Natural history of Achalasia: A descriptive study

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Introduction

Achalasia is a rare disorder of the esophageal smooth muscle characterized by impaired relaxation of the lower esophageal sphincter (LES) and absent or spastic contractions in the esophageal body[1]. The crude prevalence in Medicare of achalasia per 100,000 persons aged 65 or older was 162.1 in 2015[2]. High-resolution manometry (HRM) is the gold standard for diagnosis and based on this, this disease is further categorized into 3 distinct 3 types: type I, type II and type III. Treatment is aimed at relieving symptoms because there is no cure for achalasia[3]. This may be achieved through calcium channel inhibitors, pneumatic dilation of the LES, botox injections, peroral endoscopic myotomy (POEM), Heller myotomy and ultimately, esophagectomy.

Aim

This study aims to delineate the natural history of achalasia, emphasizing the infrequency of esophagectomy as a treatment outcome, examining the spectrum of therapeutic alternatives preceding esophagectomy, and establishing a timeline for disease progression. Another objective of the study is to try to identify variables may predict therapeutic failure.

Methods

We conducted a retrospective descriptive study of all patients diagnosed with achalasia between 2019 and 2025 at our institution. Data was extracted from institutional medical records. Achalasia was diagnosed using a timed barium swallow (TBS) study and/or HRM. Patients who had an esophagectomy for any other diagnosis that was not achalasia, were ruled out of the study. Statistical significance of the mean of quantitative variables was measured with paired-t tests and chi square and Fisher's test were used for comparison of qualitative values.

Results

A total of 147 patients were included. Five patients underwent esophagecrtomy for achalasia (**3.4%**), while 142 patients were treated with Heller myotomy. Patient demographics, comorbidities, and presenting symptoms are summarized in Table 1.

In the esophagectomy cohort, four patients had a prior myotomy before esophagectomy. The average duration of symptoms prior to esophagectomy was 13 years. Among those with prior myotomy, the average interval to esophagectomy was 10.5 years. All patients in this cohort had end-stage achalasia. Procedures were robotic, with one case requiring an open thoracic portion and one conversion to open. Two patient developed postoperative leaks, and four experienced symptom recurrence, with an average time of 145 days after esophagectomy.

In the Heller myotomy cohort, patients reported an average symptom duration of 7.5 years prior to intervention. Prior treatments included Botox injection (32), esophagogastroduodenoscopy (EGD) dilation (89), POEM (3), and percutaneous endoscopic gastrostomy (PEG) tube placement (1). At <6 months follow-up, 1.49% of patient reported worsening symptoms compared with preoperative status. At last follow-up, 4 patients reported worse symptoms than preoperatively. Symptom recurrence occurred in 27 patients, with an average time to management of 383 days after myotomy.

Conclusions

Esophagectomy is a rare outcome in the treatment of achalasia. In our cohort, Heller myotomy provided excellent symptom control at six month and at last follow-up. No predictive variables for therapy failure were identified.

References

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Table 1. Patients demographics, comorbidities and symptoms per patient group

		Patients requiring esophagectomy	Patients not requiring esophagectomy	P value
N		5	142	
Sex (%)	Male Female	2 (40) 3 (60)	91 (64) 51 (36)	0.5194
Race (%)	Asian Black or African American Other White	0 0 1 (20) 4 (80)	3 (2.1) 12 (8.4) 24 (16.9) 103 (72.5)	0.8982
Hispanic ethnicity (%)		2 (40)	23 (16.1)	0.4013
Age at procedure (mean)		55.4	56.69	0.8024
Hypertension (%)		0	64 (45)	0.4367
GERD (%)		4 (80)	70 (49)	0.3749
Diabetes (%)		0	14 (9.8)	0.8785
Hyperlipidemia (%)		1 (20)	45 (31)	>0.9999
Smoking (%)		2 (40)	57 (40.1)	>0.9999
Alcohol use disorder (%)		0	7 (4.9)	0.5059
Heartburn (%)		3 (60)	31 (21.8)	0.1625
Cough (%)		1 (20)	16 (11.3)	0.9280
Weight loss (%)		4 (80)	67 (47)	0.3253

Table 2. Recurrence rate, recurrence management and prior interventions in patients who had only a Heller myotomy

		Patient who underwent Heller myotomy
N		142
Prior Botox (%)		32 (22.5)
Average # of Botox		1.5
Prior EGD dilation (%)		89 (62)
Average # of dilations		1.7
Prior POEM (%)		3 (2.1)
Prior EGD with PEG tube (%)		1 (0.7)
Average days from Heller myotomy to last office visit		227
Recurrence of symptoms (%)		27 (19)
Management of recurrence[1]	Redo Heller myotomy (%) Botox injection (%) EGD with dilation (%) Medications (%) Other (%)	3 (11.1) 2 (7.4) 10 (37) 8 (29.3) 8 (29.3)
Average days from myotomy to management of recurrence		383

[1] Patients may have had more than one treatment for recurrence

Figure 1. Patient-reported symptoms at <6 months follow-ups