CASE REPORT

Uncommon cause of dysphagia: paraneoplastic achalasia

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SUMMARY

Achalasia secondary to underlying neoplasm is a rare entity. Early recognition of secondary achalasia is important as its treatment involves management of underlying malignancy, while treatment of primary achalasia mainly involves lowering the lower oesophageal sphincter pressure with pneumatic dilatation or Heller's myotomy. We discuss an interesting case of achalasia secondary to non-Hodgkin's lymphoma.

BACKGROUND

Achalasia cardia is the motility disorder of the oesophagus diagnosed primarily on clinical grounds. The diagnosis is confirmed by oesophageal high-resolution manometry, endoscopy and radiological evaluation.

Distinction of the primary achalasia from secondary achalasia is important as these two differ in the treatment.¹ While the treatment of primary achalasia primarily involves lowering the resistance across lower oesophageal sphincter (LOS), management of secondary achalasia is treatment of underlying cause. Secondary achalasia is most commonly due to carcinoma of gastric cardia. Other causes include carcinoma of lower oesophagus, Chagas disease and metastatic disease.²

Figure 2 High-resolution manometry showing type II achalasia.

Early identification of secondary achalasia is of immense importance as delayed diagnosis may lead to progression of underlying malignancy with poor survival.³

CASE PRESENTATION

Fifty-seven-year-old, non-smoker male patient with diabetes presented with progressive, painless dysphagia more to liquids than solids and progressed rapidly over 6-month duration. On careful general examination, he was found to have firm, non-tender enlarged lymph nodes in posterior triangle of the neck. His routine blood investigations were normal. On upper gastrointestinal endoscopy, oesophagus was dilated with resistance to the passage of the endoscope across LOS (figure 1). Based on these endoscopic findings, there was suspicion of oesophageal motility disorder. Oesophageal manometry was done to confirm the oesophageal motility disturbance. High-resolution oesophageal manometry showed panoesophageal pressurisation with raised LOS median integrated relaxation pressure consistent with type II achalasia cardia (figure 2). On the basis of lymphadenopathy and rapid progression of dysphagia, patient was further evaluated to rule out paraneoplastic dysphagia. Cervical

Figure 1 Endoscopic image showing dilated oesophagus.

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Figure 3 PET CT image showing FDG avid lymphadenopathy.

lymph node biopsy revealed diffuse large B cell non-Hodgkin's lymphoma.

PET CECT (Contrast Enhanced Computerised Tomography) revealed FDG (Fluoro-Deoxy Glucose) avid cervical and mediastinal lymph nodes (figure 3).

He was started on R-CHOP therapy for NHL. After six cycles of chemotherapy, his dysphagia improved, and subsequent gastrograffin study showed free passage of contrast across LOS, and oesophageal High Resolution Manometry (HRM) showed normal oesophageal manometry indicating resolution of achalasia.

INVESTIGATIONS

1. Upper gastrointestinal endoscopy: dilated oesophagus with resistance to passage of endoscope across the LOS.
2. Oesophageal high-resolution manometry: panoesophageal pressurisation with raised median IRP across LOS (type II achalasia).
3. PET CECT whole body: FDG avid cervical and mediastinal lymphadenopathy.
4. Lymph node biopsy: diffuse large B cell lymphoma.

DIFFERENTIAL DIAGNOSIS

Primary achalasia: usually long duration of symptoms and seen in elderly age group.

TREATMENT

Patient was transferred to medical oncology and started on R-CHOP therapy.

OUTCOME AND FOLLOW-UP

After six cycles of R-CHOP, patient condition improved with significant reduction in lymph node size and substantial relief of dysphagia obviating need for endoscopic intervention.

DISCUSSION

Secondary achalasia is an uncommon entity. In a large series of 2000 patients, secondary achalasia was reported in 1.5% of cases.⁴ It is difficult to distinguish between primary and secondary achalasia but the age of onset is younger, and duration of symptoms is longer in the former compared with the latter. There are some endoscopic features such as mucosal ulcerations, nodularity, submucosal or extrinsic bulge, which

Learning points

- Oesophageal high-resolution manometry is the gold standard to evaluate cases of dysphagia with suspected oesophageal motility disorder.
- Pseudoachalasia should be suspected if dysphagia is rapidly progressive.
- In a patient of achalasia, physical examination is of paramount importance; any neck mass or palpable lymph node may point towards underlying malignancy with associated pseudoachalasia.

are more common in secondary achalasia.^{5–7} Achalasia can occur secondary to carcinoma of gastric cardia, lower oesophagus, as paraneoplastic manifestation of malignancy such as small cell carcinoma lung, and lymphoma.^{2,8} Pastor *et al*⁹ reported an interesting case of pseudoachalasia associated with retroperitoneal diffuse large B cell lymphoma. Another case of pseudoachalasia was reported in a paediatric patient with Hodgkin's lymphoma.¹⁰ Gockel *et al*¹¹ in a case series on pseudoachalasia reported compression of lower esophagus by mass as one of the suggested mechanisms of dysphagia in cases of pseudoachalasia. Another factor that have been suggested is the infiltration of the myenteric plexus by the malignant cells. Liu *et al*¹² in a clinicopathological case series on pseudoachalasia showed that on histopathological examination of oesophagus, there was infiltration of myenteric plexus by malignant cells with reduced number of the ganglion cells. In our case, the infiltration of oesophageal myenteric plexus by lymphoma cells might explain initial dysphagia that responded to chemotherapy, but this was not confirmed histologically.

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