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LETTER

Achalasia cardia: A diagnosis often delayed!

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Sir,

Idiopathic achalasia cardia is a primary motor disorder. It is diagnosed and subclassified into three types based on esophageal manometry findings. Pneumatic dilatation, laparoscopic Heller's myotomy and peroral endoscopic myotomy (POEM) are the commonly used treatment modalities for this disorder [1, 2]. The predominant symptoms of achalasia include dysphagia, regurgitation, chest pain and weight loss [1]. These symptoms are also noted in gastroesophageal reflux disease (GERD). It is commonly noted that diagnosis of achalasia is often delayed, sometimes even up to years, before definitive treatment is offered. The present retrospective study was done with the aim to identify factors leading to delayed diagnosis in patients with achalasia cardia.

This retrospective study was done using the medical records of 88 cases of achalasia cardia diagnosed using high-resolution esophageal manometry by the author between 2011 and 2017, using Chicago classifications (CC) 2.0 and v3.0. Tracings reported as per CC 2.0 were reanalyzed using CC v3.0 for confirmation of diagnosis and further subtyping. Details noted were age, sex, symptoms with duration, Eckardt's score at presentation, consultations and investigative procedures done prior to confirmed diagnosis. Sufficient data were not available for 25 patients and, hence, they were excluded from the analysis. The collected data were expressed as percentages, median and range. Comparison of proportions was done using chi-square test and comparison of medians was done using Mann-Whitney test. A p-value of < 0.05 was considered statistically significant.

Sixty-three patients (males 42, 67%) formed the study cohort. The median age at presentation was 44 (23–62) years. Dysphagia was present in 61 (96.8%), regurgitation in 54 (85.7%), chest pain in 34 (54%), weight loss in 34 (54%) and other symptoms like cough and chest infections in 8 (13%). The diagnosis was confirmed after a median period of 18 months (6–36 months) after the onset of symptoms. The vast majority (52, 82.5%) had consulted non-gastrointestinal (GI) specialists like general physicians, ear, nose and throat (ENT) specialist, cardiologists and general surgeons at first presentation. Three-fourths (47, 74.6%) of the cases had consulted more than two doctors for their symptoms. Barium swallow and esophagogastroduodenoscopy (EGD) had been done in 52 (82.5%) and 61 (96.8%), respectively. EGD had been repeated more than once in 8 cases (13%).

Twenty-one cases were diagnosed within 12 months of onset of symptoms. These were labelled as group I and compared with those diagnosed beyond 12 months (group II). Most patients in group II had been labelled as GERD (38/42, 90.5%) prior to diagnosis of achalasia. EGD and barium swallow was done in 42 (100%) and 36 (85.7%), respectively. EGD was reported as normal in most cases (36, 85.7%) and low-grade esophagitis was noted in the remaining. Candida infection was noted in 3 cases. Barium swallow was reported as non-specific esophageal motility disorder, normal and lower oesophageal luminal narrowing in 24 (66.7%), 6 (16.7%) and 6 cases (16.7%), respectively. The factors that determined the earlier diagnosis (< 12 months from symptom onset) were an urban residence, first presentation to GI specialist and greater symptom severity as assessed by higher Eckardt's score (Table 1). Based on high-resolution esophageal manometry, it was noted that type 1 achalasia was the commonest subtype (39.7%), followed by type 3 (20, 31.7%). However, there was no statistically significant difference between the three subtypes in terms of time taken to reach the confirmed diagnosis (p-value = 0.69). Achalasia cardia presents with classical symptoms as discussed earlier. However, the time interval between the onset of symtoms and the correct diagnosis gets prolonged. This period is frustrating for the patients and associated with avoidable expenditures. Despite the presence of a straightforward clinical investigation for dysphagia (high-resolution esophageal manometry), it is alarming to note that patients go on without definitive diagnosis for long periods, despite consulting many doctors. The delay in diagnosis

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Table 1 Comparison of study parameters between group I and II

Parameters	Group I (n=21)	Group II (n=42)	p value
Age in years (median, range)	42 (23-59)	46 (28-62)	0.23
Males	13 (61.9%)	29 (69%)	0.57
Residence (urban)	16 (76%)	12 (28.5%)	0.0004
Symptoms at presentation			
Dysphagia	21 (100%)	40 (95.2%)	
Regurgitation	18 (85.7%)	36 (85.7%)	0.68
Chest pain	15 (71.4%)	19 (45.2%)	
Weight loss	14 (66.7%)	20 (47.6%)	
	9 (7-10)	7 (5-9)	
Eckardt score at presentation (median, range)			0.03
First presentation directly to GI specialist	8 (38.1%)	3 (7.1%)	0.002
Use of endoscopy +/-barium swallow			
Endoscopy	19 (90.5%)	42 (100%)	NA
Barium swallow	16 (76.2%)	36 (85.7%)	0.35
Sub type of achalasia			
Type 1 (n=25)	7 (28%)	18 (72%)	
Type 2 (n=18)	6 (33.3%)	12 (66.7%)	0.69
Type 3 (n=20)	8 (40%)	12 (60%)	

GI gastrointestinal, Type 1, 2, 3 are subtypes of achalasia based on Chicago classification v3.0

has been reported to be 4.7 years in earlier studies [3]. A recent study from Germany noted that it still takes almost 2 years until the correct diagnosis of achalasia is confirmed [4]. An earlier Indian study also noted that duration of dysphagia was significantly longer (nearly 21 months) in patients with achalasia cardia compared to those with normal manometry study [5].

We noted that urban residence, greater severity of symptoms and first presentation to a GI specialist led to early diagnosis of this disorder. This stresses the point that access to proper and specialized medical care helps to make an early diagnosis. As GI specialists, it is our duty to educate medical professionals regarding this condition, especially regarding the use of clinical scoring systems like Eckardt's score. Moreover, frequent utilization of high-resolution esophageal manometry in patients with non-resolving reflux symptoms and dysphagia may help to correctly diagnose the condition.

The present study is limited by small sample size and a retrospective design. Despite this, it emphasises that the time to first diagnosis of achalasia still takes far too long. Tests like esophageal manometry are not utilized fully, probably due to lack of availability or knowledge regarding these tests among doctors. Early diagnosis is likely to minimize prolonged symptomatic period, prevent long-term consequences of the disease and reduce health care costs.

Compliance with ethical standards

Conflict of interest MJ declare that they have no conflict of interest.

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