Case report: Achalasia-like dysmotility secondary to oesophageal involvement of sarcoidosis

Albert J Bredenoord,1 Jafar Jafari,2 Saleheen Kadri,1 David E Simcock,3 Daniel Sifrim,2 Sean L Preston1

ABSTRACT

We present a case of a patient with sarcoidosis and who subsequently developed dysphagia for solids, and some difficulty in swallowing liquids. High-resolution manometry of the oesophagus showed absent peristalsis in the oesophageal body and incomplete relaxation of the lower oesophageal sphincter. The diagnosis of sarcoidosis with oesophageal involvement was made and treatment with prednisolone 30 mg OD initiated. The patient improved symptomatically and high-resolution manometry was repeated showing complete recovery of oesophageal peristalsis and a deeper relaxation of the lower oesophageal sphincter. This is thus the first description of high-resolution manometry in sarcoidosis-induced changes of the oesophagus and of the effect treatment has on these motility changes. Oesophageal involvement of sarcoidosis is extremely rare and only a few cases have been reported. The symptoms and manometric pattern of this disorder mimics that of achalasia. However, we show that treatment with prednisolone results in a completely disappearance of the symptoms of dysphagia and subsequently lead to a large improvement of oesophageal motility.

CASE HISTORY

A 29-year-old man was referred to the Gastroenterology Clinic with symptoms of dysphagia. He was just previously diagnosed with a lateral medullary syndrome due to sarcoid vasculitis on enhanced MRI scanning in the absence of occlusive vascular disease which was suspected to be secondary to sarcoidosis. A CT scan of the chest had been performed because of chest discomfort and dyspnoea and revealed marked mediastinal and hilar lymphadenopathy, with nodular lung parenchymal infiltrates suggestive of stage II sarcoid, as well as splenomegaly. His serum ACE was 151 IU/l with a CRP of less than 5 mg/l. Bronchial biopsies had shown non-necrotising granulomas; both Ziehl Nielsen and PAS staining of the biopsies were negative for tuberculosis. Spirometry was supersupnormal, but CO gas transfer was reduced at 73%, predicted normal at presentation. He was taking both dipyridamole and aspirin for secondary stroke prevention.

The patient suffered from significant dysphagia for solids and some difficulty in swallowing liquids. There was no history of heartburn, regurgitation or chest pain and no nausea and vomiting. He lost approximately 5 kg in weight. Physical examinations of the abdomen and neck were unremarkable.

Upper endoscopy revealed that the oesophagus was partly filled with food. The lower oesophageal sphincter (LOS) was tightly contracted; however, the stomach could easily be reached. Erythematous gastritis of the whole stomach was noted. Oesophageal biopsies showed mild chronic inflammation but no granulomata, while gastric biopsies revealed non-caseating granulomata within the lamina propria and moderate chronic inactive inflammation. A barium oesophagogram showed a slightly dilated proximal oesophagus with barium hold-up in the distal oesophagus (figure 1).

High-resolution manometry of the oesophagus was performed and revealed complete lack of peristalsis in the oesophageal body and a normotensive LOS (29.9 mm Hg) that showed incomplete relaxation after swallowing (residual pressure of 78%, integrative relaxation pressure (IRP) 28.9 mm Hg) (figure 2A). The diagnosis of pulmonary sarcoidosis with vasculitis and oesophageal involvement was made and treatment with prednisolone 30 mg OD was started.

His respiratory symptoms improved soon after the start of treatment. One year after diagnosis, the patient reported that he has become completely asymptomatic and had no difficulties with swallowing anymore. Pulmonary gas transfer improved to normal range and serum ACE at that time dropped to 17 IU/l. High-resolution manometry was repeated and revealed complete recovery of oesophageal peristalsis (100%) and significant improvement in LOS relaxation (residual pressure 67%, IRP 21.0 mm Hg) (figure 2B). A slightly higher than normal intrabolus pressure (IBP) was observed (21.0 mm Hg) illustrating the resulting incomplete LOS relaxation, however, this was not accompanied by persistent symptoms of dysphagia.

DISCUSSION

While gastrointestinal involvement of sarcoidosis is seen very infrequently, oesophageal involvement of sarcoidosis is extremely rare. A study of 160 patients with sarcoidosis showed only one case with gastrointestinal involvement and no cases of oesophageal involvement, while a review of 1254 cases with sarcoidosis revealed two cases with gastrointestinal involvement and no cases of oesophageal involvement.1 2

The first case of oesophageal sarcoidosis was reported by Kerly and less than 20 cases have been published since.3–6 Oesophageal involvement of sarcoidosis usually presents with dysphagia. There are different ways sarcoidosis can result in swallowing difficulties. Direct involvement of the myenteric plexus can result in a pattern similar to achalasia, as was seen in our patient.
Dufresne described a case of achalasia secondary to sarcoid involvement of the oesophagus in which a surgical myotomy is performed with good result. Surgical biopsies of the muscle layers revealed abnormal myenteric plexus with inflammatory infiltration and complete demyelinization and about 50% axon loss with evidence of active degeneration. No granulomas were found in the oesophagus.

Other mechanisms include myopathy, caused by infiltration of the skeletal muscle of the cricopharyngeal and proximal oesophageal wall muscles. Development of strictures secondary to sarcoid involvement of the oesophagus have also been described. Dysphagia can also be related to extrinsic compression on the oesophageal body caused by enlarged lymph nodes.

In the majority of patients with oesophageal sarcoidosis, an achalasia-like pattern is recognised with absent peristalsis of the oesophagus and insufficient relaxation of the LOS. Our case illustrates that the symptom of dysphagia is clearly correlated to the manometric abnormalities, and both symptoms and oesophageal motility subsequently improved gradually under prednisolone treatment. This is the first report of high-resolution manometry recording of these changes and, as demonstrated, high-resolution manometry can be used to follow-up the effect of treatment and is able to detect the subtle remaining abnormalities afterwards.

The high-resolution manometry after treatment still showed an elevated IBP suggesting a remaining functional oesophago-gastric junction obstruction. Given his symptomatic relief, however, we decided to opt for a conservative approach for now but we will repeat oesophageal function tests in future to follow-up on this manometric abnormality.

As described, surgery has been suggested for oesophageal sarcoidosis, however, since we show that the achalasia-like motility pattern in our patient is almost completely reversed, we think that medical treatment is certainly preferable. Surgery would only be advisable for those in which medical treatment has failed.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; not externally peer reviewed.
REFERENCES
1. Israel HL, Sones SM. Sarcoidosis; clinical observation on one hundred sixty cases. Arch Intern Med 1958;102:766—76.

Editor’s quiz: GI snapshot

Yet another case of haemochromatosis?

CLINICAL PRESENTATION
A 38 year old Caucasian woman whose father and two brothers had been diagnosed with hereditary haemochromatosis was found to have chronic fatigue and a serum ferritin of 1490 µg/L. Her transferrin saturation was 32% and her full blood count, liver biochemistry, international normalised ratio and C-reactive protein were all normal. She had regular menstrual periods and no other medical history of note. She had no specific risk factors for chronic liver disease, her alcohol consumption was less than 5 units a week and there was no history of receipt of blood transfusion. Clinical examination and abdominal ultrasound scan were unremarkable. HFE genotyping was normal. A liver biopsy was performed, the histology of which is shown in figure 1.

QUESTION
What is the diagnosis?

See page 188 for the answer

A J Fowell, 1 A C Bateman, 2 W J Griffiths, 3 K L Nash 1
1Department of Hepatology, Southampton University Hospitals NHS Trust, Southampton, UK; 2Department of Cellular Pathology, Southampton University Hospital’s NHS Trust, Southampton, UK; 3Department of Hepatology, Cambridge University Hospitals NHS Foundation Trust, Cambridge, UK

Correspondence to Dr Andrew J Fowell, Southampton University Hospitals NHS Trust Level E South Block (MP811), Southampton General Hospital, Tremona Road, Southampton SO16 6YD, UK; afowell@hotmail.com

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; not externally peer reviewed.

Published Online First 24 September 2010

Gut 2011;60:155; doi:10.1136/gut.2010.221499

Figure 1 Histological appearances of the patient’s liver biopsy. (A) H&E stain. Golden brown haemosiderin pigment accumulation is present within phagocytic (Kupffer) cells in the hepatic sinuses (some but not all of these cells are arrowed). (B) Perl’s stain. The iron accumulation is highlighted. Both panels are at a magnification of ×400.
Case report: Achalasia-like dysmotility secondary to oesophageal involvement of sarcoidosis


Gut 2011 60: 153-155 originally published online November 4, 2010
doi: 10.1136/gut.2010.227868

Updated information and services can be found at:
http://gut.bmj.com/content/60/2/153.full.html

These include:

References
This article cites 9 articles, 3 of which can be accessed free at:
http://gut.bmj.com/content/60/2/153.full.html#ref-list-1

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/