A dramatic example of work experience for a teenage boy

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Case history:
The 18 year gentleman presented with 10 month of being unrousable in the morning requiring his parents to feed him. He found himself in the kitchen in the middle of night eating and not knowing how he got there. He was expelled from school due to poor attendance performance. Insulinoma was suspected as he had a family history of MEN1. Unfortunately he had not had predictive genetic testing as his parents had not discussed the father’s diagnosis with him.

Investigations and method:
He underwent a 48 hour fast, becoming hypoglycaemic at 8 hours (blood glucose: 1.7 mmol/l, Insulin: 98pmol/l, Proinsulin: 13pmol/l, C peptide: 1114pmol/l). Fasting gut hormones were normal. Corrected calcium 2.38mmol/l, Parathyroid Hormone 79ng/l (14-72), IGF-1: 62.4nmol/L, Prolactin 166mU/ mU/l. MRI pancreas demonstrated a hyper vascular lesion in the tail of pancreas, but endoscopic ultrasound of pancreas demonstrated a 2 cm mass adjacent to the splenic hilum, with the rest of the pancreas diffusely abnormal suggestive of further lesions. FNA cytology of the 2cm lesion confirmed it to be a pancreatic endocrine tumour. He underwent pancreatic angiogram with insulin stimulation which confirmed a 12mm area of early arterial blush at the distal third of the pancreatic body and insulin secretion from this lesion. Genetic Testing was positive for MEN 1 gene: 1022G>A (p.Trp341X) gene mutation.

He had multiple episodes of hypoglycaemia whilst on the wards and it was deemed unsafe to discharge him prior to surgery. He did not tolerate diazoxide and required overnight 10% dextrose infusion to prevent hypoglycaemia. He underwent distal pancreatectomy and splenectomy which revealed a well differentiated pancreatic endocrine tumour (Stage T1N1) microadenomatosis and a lymph node metastasis. On a positive note after appealing to the LEA informing them of his medical condition and hypoglycaemia to which we attribute his difficulties at school, we are pleased that he has been readmitted to school and is starting A levels with a view to studying medicine.

Conclusions and points for discussion:
We illustrate a case of young male with MEN 1 who has a metastatic insulinoma, and a remaining pancreas with multiple residual neuroendocrine tumours. Currently he is asymptomatic but is at high risk for developing further symptomatic insulinomas as well as non-functioning neuroendocrine tumours. He has metastatic disease so he is at risk of developing further metastases. Currently there are no adjuvant therapies recommended. We will discuss the management dilemmas we faced and the potential options for further treatment including further surgery, somatostatin analogues as potential treatment of hypoglycaemia and tumour stasis, targeted radionuclide therapies and newer biological agents currently being used in the management of pancreatic NETs. This case also highlights the impact of insulinoma and MEN1 in a young person and the dilemmas within a family of revealing one member has a hereditary disease.