Pseudoachalasia: a diagnostic challenge

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Abstract

Here, we present a case of a 78-year-old man that underwent gastrointestinal endoscopy because of one-month history of dysphagia to liquids and solid foods with accompanying weight loss. On endoscopy, there was distal esophageal stenosis. Multiple biopsies were obtained. Histologic examination of the samples revealed normal tissue. The stenosis was treated by dilatation and abdomino pelvic computed tomography scanning was performed to search for an underlying malignant lesion that showed a mass adjacent to distal esophagus. We did endosonography-guided fine needle aspiration of the mass. It was a squamous cell carcinoma (SCC). Malignancy is a challenging diagnosis in patients with dysphagia and near normal endoscopy. To our knowledge, there are a few reports of SCC to cause it.

Keywords: Achalasia, Dysphagia.


Introduction

Achalasia is a primary esophageal motility disorder presenting with dysphagia to liquids and solid foods. It is usually diagnosed using barium swallow study and manometry. Barium study shows narrowing of esophagogastric junction (EGJ) while manometry shows decreased or absent distal esophageal peristalsis with increased lower esophageal sphincter (LES) pressure and impaired LES relaxation with deglutition. Esophagus appears to be normal on endoscopic examination (1).

Pseudoachalasia is characterised by achalasia-like symptoms caused by secondary etiologies. Clinical, radiologic and endoscopic findings resemble those of achalasia but treatment and prognosis are different in these conditions. Therefore, discriminating between these two disorders is important (1-5).

Case report

A 78-years-old man was admitted to our hospital due to 10-days history of intractable postprandial vomiting. He complained of progressive dysphagia presented initially to solid foods and then to liquids for a month. During this time, he had 15 kilogram weight loss.

He smoked for decades and noticed occasional productive cough with no relevant diagnostic work-up. He had history of hypothyroidism after being treated by radio–iodine for the primary hyperthyroidism when he was 60-years-old. However, at the time of presentation, he was in euthyroid state.

On clinical examination, he appeared ill and dehydrated. Except for bilateral temporal wasting, the whole clinical examination was normal. Tests of blood and elec-
trolytes showed white blood cell (WBC): 9,700 /µl, Hb: 11.2 g/dl, MCV: 86/fl: Plate- 
et count: 283,000 /µl: ESR; 125 mm/ hour, Na: 137 and K: 3.6 meq/ l. Renal and liver 
function assays were normal as serum pro- 
tein electrophoresis study.

There was no evidence of hepatobiliary or 
splenic abnormality on abdominal sonogra- 
phy.

On upper endoscopy, esophagus appeared 
to be diffusely erythematous with multiple 
white plaques compatible with candidiasis; 
due to tight LES stenosis, attempts to pass 
the endoscope into stomach was futile. A 
fter dilatation with the Savary dilators, all 
parts of stomach were visualized that 
seemed to have normal mucosa. Multiple 
biopsies were obtained from distal esophag- 
us and proximal stomach (both side of 
LES); but histologic examination revealed 
normal mucosa.

Abdominal and chest CT scan showed an 
abnormal soft tissue mass around distal 
esophagus with a solitary 2 cm pulmonary 
nodule (SPN) near right lung hilum. His 
medical records were unavailable to eval- 
uate the growth rate of SPN.

On endosonography (EUS), a 42 mm hy- 
poechoic mass was seen at LES extending 
beneath the mucosa ,involving all layers of 
esophageal wall albeit without any regional 
lymph node abnormality. Fine needle aspi- 
ration (FNA) guided by EUS was done 
(Fig.1). Histological study of samples 
showed a well differentiated squamous cell 
carcinoma (Fig. 2). We did not evaluate the 
SPN as he refused to undergo further diag- 
nostic work-up.

The tumor stage was T3N0M0. There- 
fore, he received chemo radiotherapy with 
platinum-based regimen. Thereafter, dys- 
phagia improved significantly. Unfortu- 
nately, he died six months after treatment 
due to a heart attack.

Discussion

Motility type dysphagia defined by diffi- 
cult passage of both liquids and solid foods 
to stomach is a diagnostic challenge to phy- 
sicians. The differential diagnosis includes 
a wide spectrum of various disorders. At 
one end are benign motility disorders such 
as diffuse esophageal spasm, hypertensive 
LES and achalasia, and at the other ex- 
treme, are ominous malignant cancers mim- 
icking achalasia. Dysphagia to both liquid 
and solid foods that is not due to primary 
neuronal disease and instead is due to sec- 
ondary malignant or non-malignant causes 
is called pseudoachalasia; nonmalignant 
diseases include pancreatic pseudocyst, 
amyloidosis, sarcoidosis, neurofibromato- 
sis, aortic aneurysm, juvenile Sjogren syn- 
drome, chronic idiopathic intestinal pseudo 
obstruction, familial glucocorticoid defi- 
ciency syndrome, and a few surgeries (va- 
gotomy, bariatric, Nissen fundoplication) 
(2, 5-12). Esophagogastric junction adeno- 
carcinoma comprises the most common 
malignant cause of pseudoachalasia (2, 3, 
13); the others include cancers of pancreas, 
esophagus, lung, kidney, hepatobiliary,
lymphoma and mesothelioma (2). Malignant neoplasms comprise 4% of achalasia-like syndromes (3, 13). Dysmotility in achalasia is caused by unrecognized causes that destroy distal esophageal and LES inhibitory neurons; these cause LES muscle contraction with no relaxation in deglutition. Suggested mechanisms for pseudo-achalasia include encircling and compressing LES by tumor, infiltration of myenteric plexus branches or vagus nerves, and para-neoplastic neuropathy without direct infiltration of nerves by malignant cells (2, 5).

Standard treatment of achalasia, including balloon dilatation and surgical myotomy are ineffective and even dangerous for pseudo-achalasia and delays timely diagnosis of malignant neoplasm (1-3, 5). So every attempts should be done to reach correct diagnosis.

Endoscopy is the first diagnostic modality in a patient with new onset dysphagia (14); it detects structural esophageal and gastric abnormalities; especially tumors. In achalasia-like syndrome, it may show mass, nodularity, irregular mucosa or ulcer, pointing to pseudoachalasia rather than achalasia (1, 15). Even in normal appearing mucosa, difficult passage of endoscope through LES suggests pseudoachalasia in contrast to achalasia that endoscope can be passed with subtle pressure through LES (1, 2).

In the case of normal appearing esophagus on endoscopy and new onset dysphagia, the physician must be vigilant to the presence of an underlying malignant lesion, especially when the patient is old, and there is significant weight loss (1, 13, 16). In such circumstances, CT scan and EUS are helpful diagnostic tools. CT scan may show asymmetric wall thickening of distal esophagus and even a well-defined mass (2, 5). EUS would reveal more information as it precisely defines esophageal wall layers and any abnormal tissue; EUS also can be used for tissue acquisition by FNA, when endoscopic biopsies are inconclusive (4, 17, 18).

In the literature, squamous cell carcinoma as a cause of pseudoachalasia has been shown to be derived from esophagus, liver, lung and uterine cervix (1, 2, 5, 13). Our case was probably due to a primary lung squamous cell cancer with metastasis to distal esophagus because he was smoker and had a SPN in right lung.

**Conclusion**

Pseudoachalasia should be in mind when approaching to every patient with suspected achalasia and especially alarm signs should be questioned; on the other hand, it should be kept in mind that simple mucosal biopsy do not exclude pseudoachalasia and in suspected patients further work up such as CT scan and EUS may be warranted.

**References**
