

# Dysphagia in a patient with Addison's disease

Nitin Kapoor, Sahana Shetty, Shrinath Shetty, Thomas Vizhalil Paul

Christian Medical College,  
Vellore, Tamil Nadu, India

**Correspondence to**  
Professor Thomas Vizhalil Paul,  
thomasvpaul@yahoo.com

Accepted 15 June 2014

## 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.

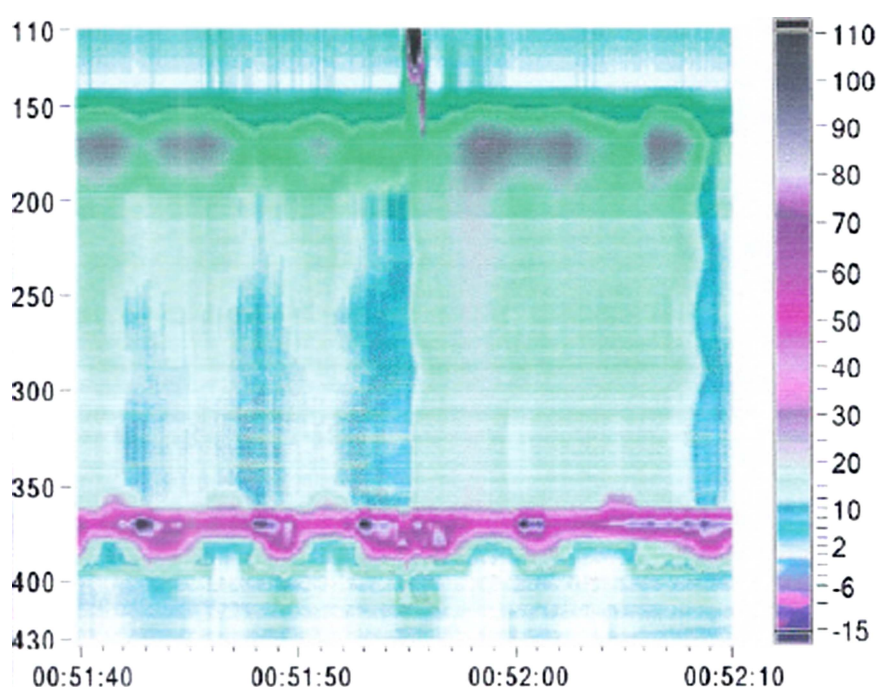


Figure 2 Oesophageal manometry.



CrossMark

**To cite:** Kapoor N, Shetty S, Shetty S, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-203672

## 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.<sup>1 2</sup>
- ▶ 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.

**Contributors** NK, SAH and SRI wrote the manuscript. NK and SAH edited the manuscript. TVP reviewed and edited the manuscript. NK, SAH, SRI and TVP approved the final version.

**Competing interests** None.

**Patient consent** Obtained.

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

## REFERENCES

- 1 Allgrove J, Clayden GS, Grant DB. Familial glucocorticoid deficiency with achalasia of the cardia and deficient tear production. *Lancet* 1978;1:1284–6.
- 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.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.  
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact [consortiasales@bmjgroup.com](mailto:consortiasales@bmjgroup.com)

Visit [casereports.bmj.com](http://casereports.bmj.com) for more articles like this and to become a Fellow