

Please read this bit first

This CPD/ CEU exercise is designed to take approximately two hours as a small group exercise within your laboratory. The Thistle QA CPD No is: **MT00025**.

Please keep a register of those taking part in the exercise. When the exercise is completed, please ask using the above email address, and we will send you a sheet showing the correct responses to each question.

Each attendee should claim two CPD points for completing the questions correctly, by retaining a copy of the relevant Thistle QA Participation Certificate as proof of registration on a Thistle QA EQA.

March 2006

Mineralocorticoid excess syndromes. (MCE syndrome)

Case Study.

A 58 year old man presented with a grand mal convulsion. He gave a history of recent weight loss, and weakness of all four limbs. He had been a heavy cigarette smoker all his life. His plasma and urinary analytes were :

Plasma

Na	137	mmol/L	(132-144)
K	2.1	mmol/L	(3.2-4.8)
CL	89	mmol/L	(98-108)
HCO ₃	36	mmol/L	(23-33)
Urea	3.8	mmol/L	(3.0-8.0)
Creat	0.08	mmol/L	(0.06-0.12)

Urine

Na	26	mmol/L
K	37	mmol/L

Comment : His ACTH level was 145 ng/L (reference range <60); his plasma cortisol concentration was > 1600 nmol/L. This case was an example of the ectopic-ACTH syndrome, which is commonly associated with lung carcinoma. The excess mineralocorticoid activity most probably represented two features :

- The mineralocorticoid activity of the excessive amount of circulating cortisol;
- The excessive production of DOC due to excessive ACTH stimulation (Williams R.H. et al, 1981).

This syndrome usually presents with pigmentation (ACTH) and hypokalaemia rather than 'full blown' Cushing's syndrome; these patients generally do not survive long enough to develop Cushingoid features.

Management of MCE syndromes.

The aetiology of the disorder determines the management, e.g. specific therapy for Cushing's syndrome and oedematous conditions; steroid replacement in enzyme defects; cancer therapy in the ectopic ACTH syndrome etc.

Primary hyperaldosteronism due to an adenoma is best treated surgically, and removal of the tumour usually results in complete cure. Chemotherapeutic treatment may also be successful in hyperaldosteronism due to an adenoma, and is the treatment of choice in hyperaldosteronism due to idiopathic bilateral hyperplasia. The drug of choice is spironolactone which usually has to be used in large doses (200 to 400 mg/day). In adrenal bilateral hyperplasia spironolactone therapy generally has to be supplemented by other antihypertensive therapy. Glucocorticoid – suppressible hyperaldosteronism is treated with dexamethasone or other similar steroids.

CPD QUESTIONS.

1. What test could be used to confirm a patient as a cigarette smoker ?
2. Discuss & describe the mechanism by which ACTH stimulates cortisol production.
3. What are Cushingoid features ?