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GI carcinoid tumours: appearance of the primary and detecting metastases

Richard M. Gore* MD

Professor of Radiology, Northwestern University Medical School; Chief, Gastrointestinal Radiology, Evanston Northwestern Healthcare

Jonathan W. Berlin MD

Assistant Professor of Radiology, Northwestern University Medical School

Uday K. Mehta MD

Assistant Professor of Radiology, Northwestern University Medical School

Geraldine M. Newmark MD

Assistant Professor of Radiology, Northwestern University Medical School; Chief, Body Imaging, Evanston Northwestern Healthcare

Department of Radiology, Evanston Northwestern Healthcare, Northwestern University, 2650 Ridge Avenue, Evanston, IL 60201, USA

Vahid Yaghmai MD

Department of Radiology, Northwestern Memorial Hospital, Northwestern University, 676 N. St Clair, No. 800, Chicago, IL 60611, USA

Gastrointestinal carcinoid tumours are notoriously difficult to diagnose in the absence of the carcinoid syndrome. The clinical presentation is typically non-specific, and patients often go undiagnosed for years. Recent advances in computed tomography (CT), magnetic resonance (MR), endoscopic ultrasound, and nuclear scintigraphy have combined to improve the diagnosis and staging of this fascinating tumour. In this chapter the applications of cross-sectional imaging in patients with gastrointestinal carcinoid tumours is presented.

Key words: carcinoid; CT; MRI; endoscopic ultrasound.

* Corresponding author. Tel.: +1 847 570 2475; Fax: +1 847 570 2942.
E-mail address: rmgore1953@aol.com (R.M. Gore).

CARCINOID TUMOURS

Carcinoid tumours are slow-growing enigmatic malignancies that have a natural history unlike that of any other neoplasm.[1] They represent a wide spectrum of tumours which originate from a variety of different neuroendocrine cell types.[2] Since the gastrointestinal (GI) tract is the largest neuroendocrine organ in the body, it is not surprising that over 90% of all carcinoid tumours originate in the GI tract. Unlike other endocrine organs, the endocrine cells of the gut are dispersed as single cells from the gastro-oesophageal junction to the rectum.[3] These cells are part of the amine precursor uptake and decarboxylation system and give this tumour its most distinctive feature: its ability to secrete biogenic amines and polypeptide hormones that cause the carcinoid syndrome.[4]

Pathological features

Carcinoid tumours are usually composed of uniform cells with moderate amounts of granular cytoplasm and round nuclei. The embryonic origin of carcinoids is the neuroectoderm argentaffin cells located in the mucosa near the base of the crypts of Lieberkühn. There are five major histological patterns: (1) insular; (2) trabecular or ribbon; (3) tubular or rosette-like; (4) atypical or poorly differentiated; and (5) mixed. These patterns correlate with the embryologic location: foregut-origin tumours show a trabecular growth pattern, midgut tumours tend to grow with an insular pattern, and hindgut tumours of the midtransverse or distal colon show mixed patterns of growth.[5,6]

Carcinoid tumours can be identified by silver impregnation stains, either argyrophilic or argentophilic. Because carcinoid tumours produce numerous polypeptides and hormones, immunohistochemistry markers are found for serotonin, gastrin, glucagon, somatostatin, insulin, substance P, and pancreatic polypeptide.[7]

The distinction between benign and malignant carcinoid tumours is more difficult than with other GI epithelial tumours. Because individual cells making up the tumour usually appear bland, and only rarely show mitoses or hyperchromatism, the diagnosis of malignancy requires gross or microscopic evidence of invasion. All carcinoids should be considered potentially malignant, even though the majority are less than 2 cm in size and those found incidentally are very likely to be benign. Factors that determine malignant behaviour include: (1) the extent of invasion; (2) tumour size; and (3) site of origin.[8,9] Metastases occur in only 50% of patients with tumours smaller than 1 cm, but 95% when tumours are larger than 2 cm.[10]

Carcinoid tumours spread locally to regional lymph nodes, liver, other intra-abdominal organs and lung, but only rarely to bone. Only a tiny minority of those that arise in the stomach or rectum spread beyond their site of origin. Appendiceal carcinoids almost never metastasize because even small tumours obstruct the lumen to produce appendicitis early, prompting surgical removal before metastases occur. Small carcinoids normally do not invade or obstruct the bowel lumen but can penetrate the muscle layer, causing marked desmoplasia that may lead to adhesions, bowel kinking and angulation, and obstruction.[11,12]

Massive fibrosis of the mesenteries, omenta, and peritoneum may result from the leakage of serotonin and other vasoactive substances independent of any intrinsic disease.[13] This fibrosis is responsible for the fixation, kinking, and angulation seen on small bowel series and the stellate soft tissue density mass on CT and MR scans.[14,15]