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Answer: Achalasia of the oesophagus

The barium swallow showed dilated oesophagus with a typical 'Bird beak' appearance of the distal oesophagus. Other features that may be observed in barium swallow study is the either lack or presence of synchronous oesophageal contractions. In advanced cases, a plain chest radiography may show a widened mediastinum from a dilated oesophagus with or without chronic lung changes. A fluid level may be seen.

Achalasia, also referred to as cardio-spasm and oesophageal aperistalsis, is a motility disorder that leads to the failure of the smooth muscle of the lower oesophageal sphincter (LES) to relax.¹⁻³ The condition is characterised by increased LES tone, incomplete LES relaxation, and lack of peristalsis of the lower oesophagus.¹⁻³ This leads to accumulations of swallowed foods resulting in symptoms and eventual dilation of the oesophagus.

The underlying pathogenesis is due to failure of the distal oesophagus inhibitory neurons. In certain parts of the world such as South America, it is important to exclude Chagas disease, an infection that leads to the same manifestation. For elderly patients, it is also important to rule out underlying malignancy infiltrating into the lower oesophagus, causing pseudo-achalasia.

The estimated incidence is 1/100,000. Symptoms associated with achalasia of the oesophagus include; discomfort in the lower chest, dysphagia and regurgitation. Weight loss commonly follows. Recurrent chest infection may occur from recurrent aspiration. Often patients will have been treated for gastro-oesophageal reflux disease before being diagnosed with achalasia.

Although such features described are specific for achalasia of the oesophagus, the gold standard for diagnosis is manometry study. Endoscopy and barium swallow study may not show the typical features in the early stages before dilatation of the oesophagus occurs. Therefore, the diagnosis can be missed in the early part of the disease unless manometry is performed, either the standard manometry or high resolution manometry. The typical findings of manometry for achalasia is shown in the Supplementary Text.

The management of achalasia is aimed at reducing the LES tone. The current gold standard is Heller's myotomy (open or laparoscopic) with formation of a wrap, either partial or complete. Endoscopic balloon dilatation also produces very good results but is associated with a slightly higher risk of recurrence. Another option is botulinum (botox) injection usually 25 units in four quadrants at the LES. This is typically reserved for those (i.e. elderly) not suited for surgery or balloon dilatation.

REFERENCES

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