Nutcracker Esophagus With Epiphrenic Diverticulae: Case Report

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Citation

Abstract
Nutcracker esophagus (NE) is a manometric pattern that is commonly seen in patients with noncardiac chest pain and/or dysphagia with normal esophageal peristalsis. We report a rare association of multiple epiphrenic esophageal diverticulae of distal esophagus with NE.

INTRODUCTION
Primary esophageal motility disorders comprise various abnormal manometric patterns which usually present with dysphagia or chest pain. Unlike achalasia other disorders like diffuse esophageal spasm (DES) and NE, have no well defined pathology and could represent a range of motility abnormalities associated with subtle neuropathic changes, gastroesophageal reflux and anxiety states. Manometric patterns of NE poorly correlate with symptoms and response to medical or surgical therapy. Epiphrenic diverticulae of distal esophagus are said to occur with motility disorders like DES, it's rarely reported with NE.

CASE REPORT
A 62-year-old male presented to our department because of intermittent non progressive dysphagia for the past 2 years. The patient reported an increasing frequency and duration of the symptoms which were independent from food intake, medication. There was no history of recurrent heart burn, nasal regurgitation, nocturnal cough or stale food vomiting. Barium swallow was performed which showed multiple diverticulae in lower end of esophagus. (Fig.1)

Figure 1
Figure 1: Endoscopic view of the diverticulae of distal esophagus above the OG Jn.

We performed an upper GI endoscopy revealed multiple outpouchings in lower third of esophagus. (Fig.2)
Esophageal manometry confirmed the presence of a nutcracker esophagus, defined by high-amplitude contractions of the distal esophagus (Fig.3). The patient was treated with nifedipine 10mg before each meal. The symptoms severity decreased but manometric pattern persisted on 6 month follow up.

DISCUSSION

Nutcracker esophagus (NE) was introduced by Benjamin and Castell in 1980. Other terms used are Supersqueezer and hypertensive peristalsis. The manometric feature proposed for a diagnosis of nutcracker esophagus (NE) is a mean distal esophageal peristaltic wave amplitude greater than 2 standard deviations above normal (i.e., greater than 180 mm Hg) in a symptomatic patient (measured as the average amplitude of 10 swallows at two recording sites positioned 3 and 8 cm above the LOS). Peristaltic contractions of long duration (>6 sec) are found commonly but are not required for manometric diagnosis of nutcracker esophagus (NE).

NE has been described in 27%-48% of patients with noncardiac chest pain (3). This manometric pattern is often unassociated with pain. The underlying mechanism for chest pain is obscure. In some patients there is coexisting acid reflux, in others, an underlying psychological problem has been observed (4). Transition of NE to other motility disorders raises speculation that it lies in the beginning of spectrum of motility disorder that ends in achalasia.

Chest pain is the predominant symptom (90%) and dysphagia is less common. (3) Symptoms intensity, frequency, and location vary. Most patients are evaluated for chest pain syndrome and referred to gastroenterologist after ruling out cardiac disease. Associated symptoms include depression, anxiety, and somatization; but not apparent on routine evaluation. Physical examination is invariably
normal.

All patients have normal peristalsis hence barium studies and radionuclide transit studies are normal. Association of epiphrenic diverticulae has not been reported with NE which was present in our case. Epiphrenic diverticulae are rare and occur in the terminal 10-15 cm of the esophagus these pulsion diverticula often occur secondary to motility disorders of the esophagus, DES (24%) or achalasia (15%). It's rarely reported along with NE. Symptoms are often due to motor disorder, rather than the diverticulum itself.

Most patients with diverticulum require no specific therapy. Surgery is usually reserved for people with significant progressive dysphagia or recurrent aspiration pneumonia. Diagnosis of NE requires manometry. Various criteria for diagnosis was used by various authors, most experts use the above mentioned criteria. Long term manometric follow up failed of show consistent findings. Evolution of NE to diffuse esophageal spasm and achalasia has been reported.

Management includes reassurance psychological intervention. Suggested approach is a trial of anti-reflux treatment followed by nitrates (ISDN 5-10 mg SL), calcium channel blockers (Nifedipine 10-30 mg QID), visceral analgesics (Imipramine 50 mg HS), sedatives (Trazadone, Alprazolam). Diltiazem 60-90-mg TID has been tried with some success. Nonresponder occasionally respond to botulinum toxin injection and myotomy.

We conclude that nutcracker esophagus is a manometric abnormality unusually associated with pulsion diverticulum of esophagus. Management of NE and epiphrenic diverticulum must be individualized based on severity of symptoms.

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