

Pseudoachalasia: A Case Report and Review of Literature

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Abstract

Achalasia cardia is due to loss of myenteric ganglion cells in the gastroesophageal junction and the etiology is idiopathic. But a similar clinical picture can be produced by other diseases, a condition termed as secondary or pseudoachalasia. A very high index of suspicion is required for the diagnosis of this condition because this is most commonly produced by a malignancy involving the gastroesophageal junction which is likely to be missed as in our case. We have presented an analysis of the literature available in this context, a knowledge of which will surely help to suspect this condition in atypical cases of achalasia.

CASE REPORT

A 23-year old female presented with chief complaint of dysphagia. She was in good health until one year before when she first noted occasional difficulty eating solid food. She used to have a sensation that food was lodged in her chest which would persist for several hours. These episodes became more frequent over time and she began having difficulty ingesting both liquids and solids. For these complaints, she underwent an upper gastrointestinal endoscopy which was normal. She also gives history of regurgitation and at times had to induce emesis for symptom relief. Apart from the above complaints, she did not have any other complaints except loss of weight.

She is a mother of three children and last child birth was 2 years before. Normal menstruation with no other significant major illness in the past. There is no similar illness in the family. On admission, her vitals were stable. Systemic examination was unremarkable. Electrocardiogram and lab values were normal. Barium swallow demonstrated tapering of distal esophagus typical of achalasia cardia (Fig 1). Upper gastrointestinal endoscopy was normal. She underwent cardiomyotomy by thorocotomy. Intraoperatively intense fibrosis was found around the hiatus with the lower esophageal sphincter being tough and fibrotic. Postoperatively, patient continued to have dysphagia for both solids and liquids which responded to nifedipine and domperidone. She was discharged on those medications.

Figure 1

Figure 1: Typical bird-beak appearance in barium swallow study.



Three months later she came back with complaint of recurrent intermittent dysphagia. After evaluation which included an upper gastrointestinal endoscopy, patient was taken for re-exploration. Laparotomy was done which showed intense fibrosis around hiatus, esophagogastric junction and upper third of stomach. Lower third esophagus and upper stomach was mobilized fully and myotomy was done well beyond the gastroesophageal junction. No intraluminal pathology could be felt from outside. Symptom relief was satisfactory in the postoperative period and she was discharged with an advice to come back for an evaluation of possible stomach pathology.

When the patient came for review, she was symptomatically better and underwent an upper gastrointestinal endoscopy. An ulcer with grayish slough was found at the gastroesophageal junction which was biopsied. The report

took us by surprise showing adenocarcinoma following which a Computerised Tomography(CT) scan of abdomen was ordered. CT showed a diffuse thickening of stomach wall in the fundus and adjacent body region suggesting a possibility of a diffuse neoplastic lesion. No lymphatic or other metastases were found.

Esophagogastrectomy was performed by a left abdominothoracic approach. A 5 cm proximal and distal margin was given from the palpable edge of the thickening of the bowel wall. Two-layered esophagogastric anastomosis was performed with a pyloroplasty. No lymphadenopathy, ascites, peritoneal or liver metastases were found.

Patient had an uneventful postoperative recovery. Histopathology revealed a moderately differentiated adenocarcinoma with a negative resected margin. She was started on 5-Fluorouracil based chemotherapy. Unfortunately, she developed pulmonary metastasis with malignant pleural effusion after 6 months and died.

DISCUSSION

A complaint of dysphagia and chest discomfort may be associated with any of the esophageal motility disorders including diffuse esophageal spasm, nutcracker esophagus and achalasia cardia. While in former two, chest pain aggravated by hot or cold food lasts for minutes and does not worsen with time, achalasia is characterized by progressively increasing chest pain associated with weight loss, aspiration pneumonia and regurgitation.

In true or primary achalasia, there is loss of myenteric ganglion cells with loss of peristalsis in esophageal body and failure of lower esophageal sphincter to relax when swallowing. Barium swallow shows a typical bird's beak appearance. Manometry is diagnostic with the findings being: increased lower esophageal sphincter (LES) pressure, failure of LES to relax in relation to swallowing, loss of peristalsis in esophagus and increased esophageal pressure. These findings can also be seen in pseudoachalasia or secondary achalasia which is clinically, manometrically and radiographically mimics true achalasia.

Pseudoachalasia accounts for 2.4-4% of patients who present with symptoms consistent with achalasia¹. A review of literature from 1968 to 2002 yielded a total of 264 cases with malignancy accounting for around 60% of lesions. An analysis by Gockel et al², showed the following as causes of achalasia: primary malignancy of esophagus or gastroesophageal junction, 53.9%, secondary malignancies

such as metastasis from lung or breast, 14.9%, benign disorders like mesenchymal tumors, secondary amyloidosis and peripheral neuropathy, 12.6%, and as a postoperative complication following antireflux surgery, 11.9%. Other rare causes are neurological disorders like meningomyelocele, brain metastasis, infiltration by lymphoma and paraneoplastic syndromes associated with small cell carcinoma lung, bronchial carcinoids and pleural mesothelioma.

Differentiating pseudoachalasia from true achalasia is often very difficult. A very high index of suspicion is the only tool helpful in making a preoperative diagnosis. The diagnosis is likely to be missed even intraoperatively as in our patient who presented at a very young age of 23. However, there are certain subtle features that help in distinguishing these two and will lead on to appropriate clinical actions.

The patients with pseudoachalasia tend to be older in age (>60 years), have shorter duration of dysphagia (<6 months) and have more substantial weight loss. Barium swallow may reveal a nodular or shouldered segment of distal esophageal narrowing. The length of the narrowed esophageal segment was found to be longer than 3.5 cm in 80% of the patients with pseudoachalasia which is the most important feature in radiological differentiation³. A CT scan may show asymmetric thickening of the esophageal wall or cardia, mediastinal lymphadenopathy or may identify primary malignancy in secondary achalasia.

Esophageal manometry does not help much in distinguishing these conditions except when amyl nitrite test was performed. Inhalation of amyl nitrite decreases the lower esophageal sphincter pressure substantially while not much so in malignant stenosis⁴. While upper gastrointestinal endoscopy is also not very helpful in making a distinction, findings like mucosal ulceration, nodularity and inability of the scope to negotiate the esophagogastric junction may be suggestive⁵. In those circumstances, endoscopic ultrasound to look for submucosal tumor and lymph nodes, if any may be helpful. A deep biopsy can also be done but has been shown to have a low diagnostic yield again.

Our patient probably represents the youngest reported case of pseudoachalasia and illustrates a perfect example for how misleading will a presentation of pseudoachalasia be. Authors would like to suggest that a good knowledge of this condition and a very high degree of clinical suspicion based on the aforementioned facts will help make an early diagnosis. We would like to conclude stating that a clinical

suspicion at least will help in changing the course of action wherein a surgical exploration will be a sound choice compared to an ill-advised pneumatic dilatation in such cases.

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