CASE REPORT

Recurrent episodes of esophageal candidiasis without dysphagia post-Guillain-Barré syndrome: an unusual presentation of achalasia

Daniel Runggaldier, Michael Fried, Daniel Pohl

Department of Gastroenterology and Hepatology, University Hospital of Zurich, Zurich, Switzerland

Correspondence to
PD Dr. med. Daniel Pohl,
Daniel.Pohl@usz.ch

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SUMMARY

Here, we present a case of a 50-year-old male with a history of a Guillain-Barré-syndrome, who was referred to our clinic with recurrent esophageal candidiasis and long-standing intermittent retrosternal cramps for further evaluation. Other symptoms such as dysphagia, regurgitations and weight loss were denied, and prior repeated endoscopy was otherwise unremarkable. Using high resolution impedance manometry, we could demonstrate a panesophageal pressure increase on water swallows and complete aperistalsis of the tubular esophagus, indicating achalasia type II. However, due to the patient's extraordinary body height and resulting length of the esophagus, endoluminal functional lumen imaging probe analysis supplementary to high resolution impedance manometry needed to be used to assess distensibility of the esophagogastric junction and to secure the diagnosis of achalasia before appropriately treating the patient with pneumatic dilation.

BACKGROUND

Achalasia is a rare esophageal motility disorder characterised by aperistalsis and impaired relaxation of the lower esophageal sphincter (LES). The incidence of the disorder is estimated to be about 1.6 cases and prevalence about 10 cases per 100 000 population.^{1 2} The major clinical manifestations include dysphagia for both liquids and solids, retrosternal pain as well as instantaneous regurgitations on deglutition and may be complicated by progressive weight loss.³ Achalasia is

caused by neurodegeneration of ganglion cells along the wall of the esophagus and the region of the LES, which is associated with inflammatory and fibrotic changes. Viral infections such as Herpes Simplex or Varicella Zoster triggering autoimmune processes are currently considered as the potential underlying pathogenesis of the disease.⁴⁻⁶ Various autoimmune disorders such as Sjogren's syndrome, systemic lupus or rheumatoid arthritis have all been associated with achalasia.⁷

The gold standard in diagnosing achalasia is esophageal manometry, detecting functional obstruction at the level of the LES and aperistalsis of the esophageal body.⁸ The endoluminal functional lumen imaging probe (endoFLIP) has recently emerged as a new diagnostic modality, which measures the distensibility as well as the opening dynamics of the esophagogastric junction (EGJ). However, due to a current lack of prospective randomised trials supporting its utility as well as a paucity of standardised protocols, its clinical relevance in large patient sets is yet to be confirmed.⁹

Here, we present a patient with a history of Guillain-Barré syndrome (GBS) developing achalasia with an atypical and unusual symptomatology.

CASE PRESENTATION

A 50-year-old 7 feet tall male was referred to our clinic with a history of recurrent esophageal candidiasis and progressive intermittent retrosternal cramps. The patient reported a history of Guillain-Barré Syndrome GBS without respiratory

Figure 1 (A) HRM Clouse plot showing aperistalsis and pan-esophageal pressure increase instantaneously on water swallows. Due to the significant length of the esophagus, both esophageal sphincters cannot be completely covered by the catheter. Hence, important HRM parameters such as the integrated relaxation pressure could not be precisely determined. (B) Combined HRM Clouse and impedance plot showing aperistalsis and ineffective clearance during the rice meal. HR(I)M, high-resolution (impedance) manometry; LES, lower esophageal sphincter; UES, upper esophageal sphincter.

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Figure 2 (A) Endoluminal functional lumen imaging probe analysis showing a restricted and pathologic distensibility of the esophagogastric junction (EGJ). (B) In comparison, a non-pathologic distensibility of a normal EGJ.

insufficiency at age 18 and 38. At age 42, the patient developed intermittent periods of retrosternal cramps predominantly at night and independent of physical exertion or food intake. Dysphagia, regurgitation, heartburn, and weight loss were denied. An extensive cardiological workup showed no evidence of myocardial ischemia. Upper endoscopy was unremarkable except for esophageal candidiasis, which was treated with fluconazole. Subsequent HIV testing showed a negative result. The patient's condition did not improve and the patient was repeatedly (age 45, 47, 49) scoped. Again, each time an esophageal candidiasis was found and treated with fluconazole, however, without achieving relevant symptom amelioration. Due to the recurrence of fungal infections of the esophagus, rare immunological disorders were ruled out with appropriate workup.

INVESTIGATIONS

After referral to our clinic, we performed esophageal high resolution impedance manometry (HRIM) to assess a possible esophageal motor dysfunction. HRIM demonstrated complete aperistalsis and pan-esophageal pressure increase on standardised assessment with single swallows as well as insufficient esophageal clearance, as assessed by impedance measurements during a 200g rice provocation meal (figure 1). Due to the extraordinary body height and the resulting length of the esophagus, our manometry catheter was too short to completely cover the tubular esophagus and LES and hence accurately determine

the integrated relaxation pressure (IRP). A complementary endoFLIP analysis verified restricted and pathologic distensibility of the EGJ of 2.1 mm²/mm Hg (figure 2A). Compatible with these results, a barium swallow demonstrated stagnant contrast-medium column in the distal esophagus after 1, 3 and 5 minutes (figure 3A).

TREATMENT

Based on our test results suggesting the presence of achalasia type II, the therapeutic options were presented to the patient. On request, we performed a pneumatic dilation. At first, we used a 30 mm Rigiflex balloon dilating the EGJ for 1 min with 20 Psi under fluoroscopic control to minimise the risk for esophageal perforation followed by a 35 mm Rigiflex balloon dilation 3 months later. Progress monitoring with a barium swallow and improvement of the Eckardt score confirmed the successful intervention (figure 3B, table 1). Moreover, during the second dilation, no more esophageal candidiasis was observed.

OUTCOME AND FOLLOW-UP

Two months after second dilation, the patient was doing significantly better and only complained of infrequent retrosternal cramps of mild intensity. Subjectively, the patient described

Figure 3 (A) Barium swallows showing an impaired relaxation of the lower esophageal sphincter (LES) as well as a stagnant contrast-medium column in the distal esophagus after 5 min. (B) After the second dilation, a significantly improved passage of the contrast medium through the esophagogastric junction and complete transit after 5 min could be observed.

Table 1 Eckardt score before and after dilation treatment

	Weight loss (kg)	Dysphagia	Retrosternal pain	Regurgitation	Total score
Before first dilation	0	0	3	0	3
After second dilation	0	0	1	0	1

an overall 80%–90% improvement of symptom severity. Follow-up by symptom assessment and barium swallow is now planned in 12 months.

DISCUSSION

The above-described case of achalasia is in several ways remarkable. At first, our patient has a history of GBS. After an extensive literature review, we could find three cases reporting onset of achalasia with or after the development of a GBS.^{10–12} A common pathogenesis or a causal relationship, however, remains to be elucidated.

Both diseases are rare and a concomitant appearance in one individual patient estimated to be unlikely.¹² Mechanistically, a common autoimmune mechanism, potentially triggered by a gastroenteritis or respiratory tract infection is discussed as trigger in the literature.^{10–13} In our case, there was a long duration of approximately 4 years between the GBS and the onset of the first symptoms of achalasia. This could be presumably explained by a very slow disease progression with relatively mild symptoms, which remained atypical until our diagnosis 12 years later. Further, purely speculative and without any literature data, a ‘late-onset GBS associated achalasia’ could be proposed in our patient.

Second, the patient presented an unusual symptomatology. The major complaint was an intermittent feeling of retrosternal cramps for years mainly independent of food intake and accompanied with recurrent esophageal candidiasis. There were no other classical symptoms of achalasia such as dysphagia, regurgitation or weight loss reported. This proved to be of relevance in two ways. On the one hand, the atypical presentation can be considered accountable for the long duration of about 7 years with numerous diagnostic workups till the correct diagnosis was finally made. It clearly demonstrates that achalasia should be considered as a rare differential diagnosis of recurrent esophageal candidiasis with retrosternal cramps. Furthermore, it is interesting to note here that the esophageal candidiasis completely vanished after the dilation treatment making it likely that the candidiasis was caused by the persistent esophageal stasis associated with the impaired relaxation of the LES. On the other hand, the atypical symptomatology showed that the Eckardt score, which has so far been established as a useful disease severity scoring tool,¹⁴ does have certain limitations especially if atypical symptomatology of achalasia results in a very low initial score. This makes usage of the Eckardt score also problematic if monitoring of therapeutic success is intended (table 1).

Third, due to the patient’s extraordinary body height and the resulting length of the esophagus, the HRIM catheter was too short to fully cover the LES region. Hence, apart from aperistalsis and pan-esophageal pressure increase on swallowing, other parameters, such as the IRP, could not be precisely determined. However, according to current classification, the diagnosis of achalasia requires the detection of complete aperistalsis along with impaired deglutitive relaxation of the LES

with an IRP >15 mm Hg.⁸ By using the endoFLIP technology as a supplementary tool, we could further evaluate the EGJ and by detecting a pathologic distensibility secure the diagnosis (figure 2A). In comparison, a normal distensibility of the EGJ can be observed in a healthy esophagus (figure 2B). This demonstrates that endoFLIP is a powerful supplementary tool to further characterise esophageal motility disorders especially in cases where technical limitations of HRIM hamper a correct diagnosis.

Learning points

- ▶ The pathogenesis of the Guillain-Barré syndrome and achalasia might share common autoimmune and neurodegenerative processes.
- ▶ Achalasia should be considered as a rare differential diagnosis in recurrent esophageal candidiasis.
- ▶ In achalasia with asymptomatic presentation, the Eckardt score might be an insufficient tool in assessing disease severity and in monitoring the treatment success.
- ▶ Endoluminal functional lumen imaging probe is a novel and useful tool to evaluate the distensibility of the esophagogastral junction especially in cases where high resolution impedance manometry (HRIM) does not provide clear results or where the catheter is not well tolerated.

Contributors DR was involved in the writing of manuscript and data analysis. MF was responsible for the editing of manuscript and patient care. DP was involved in the data analysis, editing of manuscript and patient care.

Competing interests None declared.

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