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Imaging and localization of islet-cell tumours of the pancreas on CT and MRI

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Islet-cell tumours are neuroendocrine tumours that arise from the endocrine pancreas. They may be associated with a variety of syndromes and are subclassified into functioning and non-functioning tumours. They range from benign to malignant. They demonstrate characteristic features when imaged with both computed tomography (CT) and magnetic resonance imaging (MRI). Sensitivity and specificity, as well as detection of extrapancreatic extension, are generally superior with MRI. However, CT is currently still more readily available to patients. Multiphase, post-contrast series are commended for the evaluation of islet-cell tumours with either modality.

Key words: pancreas neoplasms; neuroendocrine neoplasms; computed tomography; magnetic resonance imaging.

Islet-cell tumours are a subgroup of gastrointestinal neuroendocrine tumours that occur within the endocrine pancreas.[1] Islet-cell tumours of the pancreas occur very rarely, accounting for only 1–5% of all pancreatic tumours[2]; they have an annual incidence of less than 1 in 100 000.[3–6] The tumours may occur in association with a variety of syndromes, including Zollinger–Ellison, multiple endocrine neoplasia type 1,

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von Hippel–Lindau, neurofibromatosis type 1 and carcinoid syndrome[2,4,7], and they may be functioning or non-functioning. Non-functioning islet-cell tumours account for 15–52% of islet-cell tumours.[2,4,5] Functioning tumours tend to present early in the course of the disease when the tumour is small, secondary to the clinical manifestations of the hormone produced, whereas non-functioning tumours tend to present when they are large in size due to the mass effect of the tumour.[3] The frequency of malignancy varies. Insulinomas are most commonly benign, while 60% of gastrinomas are malignant. The majority of the remaining tumours, including the non-functional lesions, are malignant.[1] Malignancy of an islet-cell tumour often cannot be diagnosed histologically. Aggressive potential is determined by the presence of metastases or local invasion beyond the substance of the pancreas.[1]

FUNCTIONING ISLET-CELL TUMOURS

Islet-cell tumours arise from cells in the islets of Langerhans. Normal islets contain B cells, which produce insulin; A cells, which produce glucagon; D cells, which produce somatostatin; D1 cells, which produce pancreatic polypeptide; and D2 cells which produce vasoactive intestinal polypeptide (VIP). Functional tumours arising from these cells are named for the predominant hormone produced (often a tumour may secrete more than one type of hormone, but usually one hormone predominates).[4] Diagnoses of functioning islet-cell tumours are almost always established biochemically when a lesion is small.[5] Insulinomas and gastrinomas generally present when small; 90% are diagnosed when <2 cm in size. These tumours may also be multiple.[6]

Insulinoma

These are the most common of the islet-cell tumours, comprising 50% of the total. The typical patient age range is 30–60 years. There is an equal sex distribution, although at least one reference cites a female predominance.[8] They are usually solitary. Ninety per cent are located within the pancreas, and there is an equal pancreatic distribution between the head, body and tail; 6–10% of insulinomas are malignant. Multiple tumours present in 10% of cases and may be associated with multiple endocrine neoplasia type 1 (MEN-1); 4% of insulinomas overall are associated with MEN-1.[4,8]

Insulinomas tend to be small tumours: 90% are <2 cm, 66% are <1.5 cm, and 40% are <1 cm.[4]

Gastrinoma

Gastrinoma is the second most common islet-cell tumour, accounting for 20% of the total.[4] There is a male predominance, and presentation is most common during middle age.[4] Gastrinomas cause hypersecretion of gastrin, which results in hyperacidity and Zollinger–Ellison syndrome, abdominal pain and diarrhoea.[4]

Gastrinomas, relative to insulinomas, are more often extra-pancreatic and multiple. They also tend to be even smaller and less vascular than insulinomas. These features make them even more difficult to localize than insulinomas.[4] Ninety per cent of these tumours are located within the ‘gastrinoma triangle’ which is formed by the junction between the neck and body of the pancreas medially, the second and third portions of the duodenum inferiorly, and the junction of the cystic and common bile ducts