

Case

A 71-year-old man presented with a 2-month history of acute-onset, progressive pain in the right upper abdomen, associated with nausea. He had no other symptoms or prior co-morbidities and a random plasma glucose level checked 6 months earlier had been normal. On examination, he had tender hepatomegaly with a hard consistency and nodular surface that extended 4 cm below the costal margin. He was found to have random plasma glucose level of 47 mmol/L, and was started on insulin. However, he had no symptoms of hyperglycaemia (polyuria, polydipsia, or polyphagia). Despite treatment with 80 to 100 IU of premixed insulin, his plasma glucose remained uncontrolled. Serum amylase and lipase were normal. He underwent magnetic resonance imaging of his abdomen (Fig 1). His fasting plasma glucagon levels were 182 pmol/L (reference range, 22.7-65 pmol/L). He underwent

an ultrasound-guided biopsy of the liver lesion which was reported as neuroendocrine neoplasm with a MIB (Mind Bomb Homolog-1) proliferation index of 10 to 15% (Fig 2). The patient was treated with chemotherapy (carboplatin and etoposide, 6 cycles) and has been on follow-up for a year with diabetes in remission, and is clinically well although he has persistent hepatic metastases (revealed by Tc99m-octreotide scintigraphy) [Fig 3]. He was subsequently commenced on treatment with long-acting octreotide on a monthly basis.

Discussion

This patient was diagnosed to have a malignant glucagonoma in the pancreas with liver metastasis. Glucagonoma is a rare tumour with less than 250 cases having been reported in literature. About 75% are malignant and 50% have metastasised at the time

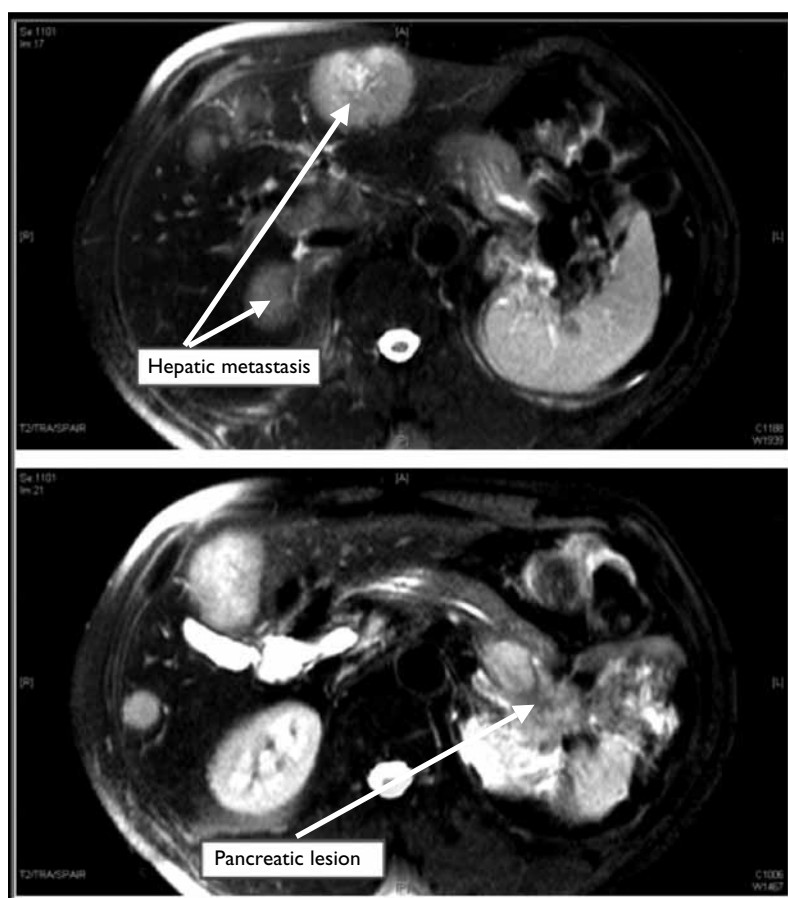


FIG 1. Magnetic resonance imaging of the abdomen: irregular pancreatic hyperintense lesion measuring 5 × 3.2 cm present in the distal body and tail with multiple hyperechoic lesions scattered in the liver of various sizes

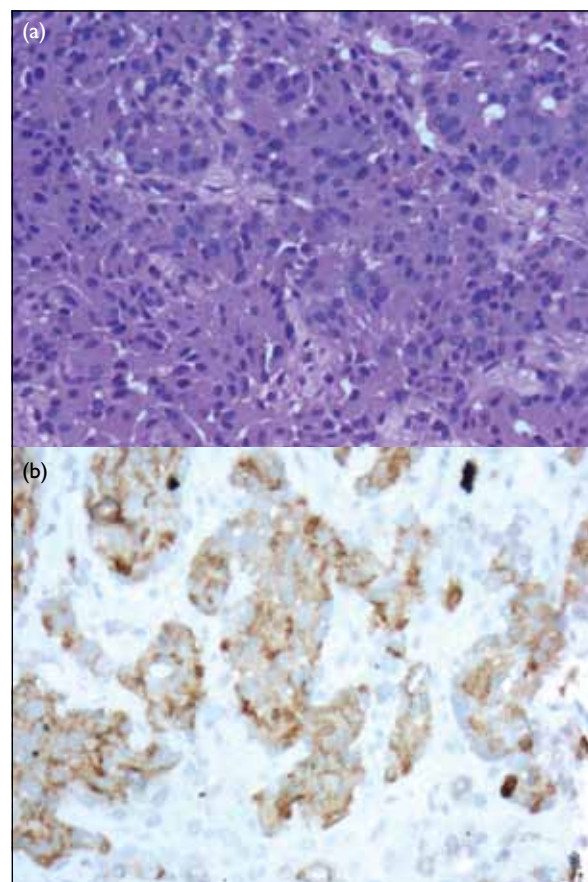


FIG 2. (a) Cluster of tumour cells (H&E, x 200). (b) Tumour cells staining positive for chromogranin (x 400)

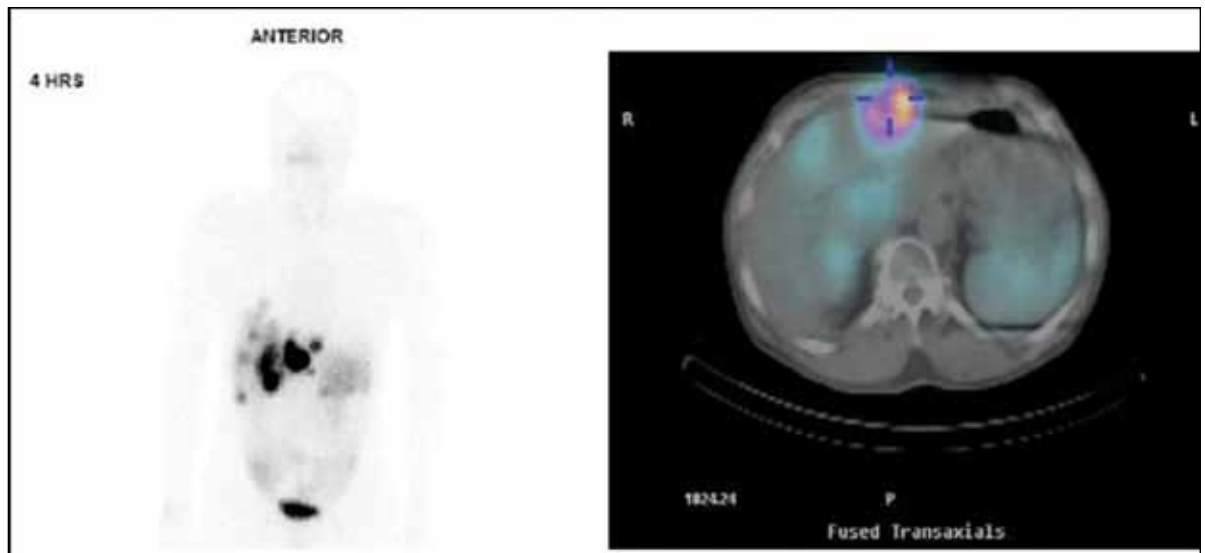


FIG 3. Tc99m-octreotide scintigraphy with single-photon emission computed tomography showing persistent multiple metastases in the liver

of diagnosis as in the patient reported here.¹ In our patient, the diagnosis was suggested by new-onset diabetes mellitus and a possible neuroendocrine tumour with liver metastasis revealed by imaging,² and confirmed by the presence of elevated glucagon levels. This patient did not have necrolytic migratory erythema, which is reportedly absent in 20% of persons having glucagonomas.³ Surgical resection is commonly not an option, due to late presentations. Therapy with a somatostatin analogue may be useful in the setting of metastatic disease. Patients with non-resectable or recurrent disease can be treated with chemotherapeutic agents such as streptozotocin and dacarbazine, interferon, or the selective use of arterial embolization.

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