Published online: 2019-09-26

Case Report

Dysphagia as initial presentation of primary amyloidosis

Piyush Ranjan, Mohan Goyal, Mandhir Kumar, Munish Sachdeva

Department of Gastroenterology, Sir Ganga Ram Hospital, New Delhi, India

Abstract

Amyloidosis involves all parts of the gastrointestinal tract including the esophagus. The esophageal involvement in amyloidosis has been reported to vary from 13% in a radiology study to 22% in an autopsy series; however, such patients have symptoms of gastroesophageal reflux. Dysphagia is an uncommon presentation of amyloidosis. We report a 64-year-old patient who presented with progressive dysphagia of 4 months duration which was confirmed to be due to primary amyloidosis with multiple myeloma. The esophageal involvement by amyloidosis was confirmed by esophageal mucosal biopsies, and 22-channel high-resolution manometry.

Key words

Amyloidosis, dysphagia, esophageal manometry, multiple myeloma, upper GI endoscopy

Introduction

Dysphagia occurs most commonly as a result of obstruction of esophageal lumen. Dysphagia without obstruction can be seen in neuromuscular and infiltrative disease.[1] Diagnosis in such cases depends on the findings of esophageal manometry and esophageal mucosal biopsies. Esophageal involvement in amyloidosis can rarely lead to dysphagia.[2] We report a rare case of primary amyloidosis presenting primarily as dysphagia.

Case Report

A 65-year-old lady presented with progressive dysphagia both for solid and liquids for 4 months. She had significant anorexia and weight loss of 20 kg. Her upper gastrointestinal (GI) tract endoscopy and barium swallow done prior to hospitalizion were normal. After admission, routine investigations revealed hemoglobin level as 10.4 g/dl and serum albumin level as 1.2 g/dl. The renal function tests were normal. Esophageal manometry revealed low esophageal body amplitude and non-propulsive contractions, but the lower esophageal sphincter (LES) pressure and its relaxation was normal [Figure 1]. Urine examination showed 3+ proteinuria and the 24-h urinary protein was 2.38 g. On the advice of a nephrologist, renal biopsy using methyl violet thioflavin T stain was done which showed amyloidosis [Figure 2]. Serum protein electrophoresis revealed monoclonal gammopathy (M spike) seen in gamma globulin region. Immunofixation electrophoresis identified "M spike" in lambda region. The examination of bone marrow aspiration revealed plasmacystosis of 11–12%. Subsequently, esophageal biopsies were obtained at upper GI endoscopy which also showed amyloid deposits [Figure 3]. Hence, a final diagnosis of primary amyloidosis with multiple myeloma, involving the esophagus in the form of motility disorder was made.

Discussion

Amyloidosis involves all parts of the GI tract. This is more frequent in primary amyloidosis (13–22%) as compared to secondary amyloidosis. Wheras the amyloid protein is predominatly deposited in the mucosa in secondary amyloidosis, the site of involvement in primary amyloidosis is muscularis. Clinical manifestations such as diarrhea, GI bleeding, and motility disorder depend upon the site of involvement.[3]

Esophageal involvement in amyloidosis has been reported. The prevalence of esophageal disease in amyloidosis ranges from 13% in a radiology study to 22% in an autopsy series.[4]

Access this article online

Website: www.jdeonline.in

Quick Response Code

DOI: 10.4103/0976-5042.129976

Address for correspondence:
Dr. Piyush Ranjan, Consultant Gastroenterologist, Sir Ganga Ram Hospital, New Delhi, India. E-mail: piyushranjan70@gmail.com

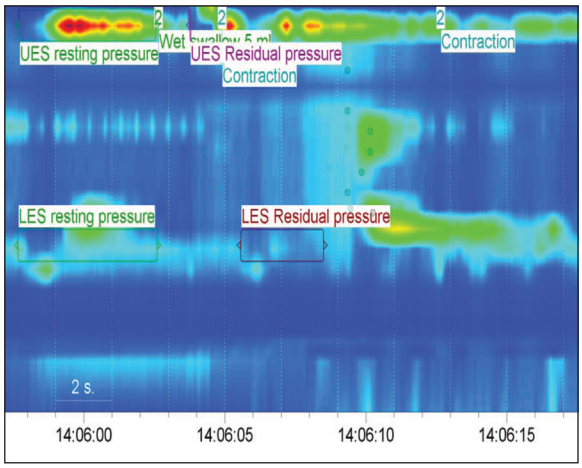

Figure 1: High-resolution esophageal manometry shows isobaric contractions suggestive of simultaneous contractions in the body with low amplitude. Lower esophageal resting pressure and relaxation were normal

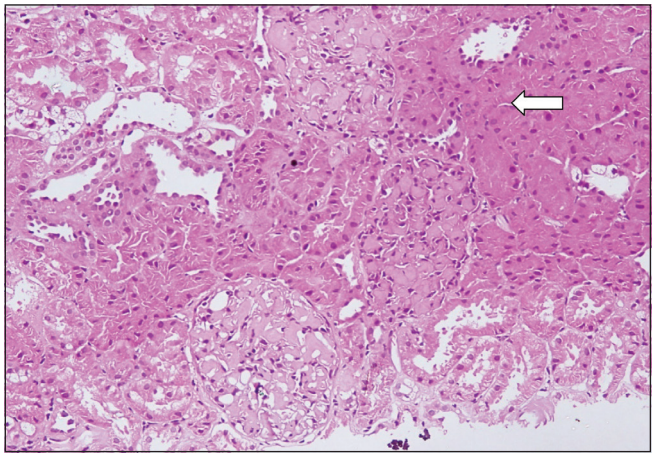

Figure 2: The glomeruli of kidney biopsy show amorphous eosinophilic material deposited in the extracellular space (hematoxylin and eosin staining, ×40)

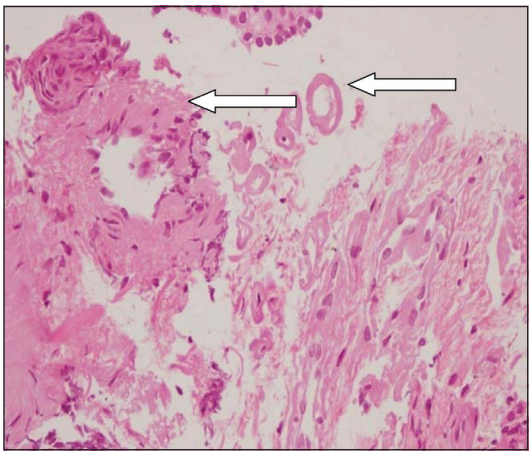

Figure 3: Esophageal biopsy shows medium-sized blood vessels with deposition of amorphous eosinophilic material within the media and extracellular space (hematoxylin and eosin staining, ×40)

However, the symptoms in these cases are those of gastric reflux. Our patient presented with dysphagia as the primary symptom. There is only one reported case of dysphagia as the presenting symptom of amyloidosis.

Manometry in patients with amyloidosis reveals various abnormalities. Manometric changes are more common in AL than in AA amyloidosis. The conventional manometry shows normal LES pressure, upper esophageal sphincter, and pharynx, but the amplitude of contraction is decreased.[5] Our case underwent 22-channel high-resolution manometry; the manometric findings showed some similarity to achalasia in the form of low-amplitude contraction and simultaneous and non-transmitted contractions, and LES pressure showed normal pressure as well as relaxation.

There are various hypotheses about the pathophysiology of esophageal involvement in amyloidosis. First, it may occur as a result of deposition of amyloid in the muscularis. Second, the deposition of amyloid in the submucosal vessels reduces the blood flow which causes deterioration in myentric plexus function. Third, vagal neuropathy can also compromise esophageal motility.[6]

Conclusion

To conclude, we have reported here a case of primary amyloidosis due to multiple myeloma, presenting with dysphagia as the presenting symptom. Dysphagia as the presenting symptom in primary amyloidosis due to esophageal involvement is rare. The deposition of amyloid results in manometric abnormalities, which in turn is responsible for dysphagia.

References

1. Richter JE. Oesophageal motility disorders. Lancet 2001;358:823-8.
2. Lefkowitz JR, Brand DL, Schuffler MD, Brugge WR. Amyloidosis mimics achalasia's effect on lower esophageal sphincter. Dig Dis Sci 1989;34:630-5.
3. Ebert EC, Nagar M. Gastrointestinal manifestations of amyloidosis. Am J Gastroenterol 2007;103:776-87.
4. Legge DA, Carlson HC, Wollaeger EE. Roentgenologic appearance of systemic amyloidosis involving the gastrointestinal tract. AJR Am J Roentgenol 1970;110:406-12.
5. Rubinow A, Burakoff R, Cohen AS, Harris LD. Esophageal manometry in systemic amyloidosis. A study of 30 patients. Am J Med 1983;75:951-6.
6. Tada S, Iida M, Iwashita A, Matsui T, Fuchigami T, Yamamoto T, et al. Endoscopic and biopsy findings of the upper digestive tract in patients with amyloidosis. Gastrointest Endosc 1990;36:10-4.

How to cite this article: Ranjan P, Goyal M, Kumar M, Sachdeva M. Dysphagia as initial presentation of primary amyloidosis. J Dig Endosc 2013;4:82-3.

Source of Support: Nil, **Conflict of Interest:** None declared.