



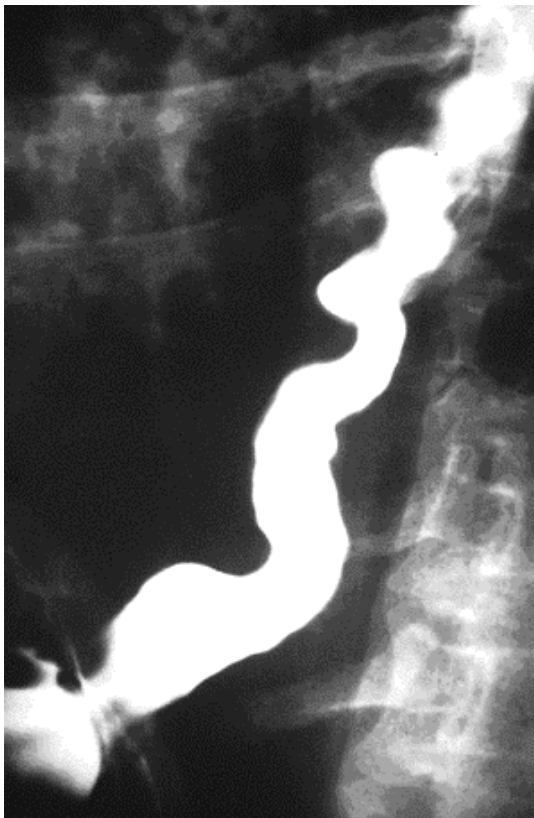
Holy C.O.W.!

It's...

Clinical Question of the Week #19
November 3rd, 2008 through November
10th, 2008

Please e-mail your answers to Kuo, Tim, Wendy, and Kevin (klian@mednet.ucla.edu; tprovias@mednet.ucla.edu; wsimon@mednet.ucla.edu; kbreger@mednet.ucla.edu) by 0800 on Monday, November 10th, 2008. The resident or intern with the most correct answers at the end of each month will receive a prize!

Case: A 75-year-old woman presents to the Emergency Department with chest pain for the third time in the past four months. Each time she is found to have no significant EKG changes and negative cardiac enzymes, and is discharged. Her pain subsided, however, after sublingual nitroglycerin the last two times she was seen in the ED. She underwent an adenosine myoview two years ago, which revealed no evidence of reversible ischemia. She is now admitted to the Observation service, where the intrepid resident orders an upper GI series, shown below.



Upper GI series with barium contrast

Questions:

1. What is the diagnosis? What are two other related syndromes?

Diffuse esophageal spasm (DES), considered one of the esophageal motility disorders (EMD), is defined as 20% or more simultaneous contractions with amplitude >30mmHg. The primary pathologic EMD is achalasia, which is defined as absent distal peristalsis, incomplete LES relaxation, and elevated resting LES pressure. Minimal information is known on the pathophysiology of DES, although some studies have suggested an abnormality in endogenous nitric oxide synthesis or degradation, possibly explaining the observation that nitrates provide some symptomatic benefit in these patients.

In addition to DES, the other abnormal motility patterns consist of two hypercontracting phenomena - nutcracker esophagus (hypertensive peristalsis) and hypertensive LES, as well as two hypocontracting phenomena – ineffective esophageal motility and hypotensive LES. Finally, other causes of esophageal dysmotility consist of retrograde contractions, triple-peaked contractions, and isolated incomplete LES relaxation.

Esophageal dysmotility may also be associated with systemic disease, as may be the case with chronic GERD, diabetes, systemic sclerosis, Chagas' disease, and chronic idiopathic pseudo-obstruction. (1)

2. Name the three cardinal symptoms of this condition.

Symptomatic esophageal dysfunction usually manifests as heartburn, dysphagia, and chest pain. The diagnosis should be included in the differential of a patient presenting with unexplained chest pain after a cardiac etiology has been excluded. (0.5)

3. What is the finding shown in the image above?

The finding in the image is the "corkscrew" esophagus, although the term "rosary bead" esophagus has been used. The image reflects multiple tertiary simultaneous contractions interrupting the barium column in the esophagus. However, it is of note that radiologic studies may also be entirely normal in DES, thus leading to poor sensitivity/specificity of these studies. The diagnostic examination of choice is multichannel impedance testing to evaluate esophageal pressure at multiple points along the esophageal course. (0.5)

4. What is the treatment?

While no standard exists for therapy given the rarity of these conditions, treatments supported by clinical trial data include calcium channel blockers such as diltiazem and antidepressants such as trazodone and imipramine (TCAs, which probably reduce the visceral sensory perception of discomfort rather than alleviating the dysmotility). Positive anecdotal results have also been seen in nitrates (NTG, isosorbide dinitrate), anticholinergic drugs (dicyclomine), phosphodiesterase inhibitors (sildenafil), and botox injections. Physical manipulation has been marginally beneficial in bougie dilation, but not in pneumatic dilation or esophagomyotomy. (1)