Addison's disease presenting with hyperpigmentation

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DESCRIPTION

Our patient is a 60-year-old woman of Indian ethnicity with background of diabetes mellitus for 10 years. She presented with persistent vomiting, giddiness and significant weight loss for 1 month. Of note, she had recent hypoglycaemia necessitating gradual reduction in doses of glucose-lowering medications. Blood investigations revealed hyponatraemia and relative hyperkalaemia. Morning cortisol was <13.8 nmol/L with adrenocorticotrophic hormone (ACTH) >278 pmol/L. On specific further questioning, she realised that for the past few months, her skin had become significantly darker than before (figure 1). A CT scan of her adrenal glands was normal. Pancreatic autoantibodies, which were sent because she has diabetes mellitus, were negative. An anti-adrenal autoantibody screen is not available in our laboratory. She was treated for Addison's disease with hydrocortisone replacement. After replacement, her symptoms improved with reduction in her skin pigmentation (figure 2). ACTH level reduced to 9.2 pmol/L.

In Addison's disease, there is adrenalitis leading to cortisol deficiency. The resulting negative feedback to the hypothalamic pituitary adrenal

Figure 2 Resolution of skin hyperpigmentation after glucocorticoid replacement.

Figure 1 Hyperpigmentation prior to treatment.

axis leads to increased proopiomelanocortin, which is a prohormone that is cleaved into ACTH and melanocyte-stimulating hormone. This leads to melanin synthesis by epidermal melanocytes, which causes increased skin pigmentation.¹ In patients with darker skin tones, hyperpigmentation may be missed. With glucocorticoid replacement, ACTH levels decrease and hyperpigmentation improves.

Patient's perspective

I did not realise that the darkening of my skin had anything to do with my medical condition. I feel much better after the treatment was started.

Learning points

- ▶ Primary adrenal insufficiency caused by Addison's disease may present with skin hyperpigmentation.
- ▶ Hyperpigmentation in patients with darker skin tones may be missed.
- ▶ Adequate replacement of glucocorticoid in Addison's disease improves hyperpigmentation.

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