A CASE OF ACUTE MUSCULAR WEAKNESS FROM ECTOPIC ACTH SECRETING NEUROENDOCRINE TUMOUR OF THE THYMUS

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Presenting complaint

A previously healthy 16 year old male from Manipur, India, presented with raid onset quadriparesis, vomiting and diarrhoea on a 3 month history of worsening facial oedema (fig 1) and a non-pruritic, maculo-papular rash on the chest and abdomen (fig 2).

5 days prior to admission the patient developed loose stools and vomiting (no blood, non-projectile and non-bilious).

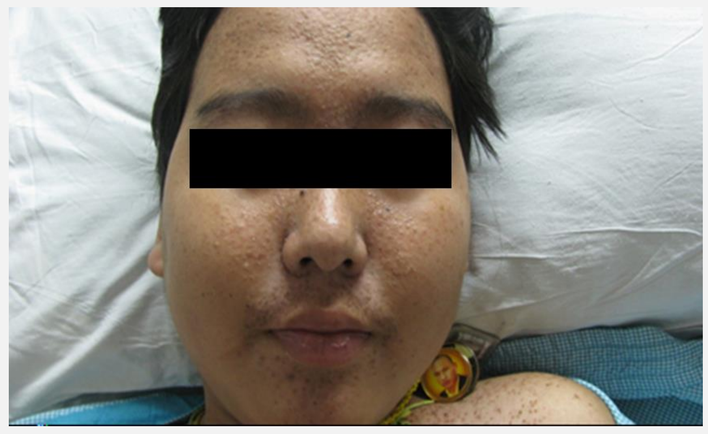

Fig 1.

Fig 2.

Investigations

Initial investigations revealed hyperglycaemia, hypokalaemia and a raised ESR.

The basal serum cortisol was 103µg/dl (N=7-22µg/dl) at 8:00am. 24-hour urinary free cortisol failed to suppress with 8mg of dexamethasone. Plasma ACTH and serum aldosterone were both markedly elevated at 263pg/ml (N=0-40pg/ml) and 528 pg/ml (N=25-31pg/ml) respectively.

Chest x ray revealed mediastinal widening (fig 3). CT scan revealed thymic mass with focal liver lesions (fig 4) and vertebral sclerotic lesions. Fine needle aspiration of the mediastinal mass revealed round cell lesions that resembled a neuroendocrine tumour.

MRI scan revealed no abnormalities (fig 5).

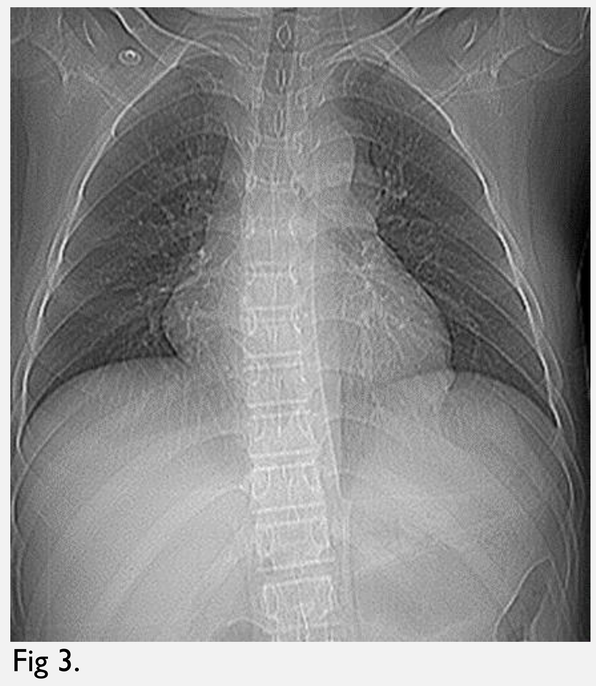

Fig 3.

Diagnosis

The patient was diagnosed with an ACTH-secreting non pituitary tumour of the thymus, with infiltration of the liver and bone. He was symptomatically managed with long acting Octeotride and followed up in clinic.

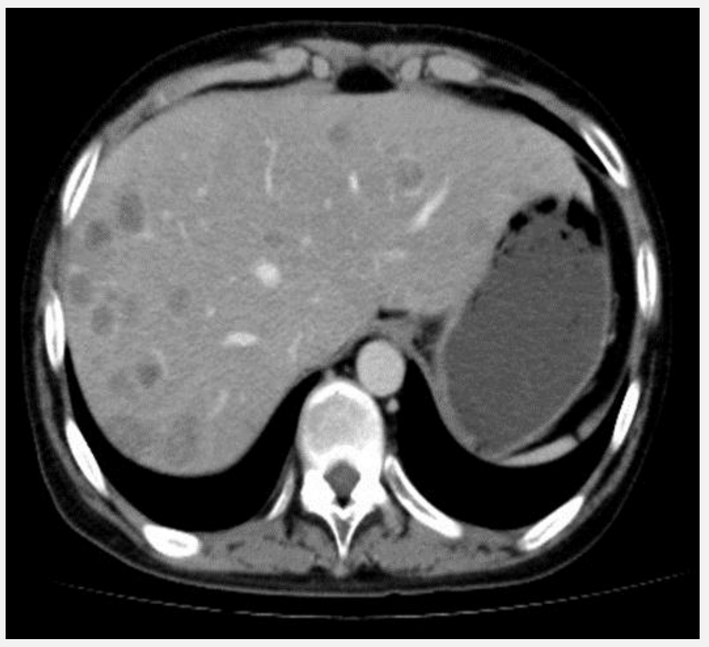

Fig 4.

Discussion

Neuroendocrine tumours (carcinoid and neuroendocrine carcinoma) of the thymus are extremely rare (1), and may present with Cushing's syndrome from ectopic ACTH excretion (2, 3)

Resection is the therapeutic modality of choice for thymic carcinoids that have not metastasised (3). When excision is not an option, medical management with octreotide (5) is favoured.

Extrathoracic metastasis has been reported in only 20-30% of cases and are associated with poor prognosis (4).

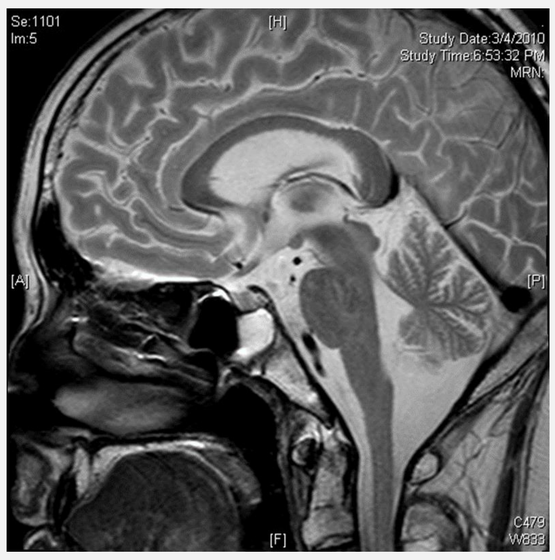

Fig 5.

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