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CORRESPONDENCE

A Rare Case of Allgrove Syndrome

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To the Editor: Allgrove syndrome is a rare autosomal recessive condition with fewer than 100 cases reported, characterised by three cardinal features -achalasia, alacrimia and ACTH resistant adrenal insufficiency [1]. We describe the first case from Coastal Karnataka.

A 17-y-old boy came for evaluation of dysphagia to both solids and liquids, weight loss and weakness for two years. Patient developed generalised hyperpigmentation over the past four years. Parents noted patient crying without tear production since childhood.

On examination, there was generalised hyperpigmentation including lips, palms, soles and buccal mucosa. His height was 163 cm, weight was 35 kg and BMI was 13.2 kg/m^2. Blood pressure was 100/60 mmHg with no postural drop. Schirmer's test was positive showing evidence of dry eyes due to alacrimia. The patient's 8 AM serum Cortisol was 1.35 mcg/dl and ACTH was 478 pg/ml. Barium swallow revealed features suggestive of achalasia cardia as shown in Supplementary Fig. S1, which was confirmed by esophageal manometry and upper GI endoscopy.

Patient underwent Heller's Cardiomyotomy under steroid coverage. Postoperatively he was started on a maintenance dose of oral hydrocortisone 15 mg/d. Improvement in symptoms, reduction in hyperpigmentation and a weight gain of 3.5 kg were noted on two-month follow-up.

Allgrove syndrome is an autosomal recessive disorder often diagnosed late in patients as few present with all three cardinal features. Our patient presented with only alacrimia in his first decade, whereas signs and symptoms of achalasia and adrenal failure only developed in his second decade leading to diagnosis. Although usually seen in the first decade of life, late presentation of adrenal insufficiency here, could be due to gradually developing adrenal destruction [2].

Allgrove syndrome is often misdiagnosed due to the variable presentation. Hence this condition should be suspected in the presence of only one or two cardinal features.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s12098-023-04840-1.

Declarations

Conflict of Interest None.

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