six-month and six-year follow-up showed good results in 57% of patients [19]. Our patient is well two years after the surgical intervention, has no dysphagia and has regained body weight. He refused suggested follow-up procedures (esophagomanometry, esophagoscopy). However, regular follow-up is absolutely recommended in these patients, since up to 10–15% of patients with achalasia may develop the so called "end-stage" achalasia (megaesophagus), especially in patients whose initial esophageal dilatation was above 6 cm, which was the case in our patient [20].

Even though achalasia requires multidisciplinary approach, its leading manifestations are gastrointestinal, which, combined with the radiological findings, should first prompt gastroenterological evaluation.

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„Залутала“ ахалазија – од гастроентеролога до пулмолога и назад

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КРАТАК САДРЖАЈ

Увод Ахалазија је ретко обољење једњака које се, с обзиром на заступљеност гастроинтестиналних и респираторних тегоба, може наћи у жижи интересовања гастроентеролога или пулмолога.

Приказ болесника Педесетогодишњи мушкарац упућен је на преглед гастроентерологу због дуготрајне мучнине, повраћања, повременог штуцања уз болове у грудима и губитак на телесној маси, промуклост и надражајни кашаљ. После радиолошког снимања грудног коша болесник је упућен пулмологу јер се посумњало да су у питању анеуризма грудне аорте, дивертикулоза једњака или инфилтрација у плућима. Увидом у радиограм грудног коша пулмолог је уочио паратрахеално проширење медијастинума с течним нивоима и посумњао на ахалазију једњака. Урађена је компјутеризована томографија грудног коша с контрастним средством *per os*, где је запажен изразито дилатиран једњак са садржајем хране у његовом лумену. Болесник је упућен грудном хирургу, који је после одговарајуће преоперационе припреме и додатних дијагностичких процедура (пасажа једњака с применом контрастног средства, езофагоманометрија, езофагогастродуоденоскопија) оперисао болесника применом Хелерове (*Heller*) миотомије. Постоперациони ток је протекао нормално и на контролном прегледу болесник је био без тегоба.

Закључак Ахалазија је обољење једњака које је удружено с постојањем типичних, пре свега, гастроинтестиналних тегоба, мада су честе и респираторне тегобе. Добра анамнеза уз правилно тумачење радиолошког налаза су предуслови за постављање дијагнозе овог обољења.

Кључне речи: ахалазија; бол у грудима; промуклост

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