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Pictorial essay

Carcinoid tumors of the abdomen: CT features

J.-P. Pelage,[1] P. Soyer,[1] M. Boudiaf,[1] I. Brocheriou-Spelle,[2] A.-C. Dufresne,[1] J. Coumbaras,[1] R. Rymer[1]

[1]Department of Body and Vascular Imaging, Hôpital Lariboisière, 2 rue Ambroise Paré, 75475 Paris Cedex 10, France
[2]Department of Pathology, Hôpital Lariboisière, 2 rue Ambroise Paré, 75475 Paris Cedex 10, France

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Abstract
Carcinoid tumors are rare neuroendocrine neoplasms that belong to a more general category of tumor called the *APUDomas*. Ninety percent of carcinoid tumors are located in the gastrointestinal tract. Abdominal carcinoid tumors are categorized according to the division of the primitive gut from which they arise. Carcinoid tumors originating from the foregut develop in the gastric wall, duodenum, and pancreas; those originating from the midgut develop from the small bowel, appendix, and right colon; and those originating from the hindgut develop from the transverse or left colon or from the rectum. This report illustrates the computed tomographic appearance of primary and metastatic carcinoid tumors of the abdomen. Among the different organs that may be involved by metastases from carcinoid tumor, special emphasis is placed on the liver.

Key words: Carcinoid tumors—Abdomen—Computed tomography.

Carcinoid tumors are tumors of the diffuse endocrine system. They represent 55% of gastrointestinal (GI) endocrine tumors. Carcinoid tumors are rare neuroendocrine neoplasms that belong to a more general category of tumor called the *APUDomas* (i.e., amine precursor uptake and decarboxylation tumors). Ninety percent of carcinoid tumors are located in the GI tract. Abdominal carcinoid tumors are categorized according to the division of the primitive gut from which they arise. Carcinoid tumors originating from the foregut develop in the gastric wall, duodenum, and pancreas; those originating from the midgut develop in the small bowel, appendix, and right colon; and those originating from the hindgut develop in the transverse or left colon or in the rectum.

This report illustrates the computed tomographic (CT) appearances of primary and metastatic carcinoid tumors of the abdomen. Among the different organs that may be involved by metastases from carcinoid tumor, special emphasis is given to the liver.

Correspondence to: J.-P. Pelage

Histological and clinical background

Macroscopically, the well-differentiated carcinoid tumor shows homogeneous yellow color on slice section. Larger tumors (<4 cm) are responsible for thickening of the intestinal wall, with stranding of the mesentery and lymph nodes of the same appearance. Microscopically, the carcinoid tumor is made of small and regular cells with clear cytoplasm containing a round nucleus without atypia or prominent nucleoli. In the absence of biological evidence of hormonal secretion, the diagnosis relies on immunohistochemical preparation or electron microscopy. Although carcinoid tumors usually grow slowly, they all have potential for being malignant. Carcinoid tumors smaller than 2 cm in the appendix, with neither serosal involvement nor metastatic lymph node, can be considered as benign. Histologically it may be difficult to distinguish between the less and the more aggressive tumors [1]. Metastases or spread to adjacent organs are the only criteria for ascertaining the malignant nature.

Carcinoid syndrome (major clinical manifestation of a carcinoid tumor) occurs in 4–10% of cases when vasoactive hormones can enter into the systemic circulation. It is especially present in cases of hepatic metastases [2]. Prolonged survival of patients with carcinoid tumor is not uncommon, with reported survival rates of more than 80% at 5 years, even in the presence of distant metastases [3]. Treatment should prevent tumor growth and limit biological effects of tumor secretion. Resection of the

tumor is the best therapeutic option. Rectal and duodenal carcinoid tumors smaller than 1.5 cm can be resected endoscopically. Hepatic metastases can be resected surgically except when diffuse involvement is present. Hepatic arterial chemoembolization is indicated when diffuse involvement is associated with a marked carcinoid syndrome. Long-acting somatostatin analogue, which allows control of diarrhea and flushing, is currently the best available noninvasive treatment of carcinoid syndrome.

CT features

Esophagus and stomach

Only a few cases of carcinoid tumor of the esophagus have been reported. Carcinoid tumors of the esophagus and stomach account for fewer than 5% of GI carcinoid tumors [4]. The tumor arising from the basal part of the mucosa has a polypoid appearance (Fig. 1). Carcinoid tumor of the stomach may be associated with chronic atrophic gastritis, especially in the case of pernicious anemia, and may have a central ulceration visible on upper GI tract series.

Duodenum

The frequency of duodenal carcinoids tumors is similar to that of gastric carcinoids. They are located mostly in the first and second portions of the duodenum. They may be associated with multiple neuroendocrine neoplasia, Zollinger-Ellison syndrome, or neurofibromatosis. They are usually low-grade malignant tumors that appear as small sessile polyps. CT shows well-defined homogeneous and lobulated soft tissue mass (Fig. 2). In the case of periampullar tumor, CT may show bile duct dilatation. Endoscopy is the best technique for diagnosis because of biopsy.

Small bowel

The small bowel is the second most frequent site of carcinoid tumors after the appendix, representing up to 20% of GI carcinoids. Ileal and jejunal carcinoid tumors are usually aggressive because of their hidden location. Thus, hepatic metastases are often present at the time of diagnosis. CT may show a polypoid or mural smoothly marginated subtle mass within a bowel loop or bowel wall thickening [5]. In some cases, dilated small bowel loops can be seen [1, 6]. CT may show virtually pathognomonic features, with a mesenteric mass in a stellate configuration associated with dense strands in the mesentery [7] (Fig. 3). The strandlike densities that radiate from the mesenteric mass tend to be directly related to the degree of fibrosis seen on tissue slides as a desmoplastic response caused by serotonin [8]. The degree of linear soft tissue densities correlates poorly with the tumor infiltration along neurovascular bundles [5, 8]. The CT finding of calcifications within a mesenteric carcinoid tumor is common in approximately 70% of cases [8]. CT may also demonstrate metastatic lymph nodes, ascites, and hepatic metastases.

Appendix

The appendix is the most common location of carcinoid tumors, representing 40% of GI tract carcinoids. Carcinoid tumors of the appendix are usually not imaged and are often discovered incidentally at appendectomy (Fig. 4). CT may depict a mass that may infiltrate the cecum and the mesentery. In general, CT plays a very limited role in the evaluation of carcinoid tumor of the appendix.

Colon and rectum

Carcinoid tumors of the rectum represent 15% of GI carcinoids. They are usually small and are discovered incidentally during proctoscopy. Carcinoid tumors of the colon are most often in the distal colon (Fig. 5), whereas carcinoid tumors of the proximal colon are often the consequence of extension of appendiceal or distal ileal carcinoid tumors. They usually appear as polypoid or mural masses. Biopsy under proctoscopic guidance provides the diagnosis and may allow resection. For large tumors, endoscopic sonography provides information regarding local extent.

Pancreas

Primary carcinoid tumor of the pancreas is exceedingly rare. From a radiologic point of view, the tumor may be indistinguishable from pancreatic islet cell tumors [9]. The carcinoid tumor is usually homogeneous on CT (Fig. 6). Calcification within the tumor is often observed [9]. In very few cases, the tumor may be responsible for dilatation of the biliary tract or chronic pancreatitis with dilatation of the pancreatic duct [10]. The frequency of hepatic metastases often present in late stages has not been reported in the literature. In fact, definitive diagnosis relies mainly on anatomopathologic techniques based on immunohistochemical studies.

Metastases

Hepatic metastases at initial presentation

Carcinoid syndrome should prompt the clinician to look for hepatic metastases. On CT, hepatic metastases may

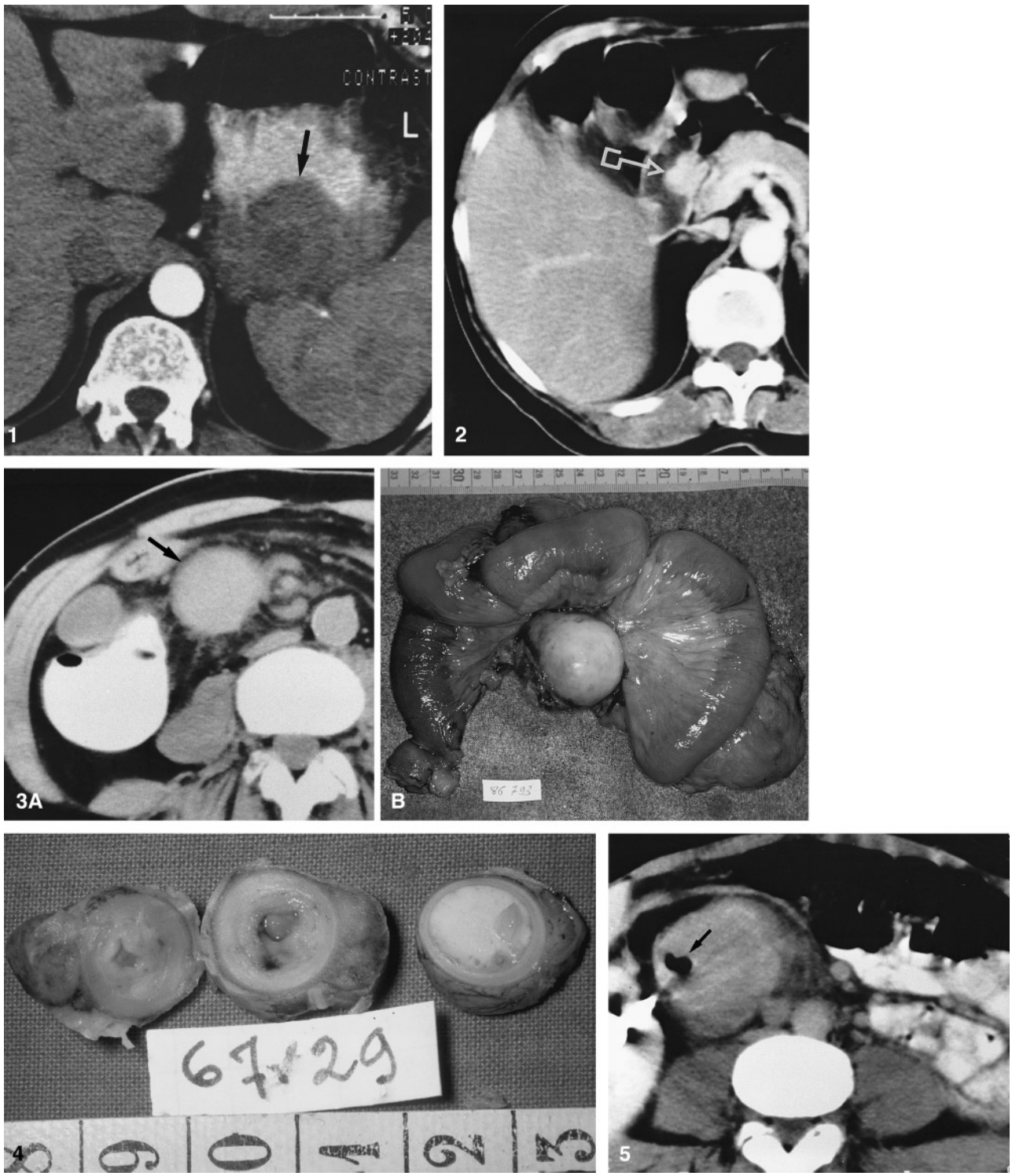

Fig. 1. A 50-year-old man with carcinoid tumor of the stomach. CT shows nonspecific parietal mass (*arrow*) arising from the gastric wall.

Fig. 2. A 69-year-old woman with primary carcinoid tumor of the duodenum. CT shows a well-defined enhancing lobulated mass (*arrow*) within the duodenum.

Fig. 3. A 68-year-old man with ileal carcinoid tumor. **A** CT shows mesenteric mass (*arrow*) with emanating strands corresponding to thickened neurovascular bundles. This feature is virtually pathognomonic for a carcinoid tumor. **B** Corresponding gross specimen after surgical resection confirms the mesenteric location of the tumor.

Fig. 4. A 29-year-old man with carcinoid tumor of the appendix. Diagnosis was made after surgery performed for suspected appendicitis. Carcinoid tumors of the appendix are often found incidentally during appendectomy, when they cause appendicitis.

Fig. 5. A 60-year-old woman referred for abdominal pain. CT depicts a large abdominal mass. Air (*arrow*) within the mass suggests a gastrointestinal origin. A carcinoid tumor originating from a duplication of the sigmoid was found during surgery.

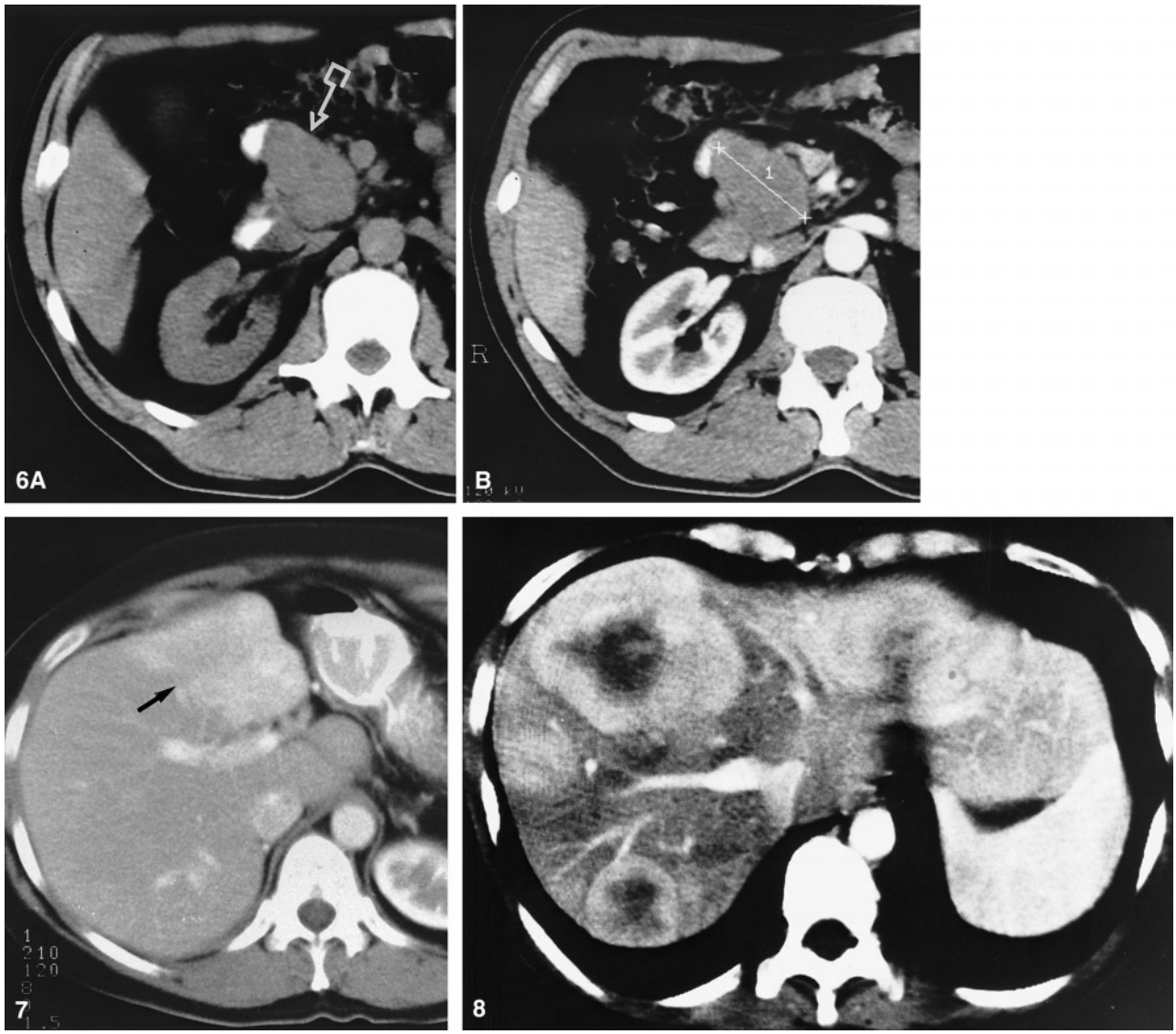

Fig. 6. A 66-year-old man with primary carcinoid tumor of the pancreatic head. **A** On unenhanced CT, the tumor (*arrow*) is isoattenuated relative to the pancreatic parenchyma. The duodenum is markedly displaced. **B** The tumor shows homogeneous enhancement after intravenous administration of iodinated contrast material. These features are different from those commonly observed with pancreatic adenocarcinoma, which is more likely to appear hypoattenuated.

Fig. 7. A 41-year-old woman with hepatic metastases from carcinoid tumor. CT obtained during the early phase of the bolus shows hypervascular lesion (*arrow*). To depict these hypervascular lesions more clearly, it is important to make the acquisition coincide with the arterial dominant phase of the bolus. (Courtesy of Elliot K. Fishman, M.D., The Johns Hopkins Hospital, Baltimore, Maryland).

Fig. 8. A 46-year-old woman with hepatic metastases from carcinoid tumor. CT obtained during the early phase of the bolus shows peripheral enhancement of the viable portions of the metastases, whereas the central necrotic portion of the metastases is hypoattenuated relative to the hepatic parenchyma.

have different features (Fig. 7) and, because of their hypervascular nature, are better depicted during the arterial dominant phase of the bolus. The tumors may appear isoattenuated when compared with the adjacent hepatic parenchyma during the portal dominant phase. In this regard, spiral CT has improved the detectability of hepatic metastases, especially in small lesions. In some cases, large metastases may have a pseudocystic pattern (Fig. 8).

Hepatic metastases after treatment

During chemotherapy, hepatic metastases may become calcified (Fig. 9). Hypervascular metastases usually be-

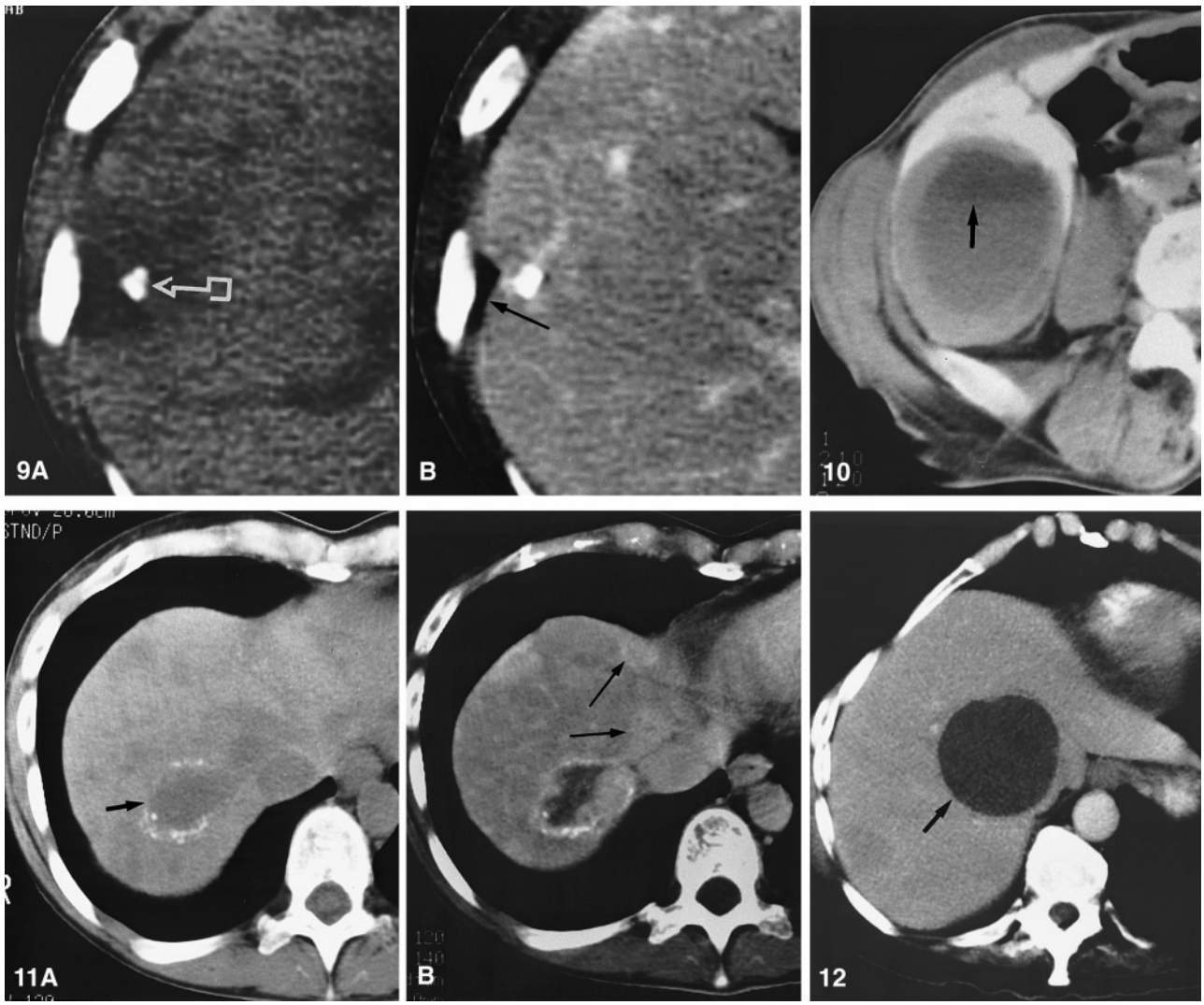

Fig. 9. A 60-year-old man who had intraarterial chemotherapy because of unresectable hepatic metastases from carcinoid tumor. **A** Unenhanced CT shows hypoattenuating lesion that contains calcification (*arrow*). **B** After intravenous administration of iodinated contrast material, retraction of the liver capsule is seen adjacent to the tumor (*arrow*).

Fig. 10. A 71-year-old man who had systemic chemotherapy because of unresectable hepatic metastases from carcinoid tumor. CT shows fluid–fluid level (*arrow*) within the lesion, indicating internal necrosis and hemorrhage.

Fig. 11. A 35-year-old woman who had systemic chemotherapy because of unresectable hepatic metastases from carcinoid tumor. **A** Unenhanced CT shows hypoattenuated lesion (*arrow*) that contains peripheral calcifications. **B** CT obtained after intravenous administration of iodinated contrast material shows peripheral enhancement of the lesion and central necrosis. Additional hepatic lesions are seen (*arrows*).

Fig. 12. A 50-year-old woman who had systemic chemotherapy because of unresectable hepatic metastases from carcinoid tumor. Secondary to marked tumor necrosis consequent to chemotherapy, CT shows a lesion with a cystic pattern (*arrow*).

come necrotic (Figs. 10, 11). In some cases, marked necrosis may lead to a pseudocystic or even cystic pattern (Fig. 12). Retraction of the liver capsule adjacent to the carcinoid tumor can be observed after intraarterial chemotherapy (Fig. 9).

Other metastatic sites

Carcinoid tumors may metastasize to different organs. The most frequent sites include heart, brain, and bone. Metastases to the heart and the brain are better detected with magnetic resonance imaging than with CT.

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