# X-ray quiz: a middle-aged female presenting with chest pain and cough

# X 光照片猜謎：一名有胸痛及咳嗽的中年女士

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## Case

A 46-year-old female had chest pain and cough for a few weeks. Her past medical history was unremarkable. She had no history of operation. She attended the Accident and Emergency Department and a postero-anterior erect chest X-ray (CXR) was performed (Figure 1).

Figure 1.

## Questions

1. What are the radiological findings?
2. What are your differential diagnoses?
3. What other investigations would be helpful?

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## Answers

1. The frontal CXR shows a tubular structure with a convex lateral border along the right mediastinal region. The upper part exhibits air lucency (Figure 1). The lower part is radiopaque, as confirmed by a lateral CXR (Figure 2). The normal gastric bubble cannot be delineated, and only a colonic shadow is seen under the left hemi-diaphragm. The most likely cause is a dilated oesophagus containing food debris. Both lung fields are clear. There are no soft tissue calcifications or surgical clips suggestive of previous operation.
2. The differential diagnoses include achalasia, connective tissue disease (CREST syndrome), tumour at the gastroesophageal junction or gastric cardia, peptic stricture, and post-vagotomy stasis. For elderly patients, presbyesophagus is a possibility. Consider Chagas disease for foreigners and for patients with a travel history to South America.
3. Other investigations include an upper gastrointestinal contrast study (barium swallow and meal), computed tomogram and/or oesophago-gastro-duodenoscopy (OGD).

Figure 2. Lateral chest X-ray showing a dilated oesophagus. The trachea is displaced anteriorly.

## Discussion

Achalasia is a well-known oesophageal motility disorder characterised by the absence of purposeful peristaltic waves with incomplete or failed relaxation of the lower oesophageal sphincter. It is caused by degeneration of the neurons of Auerbach's plexus which lies between the longitudinal and circular muscle coats. Both primary and secondary peristalsis fail, and tertiary contractions may develop.[1] This disorder generally occurs in the 35-50 year age group.[1] No known cause has been identified.[2] The patients usually present with dysphagia for both solids and liquids. Sometimes, they may complain of chest pain, which can be severe.

The CXR may reveal a right convex opacity behind the right cardiac border, as in Figure 1. This may be tethered by the azygos arch, resulting in greater dilatation above and below. An air-fluid level or a mottled lucency may be seen and is due to residual food residues. Pneumomediastinum can appear similar but the lateral border should be paper-thin. The gastric bubble can be small or even absent. As these patients have dysphagia, they can aspirate easily. Patchy alveolar opacities may be detected at dependant positions, usually the lower lobes of the lungs. Anterior displacement and bowing of the trachea can be seen in lateral CXR (Figure 2).[3]

Barium upper gastrointestinal studies will show the absence of primary peristalsis below the level of the cricopharyngeus.[3] Sometimes, there are tertiary contractions in the non-dilated distal oesophagus of early achalasia (vigorous achalasia).[3,4] Bird-beak or rat-tail deformity - smooth tapered symmetrical narrowing of the distal oesophagus caused by failed relaxation of the lower oesophageal sphincter - is characteristic (Figure 3).[5] There is a mega-oesophagus, with

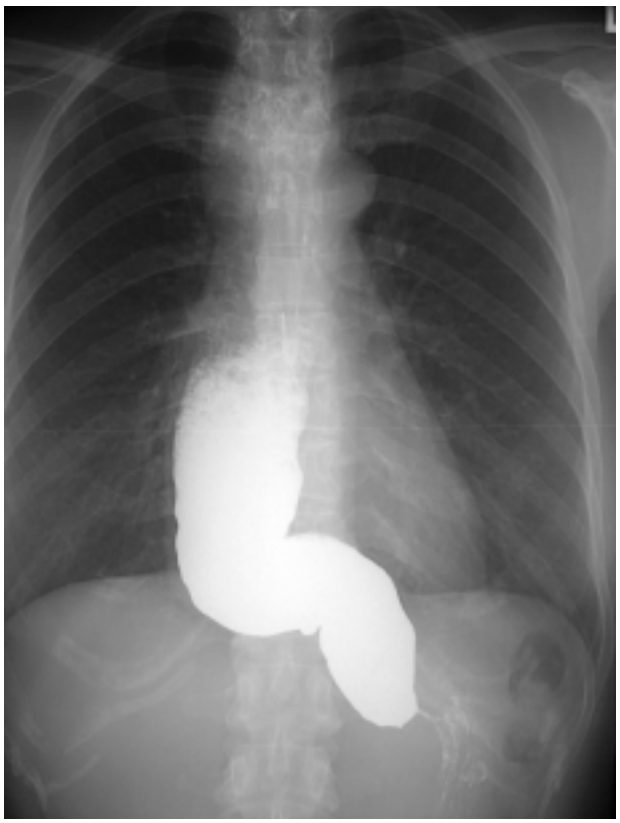

Figure 3. Barium swallow study demonstrating a dilated oesophagus down to the gastroesophageal junction where it adopts a bird-beak or rat-tail appearance. This is characteristic of achalasia.

dilatation beginning in the upper one third and ultimately the entire length.

Attention should be paid to the regions of the gastroesophageal junction and gastric cardia to exclude an underlying tumour. Connective tissue disease such as scleroderma with oesophageal involvement can mimic achalasia but the former has a patulous lower oesophageal sphincter. Soft tissue calcifications and bony changes shown in X-rays of other regions together with the clinical history also help establishing the diagnosis of scleroderma. In the case of peptic stricture, there are reflux and ulceration at and around the gastroesophageal junction. For elderly patients, presbyeosphagus has to be considered. There is defect in primary peristalsis and tertiary contractions can also be detected. Other mimickers include patients with diabetes mellitus and bulbar palsy.

Computed tomography (CT) and OGD are helpful in case the barium studies fail to evaluate the cardia and fundal region. In achalasia, CT reveals a dilated oesophagus with little or no wall thickening, and no mass at the cardia, fundus or gastroesophageal region. If there is asymmetric thickening of the distal oesophageal wall, a soft tissue mass at the cardia, or mediastinal adenopathy, neoplasia has to be excluded. In our case, CT was not performed but OGD was done and it revealed no evidence of neoplasia.

The complications of achalasia include chronic aspiration which may lead to the development of bronchiectasis, lung abscess, empyema or fibrosis. Prolonged stasis of food may cause oesophagitis with an increased risk of squamous cell carcinoma (2-7%).[3] This usually develops in the mid-oesophagus and may be obscured during the barium study. Endoscopy is helpful.[4]

Treatment includes balloon dilatation of the gastroesophageal junction or myotomy of the sphincter (Heller's operation).[3]

## References

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