Diffuse Esophageal Spasm: Transforming Into Achalasia

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Introduction

Of 130 patients diagnosed to have esophageal motility disorders, transformation of diffuse esophageal spasm (DES) into achalasia was identified in one patient. The clinical syndrome of DES was first described by Hamilton Osgood in 1889\(^1\). It is characterized by symptoms of substernal chest pain and dysphagia, tertiary contractions on barium swallow and a manometric pattern of frequent simultaneous contractions, interspersed with normal peristalsis. It accounts for 4-15\% of the esophageal motility disorders identified in patients with non-cardiac chest pain\(^2,3\). Various lower esophageal sphincter (LES) abnormalities i.e., incomplete sphincter relaxation, hypertensive LES have been reported to coexist with diffuse esophageal spasm\(^4,5\). This suggests that some of these disorders may represent a dynamic spectrum of esophageal dysmotility rarely, transforming one into the other condition\(^7\). This report describes one case transforming from DES to achalasia.

Case Report

A 35 years old male presented with a 9 month history of mild retrosternal pain associated with swallowing. Cardiac evaluation by a consultant cardiologist was normal. Barium swallow evaluated at fluoroscopy showed tertiary contractions at mid and lower esophagus (Figure 1).
Upper G.I. endoscopy (EGD) was normal. His esophageal manometry was performed using an 8 lumen polyvinyl catheter, a low compliance pneumohydraulic capillary perfusion system (Arndorfer medical specialties Greendale, USA) and a recording physiograph (Hewlett Packard, Chicago). Lower esophageal sphincter (LES) pressure was measured at the peak end expiratory phase as the mean of 4 values and found to be 10 mm Hg with complete relaxation on wet swallow (WS). Three simultaneous and 8 propagated peristaltic contractions were noted after 10 WS with normal upper esophageal sphincter pressure. Intravenous edrophonium 0.08 mg/kg was given, showing increase in amplitude and duration of contractions with intervening multiple peaks and repetitive contractions (Figure 2).
He experienced mild retrosternal chest pain, relieved after 10 minutes. A final diagnosis of DES was made for which diltiazem 60 mg 8 hourly was given with marked relief of symptoms. During one year of follow-up, he noted improvement of chest pain and choking sensation, however, he continued to have a feeling of food lodging in lower chest. Repeat barium swallow at this time showed markedly dilated esophagus with smooth tapering of the gastroesophageal junction (Figure 3).
Esophageal manometry showed resting LES pressure of 13 mm Hg, residual pressure of 3 mm Hg and aperistalsis in the esophageal body, suggestive of achalasia (Figure 4).
He underwent balloon dilatation after informed consent with a 35 mm pneumatic (microvasive) balloon. This resulted in relief of dysphagia with no recurrence of chest pain during 1 year follow-up.

**Discussion**

Transformation of DES into achalasia has been reported in the literature as a rare occurrence. Diffuse esophageal spasm (DES) accounts for 4-15% of the esophageal motility disorders identified in the non-cardiac chest pain patients\(^8\). It is a difficult condition to diagnose as most of the patients are usually labeled as “cardiac patients”, despite normal or equivocal investigations. Some patients with DES who respond to nitrates and calcium channel blockers\(^9-13\) are presumed to have ischaemic heart disease. Unfortunately the routine upper GI investigations i.e., barium swallow and EGD may be normal in many patients and its only with esophageal manometry, that the diagnosis is confirmed. Manometric findings in most patients may not show the high amplitude, simultaneous contractions unless the patient is symptomatic for chest pain at the time the test is conducted, thus necessitating provocative test as intravenous edrophonium. This may increase the contraction amplitude with reproduction of chest pain simulating the clinical situation\(^14\). This may occur in 18-30% patients with DES\(^15-16\). Transformation from DES into achalasia, as in this patient, shows a dynamic nature of motility disorders. Most patients with achalasia at the time of diagnosis have a well established disease with all features of completely developed achalasia, so one does not get a grasp on “achalasia in evolution” which may actually be more common.
than observed. This notion is further substantiated in a follow up study of DES transforming into achalasia, where this transformation took as long as 3.7 years. Others have reported similar transition, well documented symptomatically, radiologically and on manometric evaluation as presented in Table 16,18,19. Vigorous achalasia shares common presentation with DES i.e., chest pain and dysphagia but may be distinguished on manometric evaluation showing high amplitude simultaneous contractions with 10% normal intervening persistalsis whereas in vigorous achalasia normal persistaltic activity is not seen. This case further supports that DES may transform periodically into achalasia if such cases are followed-up diligently. Once established, they may respond to standard therapy for achalasia, which is usually more gratifying than the treatment of DES.

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References