A case of Pancytopenia due to isolated ACTH deficiency successfully treated with Hydrocortisone

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Background

Pancytopenia due to hypopituitarism has been reported previously in the literature. Most of these case reports relate to hypopituitarism secondary to Sheehan's syndrome. We report a case of isolated ACTH deficiency causing pancytopenia which was successfully treated with Hydrocortisone replacement.

Case history

A 71yr old lady with a known history of treated primary hypothyroidism and learning disabilities was referred to Acute Medical Unit with urinary frequency, back pain and increased confusion. Urinary tract infection (UTI) was confirmed with positive urine culture for E.coli. Her diuretics were stopped, antibiotics started and despite IV fluid resuscitation, she remained hypotensive with hyponatraemia (Na 120) and acute kidney injury (Urea 6.8 Creat 169). Her synacthen test confirmed cortisol deficiency with 0 min Cortisol 111nmol/L, 30 min cortisol 291nmol/L and 60 mins 323nmol/L. She was started on

Hydrocortisone replacement with good clinical improvement.

She was pancytopenic with WBC 2.1 Hb 98 g/L PLTS 53. Further investigations for her anaemia and pancytopenia failed to reveal any obvious cause. The Reticulocyte count was normal and HIV, Hepatitis serologies were negative.

Direct Antiglobulin test was positive, Urine Bence-Jones protein was negative. Bone marrow aspirate showed increased cellularity but no features of Myelodysplasia. Her ACTH was undetectable < 5 . Other anterior pituitary hormones showed Prolactin 77mu/L, IGF 26ug/L, LH 34.2iu/L, FSH 86.7iu/L. TSH was 0.20mu/L and Free T4 20.1pmol/L on 100mcg of Thyroxine.

The adrenal antibodies were negative and pituitary MRI was normal. A diagnosis of Isolated ACTH deficiency was made and she was discharged with outpatient Endocrine and Haematology follow-up. Within 2 months of Hydrocortisone replacement, all her counts improved with Hb concentration 113 g/L, WBC 5.7, PLTS 248 and remained stable.

Short synacthen test

	0	30	60
	mins	mins	mins
Cortisol nmol/L	111	291	323

Full Blood count

	On admission 16/11/2011	5 months after HC treatment 03/04/2012
Hb	98	122
WBC	2.1	9
PLTS	53	220
Neut	1.5	5.8
Lymph	0.4	1.5

Discussion

Isolated ACTH deficiency is a rare disorder characterised by secondary adrenal insufficiency with low or absent cortisol production, normal secretion of pituitary hormones (other than ACTH) and absence of structural abnormalities of pituitary gland. To our knowledge this is the first reported case of isolated ACTH deficiency causing pancytopenia successfully treated with steroid replacement.

References

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