A case of morning headache:Doege-Potter syndrome

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Introduction

Tumour induced hypoglycaemia is a rare disorder occurring in cancer patients caused by variety of tumours including islet and non-islet tumours. Non-islet cell tumour induced hypoglycaemia (NICTH) is rare paraneoplastic disorders normally associated with pleural solitary fibrous tumour but can also rarely occur in extra thoracic site.

Case Report

We describe 86 year old man not known to have diabetes mellitus who presented with 3 month history of early morning headache and dizziness relieved by consuming large amount of fizzy sweet drinks. On examination, he was noted to have large palpable mass on the left upper quadrant

Investigation

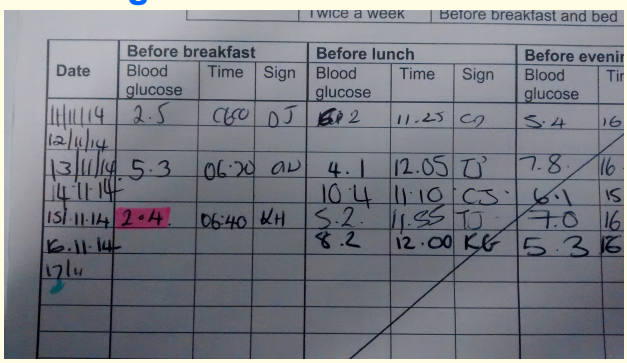

Fig 1 Capillary glucose monitoring showing pattern of early morning hypoglycaemia

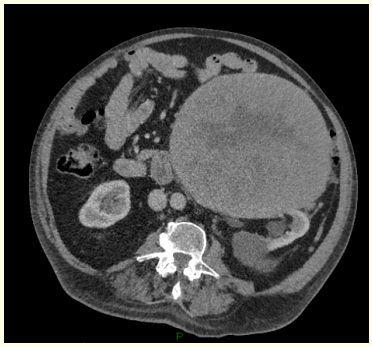

Fig 2 CT scan of the abdomen revealed huge solitary 22cm x 19cm x 16cm well defined, rounded solid heterogeneous retroperitoneal mass with internal ill-defined areas of necrosis

	Result	Normal Range
Plasma Glucose	2.4	3.9 – 5.5 mmol/l
Insulin	<1.0	17.8 – 173 pmol/l
C Peptide	81	298 – 2350 pmol/l
3-hydroxybutyrate	<0.1	0.3 – 0.5 mmol/l
Free fatty acid	0.3	0.00 – 0.72 mmol/l
IGF-1	7.1	6 – 36 nmol/l
IGF-2	94.5	nmol/l
IGF-2/IGF-1 ratio	13.3	Ratio <10
Short Synachten Test (0 minute/ 30 minute)	298 / 874	Normal response peak of >500 nmol/l
Serum Sulphonlyureas	Undetectable	
FT4	15.5	11 – 26 pmol/l
TSH	6.7	0.4 – 5.5 mU/l

Table 1. Initial inpatient investigations during symptomatic hypoglycaemia.

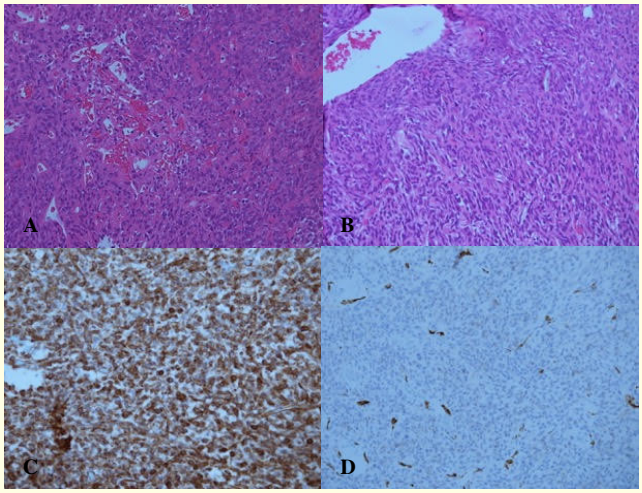

Fig 3 Core biopsy of renal mass : Haematoxylin eosin staining showing cellular spindle cell proliferation with "patternless" architecture and stag horn-like vessels (Image A, B)
Immunohistology showing positive CD-34 expression (Image C) and negative SMA (smooth muscle actin) (Image D)
Courtesy of Dr Sheikh Saleh (Consultant Histopathologist, Chesterfield Royal Hospital)

Discussion

Investigation results were consistent with diagnosis of NICTH secondary to extra thoracic solitary fibrous tumour. Solitary fibrous tumour are mesenchymal tumour thought to be of fibroblastic origin. It can be rarely associated with hypertrophic pulmonary osteoarthropathy (Pierre Marie-Bamberger syndrome) or/and refractory hypoglycaemia (Doege-Potter syndrome)[1].

Effective and ideal treatment would be surgical resection but in this case this was not feasible given patients comorbidity. Symptomatic treatment has been achieved using various modalities including administration of glucagon, somatostatin analogue, growth hormone and glucocorticoid[2]. He was managed conservatively with oral dexamethasone 1.5 mg od which resolves the recurrent hypoglycaemic episodes.

Overall mortality for 5 year and 10 year disease specific survival rates are 89% and 73% respectively.

Conclusions

In summary, we have described a rare phenomenon of Doege-Potter syndrome, a paraneoplastic phenomenon of hypoglycaemia associated with rare extra thoracic solitary fibrous tumour. It is important to investigate the aetiology thoroughly as management can be tailored individually for each patient.

Reference

1 Kalebi AY et al. Surgically cured hypoglycemia secondary to pleural solitary fibrous tumour: case report and update review on the Doege-Potter syndrome. J Cardiothorac Surg. 2009;4:45

2 Teale JD, Wark G. The effectiveness of different treatment options for non-islet cell tumour hypoglycaemia. Clin Endocrinol (Oxf). 2004;60(4):457–460