The challenges in managing a case of malignant insulinoma (MEN1)

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Introduction
Insulinomas are the most common, functioning, pancreatic neuro-endocrine tumours. The great majority (>90%) are nonmetastatic at presentation and can be cured surgically cured. The <10% patients with distant metastases have a median survival of <2 years. We present a case of a gentleman with a thirty year history of Multiple Endocrine neoplasia (MEN1), which highlights the various modalities of treatment and challenges faced due to progressive disease.

Case history
Mr RW, a 75 year old patient was first diagnosed with Primary Hyperparathyroidism causing hypercalcemia in 1972. He underwent parathyroidectomy and had a recurrence in 2002 needing repeat surgery. Genetic testing confirmed MEN 1. His son also was found to have same mutation. In 2005, he was diagnosed with Insulinoma in view of symptomatic hypoglycaemic episodes. He had metastatic lesions in the liver and spleen in addition to the pancreatic lesion.

In 2006 he underwent surgery including distal pancreatectomy, splenectomy and resection of liver lesions In addition radiofrequency ablation of liver lesions was performed. Due to his clinical and radiologically progressive disease, he underwent 3 further radiofrequency ablations in 2007. An Octreotide trial in 2008 failed to give symptomatic relief. Progressive disease necessitated another surgery in 2009, followed by further radiofrequency ablation. Diazoxide has provided symptomatic relief but with significant side effects. His Octreotide scan was negative.

His most recent scan has shown further progression of disease. He has been referred to our oncology colleagues for consideration of Everolimus,

Conclusions and points for discussion
Management of malignant Insulinoma can be very challenging. This case gives opportunity to review evidence for various treatment modalities and new developments in this field.