Case report

Dysphagia in an elderly patient suffering from long-lasting rheumatoid arthritis.

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Abstract

idiopathic esophageal achalasia (IEA) is a rare motor disorder whose initial manifestation is dysphagia, sometimes versus liquids. IEA can be an uncommon manifestation of some rheumatic inflammatory diseases such as Sjogren syndrome, systemic lupus erythematosus, sclerodermia, rheumatoid arthritis. IEA is considered a precancerous condition and the possibility that it represents a risk factor for development of esophageal carcinoma (EC) has been more and more descripted. These same rheumatic diseases - when long-standing and/or aggressive - can give an esophageal amyloidosis (EA) with AA amyloid deposits . EA is very rarely descripted as unique manifestation of systemic amyloidosis during these diseases. IEA has a typical manometric pattern whereas in EA the anarchic arrangement of amyloid can produce unforeseen and uncharacteristc motility patterns different from the idiopathic one. Both IEA and EA are very rare conditions. For IEA, it has been extimated an annual incidence of 2/100,000 and a prevalence rate of 10/100,000 with a peak incidence between 30 and 60 years of age. The prevalence and incidence of EA are uncertain. The surgical experiences in esophageal pseudoachalasia are aneddhotic. We describe the clinical case of an elderly woman suffering from long-lasting rheumatoid arthritis in which an EA overlapped an IEA, causing the sharp worsening of dysphagia and poor prognosis despite surgical approaches. According to our knowledge, this overlap has been never descripted. ***Keywords:*** *esophageal achalasia, dysphagia, rheumatoid arthritis, amyloidotic psuedoachalasia, elderly patient*

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Introduction

Esophageal achalasia (EA) is a rare disorder characterized by loss of peristalsis and insufficient lower esophageal sphincter (LES) relaxation in response to deglutition. Dysphagia is the first clinical manifestation, in the vast majority of cases ; it has a progressive course ; sometimes it begins to liquids, only (*paradoxical dysphagia*). Material retained in the esophagus may be aspired in the respiratory tract, expecially during supine position at night, and led to an ab ingestis pneumonia. Esophageal achalasia can represent an uncommon manifestation of some rheumatic diseases such as Sjogren syndrome, sclerodermia, sistemic lupus eythematosus, rheumatoid arthritis…..(1). Besides, an esophageal amyloidosis secondary to these inflammatory diseases has been rarely descripted. In this specific condition, the anarchic arrangement of the amyloid (AA type) in the esophagus can produce unforeseen and uncharacteristic motility patterns almost different from the idiopathic one, justifying the term of *pseudoachalasia* (2,3). The possibility that an amyloidotic achalasia may overlaps an initial idiopathic achalasia has never been descripted.

Case presentation

A.M.S. was a 76-years-old woman suffering from rheumatoid arthritis (RA) for 20 years, successfully treated with methotrexate (10 and then 15 mg /week) and antimalarials synthesis (plaquenil 200 mg/day). Low-dosage corticosteroids were added for short periods in presence of inflammatory flares, expecially at wrists and toes. Over the past 6 years, she began to complain of paradoxical dysphagia but it had been initially underestimated due to a long-standing gastroesophageal reflux. When dysphagia worse, after using prokinetics with little benefits, she had consulted a gastroenterologist who after endoscopic examination and esophageal manometry diagnosed EA and recommended treatment with botulinum toxin. During endoscopic examination, had been carried out random bioptic samples which did not provide any meaningful diagnostic element.

The benefit of endoscopic infiltration with botulinum toxin was excellent and she periodically repeated it for several times until to the end of 2013. In 2014, a severe three-vessel coronary artery disease was diagnosed and she was in III° NYHA functional class even after a heart surgery operation. In september of 2014, because of a sharp worsening of dysphagia with rapid weight loss, she was hospitalized. An endoscopic control showed some areas of increased thickness of the wall suspicious for cancer development but the bioptic examination revealed the presence of amyloid deposits (figura 1a). The lost of affinity for Congo red after treatment with potassiun pergamanato (figure 1b) allowed the diagnosis of AA amyloid. Laboratory tests showed proteinuria (1.5 gr/24 hours) , modest elevation of creatinine (1.60 mg/dl vs < 1.10) – previously always normal and elevated levels of serum amyloid A (25 mg/L vs < 10). Normal were abdominal and pelvic US and all other laboratory tests. Therapy with infliximab (that has documented efficacy in the treatment of secondary amyloidosis in RA) was excluded for the heart disease. It was carried out an esophageal pneumatic dilation (with no results) and then a per-oral endoscopic myotomy but the patient died after 6 months for an aspiration pneumonia.

Discussion

Femoral neck fractures are one of the important achalasia is an uncommon esophageal motility disoder. It has been extimated an annual incidence of 2/100,000 and a prevalence rate of 10/100,000 with a peak incidence between 30 and 60 years of age (4). Morphologic studies have showed the loss of myenteric gangion cells in the esophageal myenteric plexus. In severe cases, the myenteric plexus had been replaced by collagen (5). Many studies suggests a significantly increased cancer risk and some authors think that achalasia represents a precancerous condition (6). Tumors in the gastric cardia or those infiltrating the myenteric plexus such as pancreatic, breast, lung or hepatocellular cancers can be considered in the differential (7). The possibility that RA (such as other inflammatory chronic conditions) can develop an secondary amyloidosis with formation of amyloid type AA is well-known. This possibility is strongly linked to duration and severity of RA. Amyloidosis secondary to RA is different from amyloidosis (so-called AL amyloid) secondary to cancer or lymphoproliferative malignancies : the lost of affinity to Congo-red colorant after treatment of the bioptic specimen with potassium permanganate is characteristic of amyloid AA and not for the AL-one. Esophageal amyloidosis secondary to RA has been rarely descripted (3,8) . In some cases the radiologic, clinical and endoscopic pictures are similar to idiopathic achalasia while manometric pattern can be different . In our patient, the first upper endoscopy with random biopsies excluded the presence of amyloid deposits. The manometric pattern had been typical for achalasia. The second endoscopy had showed a thickening area of the esophageal wall and its biopsy had revealed the presence of AA amyloid. A second manometry had been not possible and so the possibility that amyloid deposits was able to modify the initial manometric pattern is only speculative. Certainly the association between idiopathic achalasia and amyloidotic pseudo-achalasia has changed the course and the prognosis of our patient. The presence of a severe heart disease has strongly influenced treatment decisions.

Conclusions

idiopathic achalasia and esophageal amyloidosis are two extremely uncommom diseases. Therefore their association should be considered frankly outstanding. According to our knowledge, the possibility that an esophageal amyloidosis can overlap an initial idiopathic achalasia has never been descripted before.

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