REVIEW ARTICLE

Pseudoachalasia: A review

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Abstract

Pseudoachalasia presents typically like achalasia. It account for only 2.4-4% of patients presenting with achalasia-like symptoms. Clinical, radiologic and endoscopic findings resemble those of achalasia but treatment and prognosis are different in these conditions. The aim of this review is to give an overview of the condition and highlight challenges in diagnosis and distinguishing features between the two conditions. A review of the publications obtained from Medline search, medical libraries, and Google on 'pseudoachalasia' and 'secondary achalasia' was done. A total of 50 articles were retrieved and used for this review. There has been tremendous efforts towards establishing the diagnosis of pseudoachalasia both clinically and with the use of modern investigative modalities but to date its still difficult to distinguish it from achalasia. Endoscopy, endoscopic ultrasonography and computerized tomography scan have shown promising results.

Key words: Dysphagia, pseudoachalasia, secondary achalasia

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Introduction

Pseudoachalasia is characterized by achalasia-like symptoms caused by secondary etiologies in most instances an occult tumor. Clinical, radiologic, and endoscopic findings resemble those of achalasia, but treatment and prognosis are different in these conditions.[1] This condition may be difficult to diagnose in the early phase because of low diagnostic yield of either barium or endoscopic findings and false-negative rate of up to 25% of endoscopic biopsies in the diagnosis of the primary cancer.[2]

It is a rare clinical entity accounting for only 2.4–4% of patients presenting with achalasia-like symptoms.

We aim to update clinicians on this rare disease entity and highlight the challenges in making diagnosis and treatment.

Methods

A literature review of pseudoachalasia was done from 1970 to date using manual library search, journal publications on the subject, and Medline using search

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terms "pseudoachalasia" and "secondary achalasia". Full texts of the materials, including those of relevant references, were collected and studied. Information relating to historical perspective, etiology, pathogenesis, clinical features, diagnosis, and treatment were extracted from these materials.

Historical perspective

Horvath first suggested this entity in 1919.[3] Ogilvie later described the syndrome in 1947.[4] He described it as a form of achalasia as a result of the involvement of the cardia region from gastric carcinoma. Park, in 1952, and Asherson, in 1953, reported on patients with apparent "cardiospasm", in whom gastric cancer was eventually diagnosed.[5,6]

Incidence and prevalence

Owing to the lack of a large series, there are no reliable epidemiological data on the incidence and prevalence of the disease. Campo *et al.* in their review reported 4.7% of patients who fulfill the manometric criteria of achalasia were eventually diagnosed with a malignant disease, directly or indirectly involving the cardia, or following antireflux surgery.[5]

Etiology

Primary malignancies of the esophagus and esophagogastric junction account for 50% of cases of pseudoachalasia.[7] This is followed by secondary malignancies (18%) such as metastases, which primarily originated from the lungs and breast. Malignancies from different parts of the body have also been reported.[8-12]

Benign causes of pseudoachalasia include benign mesenchymal tumors, secondary amyloidosis, and peripheral neuropathy. Others include pancreatic pseudocyst, sarcoidosis, neurofibromatosis,[13,14] esophageal leiomyomatosis,[15] esophageal mesenchymal tumors,[15] histiocytosis-x,[16] aortic aneurysm, juvenile Sjogren syndrome, chronic idiopathic intestinal pseudo-obstruction, familial glucocorticoid deficiency syndrome, and a few surgeries (vagotomy, bariatric, and Nissen fundoplication).[1] They account for 12% of the causes of pseudoachalasia. Three explanations have been proposed for antireflux surgery causing pseudoachalasia. These are misdiagnosed idiopathic achalasia with evidence of dysphagia just after surgery, achalasia occasionally developing for the underlying gastroesophageal reflux, and development of scar tissue and/or an overly tight fundic wrap.[17] Pseudoachalasia has also been reported in a patient that had laparoscopic adjustable gastric banding (LAGB).[9] Encapsulating peritoneal sclerosis (EPS) as a complication of peritoneal dialysis has also been reported as a cause of pseudoachalasia.[18]

Rare causes include paraneoplastic syndromes from small cell carcinoma of the lung, bronchial carcinoid, gastric carcinoma, and pleural mesothelioma.

Pathogenesis

Two patterns of tumor involvement have been described.[10] The most common type consists of malignant stricture of the cardia that acts as a physical barrier to the passage of food. A less frequent type is strictly related to the malignant submucosal infiltration with secondary impairment of inhibitory neurons of the esophageal myenteric plexus by tumor cells, which let the manometric pattern of achalasia be stable even after any treatment. Indeed, many malignancies as common causes of pseudoachalasia directly involve the esophageal myenteric plexus by neoplastic cells infiltrating the mucosa at the cardia as the main pathogenetic mechanism.[19-21] However, neuronal degeneration distant from the primary tumor site with a reduction in ganglion cells in the dorsal nucleus of the vagal nerve or in the vagal nerve itself has been also proposed.[22-24] This interaction of tumor factors with the esophageal neuronal plexus without a direct infiltration of the esophagogastric junction, even infrequently, and serological antineuronal nuclear antibody can be detected in these patients, suggesting a paraneoplastic syndrome. Another form of pseudoachalasia occurs following antireflux surgery.[17] Three explanations have been proposed: Misdiagnosed idiopathic achalasia with evidence of dysphagia just after surgery, achalasia occasionally developing for the underlying gastroesophageal reflux, and development of scar tissue and/or an overly tight fundic wrap.[25-27]

The mechanism of esophageal dilation and dysmotility postgastric banding is unknown. Lower esophageal sphincter (LES) pressure and LES residual pressure are increased after LAGB. A proposed mechanism is the high outflow resistance caused by the gastric band at the LES that creates a high-pressure area leading to a progressive weakening of the esophageal musculature. The inflammation around the laparoscopic band, with fibrosis or neuromuscular damage, may account for the variability of manometric studies.[9]

The mechanism of pseudoachalasia in EPS is unclear, but authors of the single case report of EPS causing pseudoachalasia postulated that thickened and sclerotic peritoneum on the abdominal surface of the diaphragm and the lower esophagus caused a mechanical constriction of the lower esophageal sphincter presenting as pseudoachalasia.[18]

Clinical Features

The classic symptom presentation of short-term interval weight loss, progressive dysphagia, and chest pain are secondary to impaired esophageal emptying at the distal esophagus. The demographic study has demonstrated gender predisposition with a median age of 61 years.[28] Patients with pseudoachalasia are usually over 50 years of age; they present with a short history of progressive dysphagia usually <1 year and significant weight loss

usually >7 kg and retrosternal pain.[29] They may present with other features of primary malignancy.

Diagnosis

Pseudoachalasia often presents serious diagnostic difficulty[30] and the consequent delay in instituting appropriate treatment may result in an underlying carcinoma becoming inoperable. Therefore, this demands committed clinical, radiographic, manometric, and endoscopic assessment to achieve distinction from primary achalasia. Careful preoperative assessment is imperative prior to surgical intervention

Barium swallow findings often mimic the classic primary achalasia finding of the smooth tapering distal esophagus ("Bird's beak" appearance) and dilated esophagus with barium column.[31] However, barium swallow features that may be suggestive of pseudoachalasia are a short segment of dilated esophagus, asymmetry of esophageal narrowing, asymmetry of esophageal wall thickness, rigidity of lower esophagus, deformity of the stomach, and mucosal ulceration.[32] Woodfield *et al*.[33] reported that secondary achalasia would not be suspected in most cases solely on radiologic criteria. However, the narrowed distal esophageal segment was longer than 3.5 cm in 80% of patients with secondary achalasia in contrast to narrowed segment of <3.5 cm in primary achalasia. In their series, the degree of esophageal dilatation above the narrowed segment was also a statistically significant criterion for differentiating primary from secondary achalasia. The diameter of the esophagus was 4 cm or less in 80% of patients with secondary achalasia, whereas the diameter of the esophagus was >4 cm in 90% of patients with primary achalasia presumably related to the more gradual course of the disease that allowed the esophagus to progressively dilate over a period of years.

Amyl nitrite test

This pharmacologic inhalant and smooth muscle relaxant induce a measurable increase of 2 mm or more in sphincter diameter for patients with primary achalasia. Patients with pseudoachalasia exhibit no response.[34]

Esophageal manometry

The finding is similar to those of primary achalasia that are aperistalsis of the body of the esophagus and poor lower esophageal sphincter relaxation.[31]

Endoscopy

This is probably the only investigative modality that can suggest pseudoachalasia. The findings are mucosal ulceration or nodularity, reduced compliance of the gastroesophageal junction, or an inability to pass the endoscope into the stomach.[10] Endoscopic biopsy remains the most definitive tool for the diagnosis of pseudoachalasia occurring secondary to distal esophageal or esophagogastric junction carcinoma. The hallmark pathologic characteristics of this condition are an invasion and disruption of the myenteric plexus or a paraneoplastic autoimmune-mediated depletion of myenteric ganglion cells.[27,35] Kahrilas *et al*.[16] in their series of 32 patients with pseudoachalasia, endoscopy and biopsy revealed the correct diagnosis of malignancy in only 66% of all patients with pseudoachalasia. In a further series, only 10% of all biopsies obtained in patients with pseudoachalasia suggested the presence of malignancy. Consequently, a second endoscopy with multiple biopsies is frequently necessary.[36]

Endoscopic ultrasonography

Reliable means of diagnosis for carcinoma at the cardia and distinguishing it from achalasia. Lymph node enlargement adjacent to the tumor can be detected.[37] It has low accuracy in differentiating mucosal from submucosal lesions at the lower esophagus or gastroesophageal junction.[38]

Computed tomography

May show diffuse thickening of stomach wall in the fundus and adjacent body region suggesting a possibility of a diffuse neoplastic lesion.[39] It may also depict the malignant lesion, lymph node involvement, as well as regional and metastatic spread, especially hepatic and pulmonary when it is more than 15 mm in diameter.[40,41] CT scan may also show intrathoracic malignancies infiltrating the esophagus.

Differentiating features between achalasia and pseudoachalasia:

1. History – achalasia has a long history while pseudoachalasia has a short history usually <6 months
2. Weight loss – patients with achalasia has little weight loss while those with pseudoachalasia have marked weight loss
3. Age – patients with achalasia are usually <50 years while those with pseudoachalasia are more than 50 years of age
4. Fecal occult blood test - negative in achalasia but may be positive in pseudoachalasia
5. Amyl nitrite test - induces a measurable increase of 2 mm or more in sphincter diameter for patients with primary achalasia. Patients with pseudoachalasia exhibit no response
6. Barium swallow – the length of narrowed distal esophageal segment is usually <3.5 cm in achalasia while in pseudoachalasia it is >3.5 cm. The degree of esophaeal dilatation above the narrowed segment is usually >4 cm in achalasia because of longstanding obstruction while in pseudoachalasia it is <4 cm
7. Endoscopy – endoscope can easily pass into the stomach in achalasia while in pseudoachalasia the endoscope cannot pass into the stomach
8. CT scan – there is no lesion in achalasia but there may be lesion at the gastroesophageal junction

9. Dysphagia – it is relieved by pneumatic dilation in achalasia but not relieved in the case of pseudoachalasia.

Treatment

The mechanism of pseudoachalasia is a mechanical obstruction at the gastroesophageal junction; treatment, therefore, is aimed at removing the obstruction by surgery, chemotherapy, or radiotherapy. This has been shown to allow the normal return of peristalsis.[42]

Early malignancies of gastroesophageal origin

Esophagogastrectomy via either Ivor Lewis[43] or thoracoabdominal incision.[39] Others advocate subtotal McKeown esophajectomy, especially for a middle thoracic pseudoachalasia carcinoma.[44]

Advanced malignancy

Palliative options have been advocated in the form of the use of metallic stent, especially in patients that are not suitable for surgery.[42,45-47] Despite the initial therapeutic success, the overall results were found to be disappointing, frequent complications, such as aorta-enteric fistula, esophageal perforation, stent migration, and severe reflux esophagitis being reported.[48-50]

Conclusion

Differentiating pseudoachalasia from achalasia is very important because their treatments differ. There have been tremendous efforts toward establishing the diagnosis of pseudoachalasia both clinically and with the use of modern investigative modalities but to date it is still difficult to distinguish it from achalasia. Clinical features together with the investigations mentioned should form a reliable tool for the diagnosis of pseudoachalasia.

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Conflicts of interest

There are no conflicts of interest.

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