Dysphagia in a patient with Addison's disease

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DESCRIPTION

A 19-year-old boy presented with history of progressive dysphagia and vomiting of 3 years duration. Dysphagia was more for liquids than solids. At the age of 8 he had vomiting and asthenia, and was noticed to have hyperpigmentation. At that time he was diagnosed with adrenal insufficiency and had been on glucocorticoid and mineralocorticoid replacement ever since. His mother had also noted that he had an absence of tears since early childhood for which he was evaluated and diagnosed with alacrimia and given tear substitutes. He had no significant family history of similar illness. Examination revealed generalised hyperpigmentation and a corneal ulcer in the left eye. His pubertal status was normal and rest of the examination was unremarkable. Barium swallow (figure 1) showed a dilated thoracic oesophagus above the lower oesophageal sphincter and bird beak appearance. A grossly dilated oesophagus with sigmoid appearance with mucosal breaks of 7–8 mm in the lower oesophagus and no other structural abnormality was visualised on an upper gastrointestinal endoscopy. A further evaluation by oesophageal manometry (figure 2) displayed a high median (integrated relaxation pressure >17 mm/Hg) with an absence of peristalsis confirming a diagnosis of achalasia cardia. A diagnosis of Allgrove syndrome (Triple A syndrome) was carried out as this patient had a triad of adrenal insufficiency, achalasia and alacrimia.

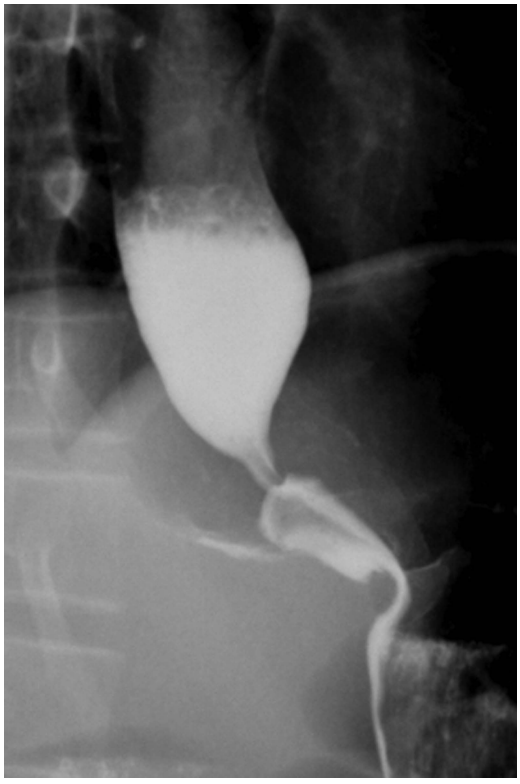

Figure 1 Barium swallow.

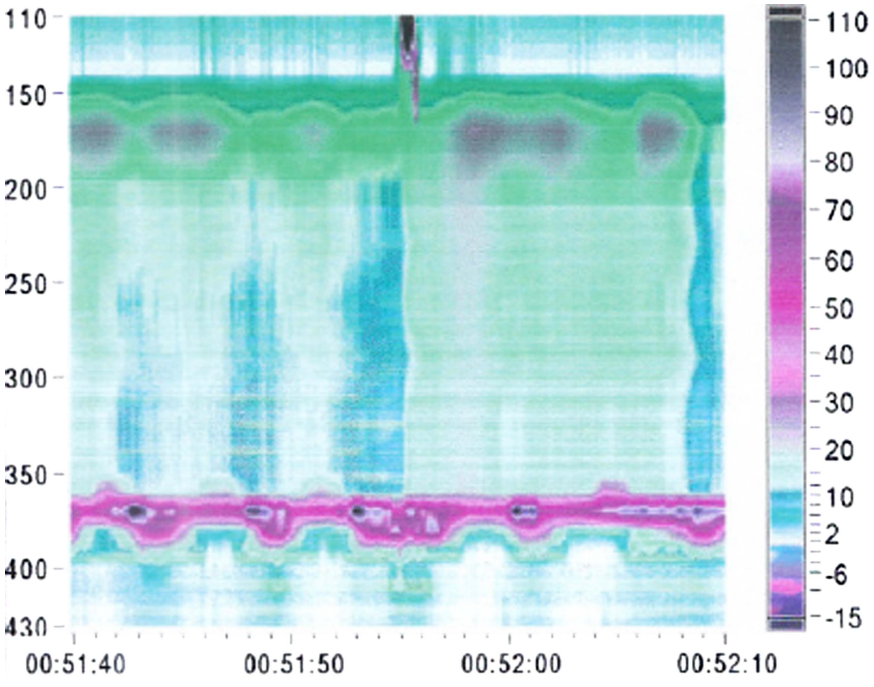

Figure 2 Oesophageal manometry.

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Learning points

- Dysphagia in a patient with Addison's disease could be due to oesophageal candidiasis as a part of polyglandular autoimmune syndrome I or achalasia cardia as a part of Triple A syndrome.
- Allgrove syndrome is a rare disorder due to mutation in AAAS gene encoding ALADIN protein on 12q13 locus and is an autosomal recessive disorder with variable presentation.[1 2]
- The primary cause of mortality in these patients is an adrenal crisis; however, an increased morbidity is seen due to corneal ulceration or oesophageal involvement.

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Competing interests None.

Patient consent Obtained.

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2 Cronshaw JM, Matunis MJ. The nuclear pore complex protein ALADIN is mislocalized in triple A syndrome. *Proc Natl Acad Sci USA* 2003;100:5823–7.