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Case Report

End-stage, Chicago/FBF Type I Achalasia in a Patient with Long-standing Untreated Dysphagia: Case Report and Pictorial Essay

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Abstract

We describe here the clinical history of a 52-year old woman presenting with a severe long-standing esophageal dysphagia of more than 10 years, for both liquids and solids with regurgitation, chest discomfort with pain and a 12 kg weight loss in the previous five years due to severe eating impairment. Apart from these symptoms, the patient's clinical history was unremarkable. The patient was seen by our Gastroenterology specialists and subsequently referred to our Radiology Unit, where Barium Swallow was performed and the suspected diagnosis of Achalasia was confirmed.

Introduction

Achalasia is a degenerative disease in which failure of distal esophageal neurons causes incomplete Lower Esophageal Sphincter (LES) relaxation, increased LES tone and progressive impairment of esophageal peristalsis [1]. In the natural history of the disease, especially, as in the case of our patient, it is allowed to go on untreated, what happens is a 'decompensation', as first described by Olsen [2] in 1969: even though the motor disfunction remains unchanged, clinically and radiologically the disease evolves naturally towards peristalsis impairment and progressive esophageal dilatation. Definitions of End-stage Achalasia are many and most of them date back in time: Peters described end-stage disease in patients with failed previous treatments occurred and persistent or recurrent dysphagia, associated with a dilated and tortuous esophagus, retained food at endoscopy, a "tree-barked" aspect of the mucosa, and/or a reflux stricture [3]; Banbury adopted a clinical approach to the definition, considering the disease to be end-stage when disabling dysphagia with regurgitation and weight loss due to important dietary restrictions occurred despite aggressive treatment, associated with esophageal dilatation and tortuosity of the esophagus [3]. Radiologically, an end-stage achalasia is considered to be present when the esophagus is dilated, the diameter is 6 cm or more and there is a sigmoid-shaped lumen [1,4]. The new integrated multidisciplinary approach suggested by our group is based on the Chicago Manometric Classificatio, Seoul Consensus on Achalasia and the consequently derived barium swallow FBF grading system [1,4], developed at the Fatebenefratelli Hospital in Benevento, Italy. This approach divides achalasia into three subtypes or syndromes, the atonic type I, panpressurizing type II and spastic type III; end-stage, sigmoid achalasia may happen in all three types but, in the case of untreated disease, it is much more likely to be the final stage of type I achalasia, with its peculiar treatment pathways and prognosis. However, all the different approaches to the disease converge, though, on one point: the megaesophagus. The importance of this stage of this disease is summarized by Orringer [5], who realized that the disease at this stage is not relieved with incision of the LES, as in the less severe stages, and always opted for more aggressive surgery, esophagectomy, when in presence of what he defined a 'sink-trap' esophagus. In this case report, we describe the peculiar case of a patient with a long-standing, untreated dysphagia that fits all the previous definitions.

Discussion

Our patient is a 52-year-old woman presenting with a severe, long-standing esophageal dysphagia of more than 10 years, for both liquids and solids with regurgitation, chest discomfort with pain and a 12 kg weight loss in the previous five years due to severe eating impairment. The patient, who had not felt the need for medical help until the day she came into our GI practice, used to help herself to many glasses of water during meals to help reduce food impaction and stimulate LES relaxation, often to no avail, with chest discomfort and pain enduring for hours after the meal, until LES opened and food passed into the stomach. The clinical hypothesis of achalasia was supported by a score of 29 out of 31 at the Quality of Life for Achalasia questionnaire [6], which highlighted the huge impact the suspected achalasia was having on her life and its quality. The patient was then scheduled for a Barium Swallow in our Radiology Department, which was carried out according the dynamic protocol

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designed in our institution [6,7], specifically modified for the patient adding still, timed evaluations at 10 and 15 minutes. All the hallmarks of classic, end-stage achalasia were present [4,7-10] and the diagnosis was confirmed within seconds: bird beak sign, esophageal dilatation (maximum diameter 63 mms, the cut-off for end-stage disease being 60 mms) (Figure 1), with absent lower esophageal sphincter relaxation that is not coordinated with esophageal contraction, stasis of barium in the esophagus, which has become atonic and sigmoidal, as typical in end-stage disease (Figure 2). The presence of uncoordinated, non-propulsive, tertiary contractions was noted, coupled with the failure of normal peristalsis to clear the esophagus of barium, with no primary waves identified (Figure 3). The hydrostatic pressure can generally overcome the lower esophageal sphincter pressure, allowing passage of esophageal content, usually with the help of a hot or carbonated drink; in our patient, we were able during the exam to visualize just partial sphincter relaxation and barium emptying, even at the timed evaluations at 10 and 15 minutes (Figure 4). The FBF score for this patient was 12, confirming the presence of a severe type I, atonic achalasia. The radiological findings of atonic, end-stage achalasia were confirmed by manometry and the patient was then referred to a specialist center to have an esophagectomy carried out [11-13].

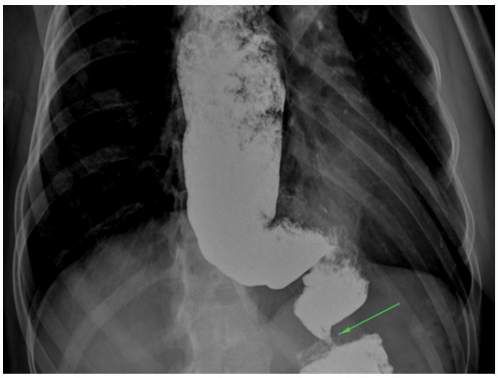

Figure 1: Frontal view. Bird beak sign shows the absent/incomplete LES relaxation, with a reduced or filiform caliber; esophageal dilatation (caliber 63 mms) is noted above the LES and along all the esophageal body.

Figure 2: Frontal view. Uncoordinated esophageal contraction, with stasis of barium in the esophagus, which has become atonic and sigmoidal, as typical in end-stage disease.

Figure 3: Right Anterior Oblique view. Uncoordinated, non-propulsive, tertiary contractions, with no primary waves identified.

Figure 4: Frontal view. Partial sphincter relaxation and barium emptying are noted, at the timed evaluation at 15 minutes.

Conclusion

This case brilliantly shows all the typical clinical and radiological hallmarks of end-stage achalasia, its peculiarity being that it represents the actual final stage of the untreated disease. As stated before, at this point the esophagus has become so atonic that there is no more role for medical therapy and conservative surgery, and only an aggressive surgical approach, an esophagectomy with reconstruction, can restore an acceptable quality of life.

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