CASE REPORT

Fever, ulcers and joint pain: an incidental finding of oesophageal dysmotility

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SUMMARY

A 41-year-old Bangladeshi man presented with a 2-month history of bilateral feet/hand swelling which intermittently resolved without medication. All blood tests performed by the general practitioner (GP) were unremarkable. Following admission to the accident and emergency department, a chest X-ray revealed a 'vail-like' opacification in the right side. High-resolution CT confirmed dilation of oesophagus with food debris and circumferential thickening of the lower oesophageal sphincter.

BACKGROUND

Rare presentation found incidentally.

CASE PRESENTATION

A 41-year-old Bangladeshi man presented with a 2-month history of fever and bilateral feet swelling to his general practitioner (GP). Swelling was observed overnight and the patient reported a dull pain around the metatarsal joints which was increased with analgesia. He previously visited Bangladesh 6 months prior to the onset of his symptoms and reported associated morning joint stiffness, dry mouth and increased thirst. He denied any history of unprotected sexual contact, dysuria, skin rash/changes and psoriasis or connective tissue disorders.

The patient reported that these symptoms had been investigated by his GP and all the tests were normal. Twelve days after these investigations the swelling had improved without any medication. Two days later he noted bilateral hand swelling over night, occasional night sweats and polyarthropathy.

On examination the patient was comfortable at rest. There was tenderness and swelling noted bilaterally in the second and third proximal intrapharyngeal joints. In addition, there was swelling and tenderness of wrists as well as elbows. No rashes were noted and joints were cool to touch.

Tenderness was also noted in heels as well as the patient demonstrated good range of movement in all limbs. Bilateral mild pitting oedema was noted.

Mouth ulcers were also noted at the base of the lower gum line. However, cardiovascular, abdominal and neurological examinations were unremarkable. No stigmata of endocarditis.

Respiratory examinations showed the trachea was central with equal lung expansion, resonant on percussion and bilateral air entry noted. No crepitations were noted.

INVESTIGATIONS

Blood results showed haemoglobin 10.5, white cell count 14.6, platelets 283, neutrophils 11.2, monocytes 1.8, erythrocyte sedimentation rate 6 mL, Na 134, K 3.8, Cl 98, urea 2.8, creatine 52, epidermal growth factor receptor 170 mL/min, C reactive protein 71 mg/L, ACE 28 and urate 248 μ mol/L. Complement C3 and C4 were 1.32 and 0.30 g/L (0.14–0.54 g/L), respectively. Antistreptolysin O titre (77). Rheumatoid factor <15 (0–20). Antibodies against the following were investigated: antineutrophil antibodies, IgG antineutrophil cytoplasmic antibodies, rheumatoid factor (<15), anticyclic citrullinated peptide (1.0 k units/L), gastric parietal cell, smooth muscle IgG, mitochondrial IgG, liver/kidney/microsomal, extractable nucleic antibodies, double-stranded DNA (dsDNA) ELISA, dsDNA immunofluorescence, HIV1 and 2 antibodies, hepatitis C virus IgG and hepatitis B surface antigen were undetectable.

A chest X-ray (CXR) was performed and was markedly abnormal with opacification with mottling throughout the entire right lung field (figure 1). X-ray of the hands and feet were unremarkable.

High-resolution CT was requested to investigate this further that showed increased opacification in the right thorax which was explained by a markedly dilated oesophageal secondary to circumferential thickening of the gastro-oesophageal junction containing food debris (figures 2–4).

DIFFERENTIAL DIAGNOSIS

Of CXR

- ▶ Tuberculosis
- ▶ Pneumothorax

Figure 1 Markedly abnormal chest X-ray demonstrating lung opacification of entire right side.

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Figure 2 Coronal slice of CT thorax.

- ▶ Atypical pneumonia
- ▶ Aspergilosis

OUTCOME AND FOLLOW-UP

The patient was seen in rheumatology and gastroenterology outpatient clinics. All investigations including autoimmune blood tests and a gastroscopy were found to be normal.

DISCUSSION

Achalasia is a Greek word that means 'failure of relaxation'.¹ The condition is characterised by aperistalsis of the oesophageal body and lack of relaxation of the lower oesophageal sphincter (LOS).¹ Presenting features include dysphagia, odynophagia, heart burn, weight loss and polyarthropathy.¹⁻³

Achalasia can be primary (idiopathic) or secondary. The exact pathogenesis of primary achalasia is still not known.¹ However, it is thought to be caused by the loss of inhibitory ganglion cells in the myenteric plexus.¹ In secondary achalasia, several studies have attempted to explore initiating agents that may cause the disease such as viral infection (measles, varicella zoster virus),

Figure 3 Sagittal slice of CT thorax.

Figure 4 CT thorax illustrating dilated oesophagus secondary to obstruction from marked circumferential 'mass-like' thickening of the gastro-oesophageal junction.

chagas disease, other environmental factors, autoimmunity and genetic factors.²⁻⁴

Investigations would include

1. Barium swallow and CXR^{1 5}
 - A. Lack of the progressive peristalsis
 - B. Delay in barium passing into the stomach
 - C. CXR may show a wide gullet
2. Endoscopy^{1 5)}
 - A. Food debris may be visible
3. Manometry^{1 5}
 - A. Small pressure sensitive tube (similar to NG tube)
 - B. Detects weak contractions of the upper gullet and sustained high pressure (spasm) of the LOS
 - C. High pressure and the failure of the LOS to relax→'balloon effect' which causes difficulty in swallowing and food remaining in the gullet.

Treatment options include:

1. Drugs^{1 5}
 - A. Nitrates or nifedipine will only produce a brief improvement in swallowing and are ineffective in some
 - B. S/E headache, which usually gets less with continued treatment
 - C. Dilation (stretching the valve)
2. Botulinum toxin ('Botox') injection^{1 5}
 - A. It is not a permanent treatment, but is useful for patients who are unable or unwilling to have surgery
3. Surgery^{1 5}
 - A. Muscle fibres that fail to relax are cut and this results in a permanent improvement in swallowing
 - B. The operation is now often performed by laparoscopy ('key-hole surgery') requiring only an overnight stay in hospital.

Previous studies have implicated antimyenteric and antimuscarinic autoantibody and found to have a higher prevalence of these autoantibodies in achalasia patients compared to controls.^{5 6} Recent attempts in Canada have been made to determine the prevalence of autoimmune disease among patients with achalasia.⁷ The authors highlighted four autoimmune disease as having a higher prevalence in the study group (n=94) of achalasia patients compared to the general population (Canada).⁷

- ▶ Type I diabetes mellitus (RR 11, 95% CI 3.0 to 39)
- ▶ Hypothyroidism (RR 5.4, 95% CI 2.1 to 13)
- ▶ Systemic lupus erythematosus (RR 44, 95% CI 2.4 to 241)
- ▶ Uveitis (RR 532, 95% CI 27 to 2891)

In conclusion, all autoimmune investigations performed as an outpatient have been negative. Interestingly, recent literature suggests younger patients with achalasia are 3.3 times more likely to suffer from any autoimmune condition (95% CI 1.8 to 5.7).⁷ Recent links with severe achalasia and arthritis may explain our patient's symptoms.⁸ Erikson *et al*⁸ reported three patients with severe achalasia and arthritis reported improvements in oesophageal and arthritic symptoms following oesophagectomy and oesophageal reconstruction. In addition, oesophageal involvement of sarcoidosis has been suggested to induce 'achalasia-like' dysmotility as symptomatic improvement, complete recovery of oesophageal peristalsis and a deeper relaxation of the LOS, was achieved following treatment with prednisolone 30 mg orally once daily for 3 weeks.⁹

Learning points

- ▶ Achalasia is a rare condition^{1 5} and even rarer to be picked up incidentally with minimal gastric symptoms.
- ▶ Demonstrated by lack of the progressive peristalsis and delay in barium passing into the stomach→'bird beak' sign may be noted.^{1 5}
- ▶ There is limited evidence regarding links with achalasia and autoimmune diseases—more research is required.^{1 5}

Contributors CAI drafted the case report. NT and AM proof read and edited the article.

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