

Diffuse Esophageal Spasm as a Cause of Chest Pain: Case Reports

Altat Alam, S. Waqar Hassan, Arshad Kamal Butt, Anwaar A. Khan
Department of Gastroenterology, Shaikh Zayed Hospital, Lahore

SUMMARY

Non-cardiac chest pain is a common clinical problem. 30% of patients presenting with chest pain have a non cardiac cause e.g. Nutcracker esophagus (N.E.), diffuse esophageal spasm (DES). We present 3 such cases who complained of dysphagia and chest pain and after complete work up were diagnosed to have diffuse esophageal spasm.

CASE 1

Fifty two year old female, a college teacher, presented in the out patient department of Shaikh Zayed Hospital-Lahore, with 3 years history of dysphagia for both solids and liquids and regurgitation of food with vomiting. In addition, she also complained of severe crushing chest pain usually after meals and sometimes on empty stomach, necessitating frequent emergency check ups. Her cardiac evaluation along with ECGs and exercise tolerance test (ETT) were reported normal on several occasions. There was no significant history of weight loss. Her physical examination was normal. Initial laboratory investigations were normal. Barium swallow showed marked segmentation of barium in the esophagus, suggestive of corkscrew esophagus (Fig. 1). Her Upper GI endoscopy was normal but esophageal manometry showed the following salient features;

- Lower esophageal sphincter (LES) pressure of 26.10 mmHg with incomplete relaxation on wet swallow (W.S).
- Simultaneous non-peristaltic contractions along with intermittent normal peristalsis in the body of the esophagus on W.S.
- Intravenous Tensilon (Edrophonium Hydrochloride), 0.08 mg/kg, resulted in increased amplitude in the esophageal body upto 250 mmHg with double and triple peaked contractions (Fig 2). It was associated with crushing chest pain, relieved after a few minutes. In normal subjects, there may be

increased contraction amplitude and duration but no pain induced with I.V. Tensilon.



Fig. 1: Diffuse esophageal spasm with typical curling or corkscrew appearance.

Final diagnosis of diffuse esophageal spasm (DES) with incomplete relaxation of LES was made and dilatation of LES was planned. After dilatation with 30 mm diameter (microvasive) pneumatic balloon, she remained symptomatic for chest pain but dysphagia was partially relieved. Diltiazem, a calcium channel blocker, 30 mg three times a day resulted in 80% relief in chest pain.

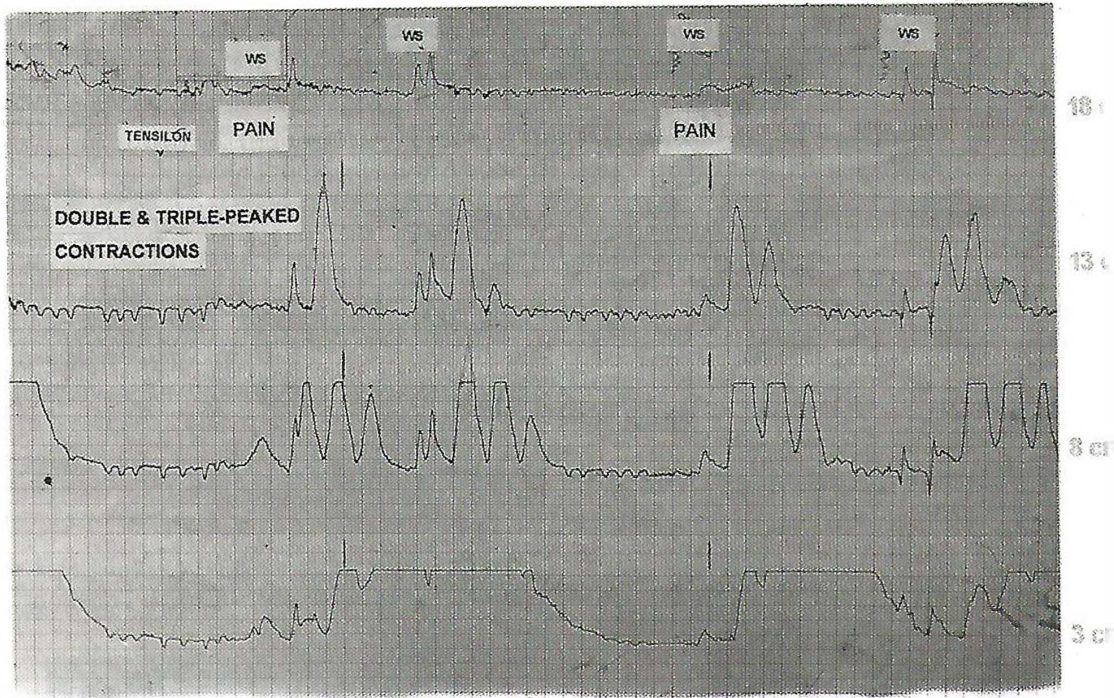


Fig. 2: Double and triple peaked contractions in a patient with diffuse esophageal spasm (case 1) after receiving edrophonium (Tensilon).

CASE 2

Thirty five year old male, dispenser by profession, presented with 9 months history of dysphagia, mild retrosternal pain provoked by swallowing. Cardiac evaluation, including ETT was normal. Barium swallow outlined whole of the esophagus with tertiary contractions in the mid part. Upper G.I. endoscopy (EGD) was normal. His manometry showed following findings;

- a. LES pressure of 10 mm Hg with complete relaxation on WS.
- b. Two simultaneous and 8 normal propagated waves after 10 WS.
- c. After giving 0.08 mg/kg I.V. Tensilon, amplitude and duration of contractions increased with intervening multiple peaks and repetitive contractions (Fig 3). Patient experienced mild retrosternal chest pain, relieved after a few minutes.
- d. Upper esophageal sphincter was normal.

Final diagnosis of DES was made and patient was prescribed Diltiazem 30 mg q 8 hourly which relieved his symptoms.

CASE 3

Thirty six year old male shopkeeper by profession, presented with one and half years history of epigastric pain radiating to retrosternal area and shoulders, non responsive to repeated courses of H₂ blockers and proton pump inhibitors. Barium swallow and upper GI endoscopy was normal. Esophageal manometry revealed the following changes;

- a. LES pressure of 15 mm Hg with complete relaxation.
- b. Normal esophageal peristaltic waves in the esophageal body.
- c. After I.V. Tensilon 0.08 mg/kg amplitude and duration of waves increased. 10 WS showed 3 simultaneous and 7 propagated peristalsis. Patient experienced mild retrosternal chest pain, relieved after a short time.

Final diagnosis of DES was established and he was prescribed Diltiazem 30 mg 8 hourly to which he responded very well.

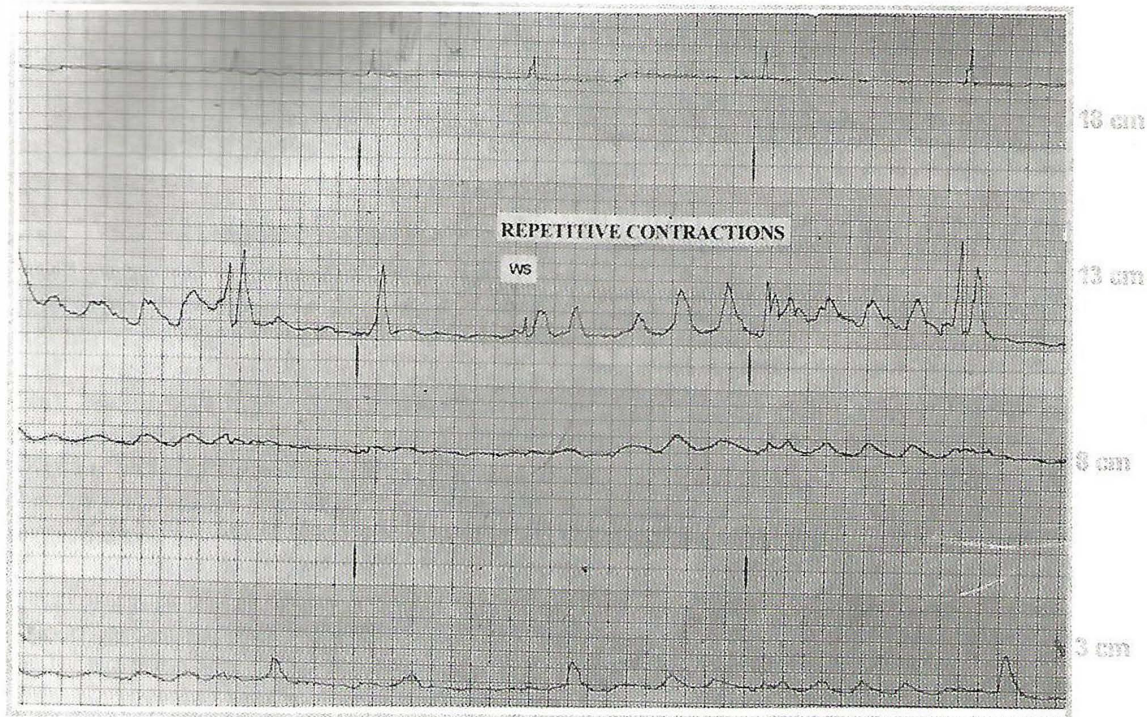


Fig. 3: Classical repetitive contractions in a patient with diffuse esophageal spasm (case 2).

DISCUSSION

These three patients presented with symptoms of chest pain and dysphagia. Cardiac evaluation and upper GI endoscopy was normal in all of them. Barium swallow showed tertiary contractions in case 1 and 2 while it was normal in case 3. Esophageal manometry and edrophonium stimulation test confirmed the diagnosis of diffuse esophageal spasm in all 3 cases with associated findings of incomplete relaxation of LES in the first case.

DES accounts for 4-15 % of the esophageal motility disorders identified in the non cardiac chest pain patients¹. It is a difficult condition to diagnose as most of the patients have already had full cardiac evaluations and many are labelled patients with coronary heart disease (CHD), despite normal or equivocal investigations. DES patients respond to nitrates and calcium channel blockers²⁻⁶, prescribed for presumed CHD. If other drugs like beta blockers are prescribed for presumed CHD, response is usually poor. Unfortunately routine upper GI investigations like barium swallow and, EGD may be normal in many patients. It is only when esophageal motility studies are performed, that a diagnosis is

confirmed. The manometric criteria for diagnosing DES include, simultaneous contractions in (>10% of wet swallows) and intermittent normal peristalsis. Associated findings may include repetitive contractions (>3 peaks), prolonged duration of contractions (> 6 seconds), which may be of high amplitude and frequent repetitive contractions. LES may show high resting pressure or incomplete relaxation. In patients suspected to have DES, provocative test with edrophonium is recommended during esophageal manometry. It produces increase in amplitude and duration of esophageal contractions, both in normals and in patients with abnormal motility disorders but chest pain is provoked in 18-30% of non cardiac chest pain patients only^{7,8}.

The mainstay treatment of this condition is pharmacological. Nitrates and calcium channel blockers²⁻⁶, will relieve pain in majority of patients. In refractory cases, pneumatic balloon dilatation and surgical myotomy have been reported to give good results^{9,10}. Psychological intervention with counseling or biofeedback therapy have shown to reduce symptoms in patients with esophageal motility disorders but have not been studied extensively.

REFERENCES

1. **Katz PO, Dalton CB, Richter JE, et al.** Esophageal testing of patients with non cardiac chest pain and/or dysphagia. Results of a three year experience with 1161 patients. *Ann Intern Med* 1987; **106**: 593-597.
2. **Shlemmel A, Priviteri CA, Poppel MH.** A study of the effects of certain drugs on curling of the esophagus. *Am J Roentgenol* 1949; **62**: 807.
3. **Swamy N.** Esophageal spasm and manometric response to nitroglycerin and long acting nitrates. *Gastroenterology* 1977; **72**: 23-27.
4. **Orlando RC, Bozyski EM.** Clinical and manometric effects of nitroglycerin in diffuse esophageal spasm. *N Engl J Med* 1973; **289**: 23-25.
5. **Blackwell JN, et al.** Effect of nifedipine on esophageal motility and gastric emptying. *Digestion* 1981; **21**: 50.
6. **Richter JE, et al.** Effects of oral calcium blocker, diltiazem, on esophageal contractions. *Dig Dis Sci* 1984; **29**: 649-656.
7. **Benjamin SB, Richter JE, Cordora CM, et al.** Prospective manometric evaluation with pharmacologic provocation of patients with suspected esophageal motility dysfunctions. *Gastroenterology* 1983; **84**: 893-901.
8. **Richter JE, Hackshaw BT, Wu WC.** Edrophonium: a useful provocative test for esophageal chest pain. *Ann Intern Med* 1985; **103**: 11-21.
9. **Ebert EC, et al.** Pneumatic dilatation in patients with symptomatic diffuse esophageal spasm and lower esophageal sphincter dysfunction. *Dig Dis Sci* 1983; **28**: 481-485.
10. **Brown M, Stacier MS, May ES.** Esophageal myotomy and treatment of "nutcracker esophagus". *Am J Gastroenterol* 1987; **82**: 1331.

The Authors:

Altaf Alam,
Assistant Professor,
Department of Gastroenterology,
Shaikh Zayed Hospital,
Lahore.

S. Waqar Hassan,
Registrar,
Department of Gastroenterology,
Shaikh Zayed Hospital,
Lahore.

Arshad Kamal Butt,
Senior Registrar,
Department of Gastroenterology,
Shaikh Zayed Hospital,
Lahore.

Anwaar A. Khan,
Professor,
Department of Gastroenterology,
Shaikh Zayed Hospital,
Lahore.

Address for Correspondence:

Altaf Alam,
Assistant Professor,
Department of Gastroenterology,
Shaikh Zayed Hospital,
Lahore.