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Pictorial Essay

Mesenchymal Tumors of the Pancreas: CT Findings

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Abstract: This article reviews the CT imaging features of the most frequent mesenchymal tumors of the pancreas and stresses important distinctive patterns that may help distinguish specific entities. Various neoplasms (lymphangioma, lipoma, teratoma, pancreatoblastoma, schwannoma, neurofibroma, lymphoma, and sarcoma) are reviewed, with key differential points (structure, fatty and water densities, calcification, pattern of contrast enhancement, vascularization, and necrotic or regressive changes) emphasized. In addition, epithelial tumors are considered in the differential diagnosis.
Index Terms: Computed tomography—Pancreas, neoplasms—Mesenchyma.

Although the great majority of both benign and malignant pancreatic neoplasms arise from pancreatic epithelial cells, mesenchymal tumors, while rare, can derive from the connective, lymphatic, vascular, and neuronal tissues of the pancreas (1). Mesenchymal tumors account for 1–2% of all pancreatic tumors and are classified according to their histologic origin (1). In selected instances, cross-sectional imaging can offer a specific diagnosis of histologic types. The aim of this essay is to describe the CT findings of these rather uncommon entities.

LYMPHANGIOMA

Pancreatic lymphangiomas constitute <1% of all lymphangiomas (2). Their histogenesis is uncertain. Proposed theories have suggested an inherited abnormality or an embryologic or traumatic origin, whereas other authors believe lymphangioma is a true neoplastic lesion (3).

The tumor is typically multicystic, with cysts of various sizes ranging from microscopic to as large as 10 cm, separated by thin septa. The cystic spaces contain serous, serous-hematic, or chylous fluid. A peripheral thin capsule of fibrous tissue is present (2). Calcifications, both parietal and intracystic, are rare (4).

Pancreatic lymphangiomas can grow within the parenchyma or may be connected to the organ by a pedicle that can be liable to torsion.

CT shows a cystic mass, uni- or multilocular, with thin enhancing endocystic septa (5). The walls are generally thin and rarely calcify (Fig. 1). The diagnosis of pedicle forms (Fig. 2) is made easier by the use of helical scanners, which permit optimal multiplanar reconstructions.

All the cystic lesions encountered in the pancreas must be considered in the differential diagnosis, including inflammatory pseudocysts, hydatid cysts, dysplastic cysts, and neoplastic cysts (microcystic serous adenoma, macrocystic adenoma/adenocarcinoma, cystic schwannoma, and cystic islet cell tumor).

LIPOMA

Visceral lipomas are commonly found in the digestive tract; however, they are very uncommon in the pancreas, with only a few cases reported in the literature (6). Histologically they are composed of lobules of mature adipose cells. They are circumscribed by a thin collagen capsule, which allows a better surgical enucleation as well as the distinction between lipoma and lipomatosis (7).

The densitometric evaluation is diagnostic. Values range from −30 to −120 HU, with homogeneous appearance and no significant contrast enhancement (Fig. 3) indicating a benign fatty structure. Thin fibroreticular septa may occasionally be seen within the mass, representing the interlobular septa (8). The absolute lack of infiltration of surrounding tissues is typical.

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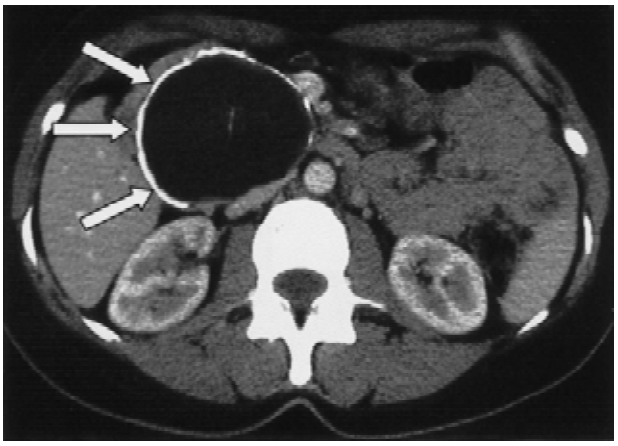

FIG. 1. Cystic lymphangioma of the pancreas. This 45-year-old woman had left renal colic. Spiral CT shows a large cystic mass in the cephalopancreatic region. In this case, the walls show atypical calcified pattern (arrows), and thin endocystic septa are demonstrated. No enhanced papillary endocystic growth or nodular solid components are seen. At pathology, the cyst contained serous and bloody fluid. Microscopically lymphatic endothelium was found to line the cystic spaces.

The differential diagnosis includes localized lipomatosis (generally showing a direct contact with the peripancreatic fat and an absent capsule), cystic teratoma (calcifications and various conglomerate tissues with heterogeneous densities present), and other fatty neoplasms such as fibrolipoma, lipoblastoma, and liposarcoma (7).

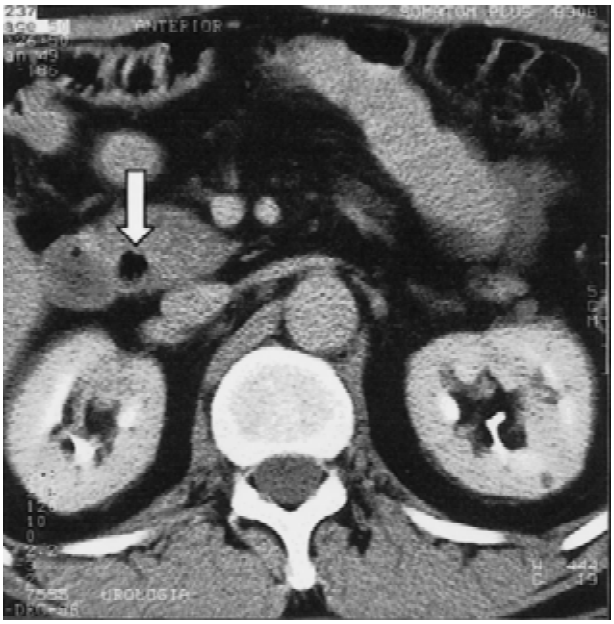

FIG. 3. Pancreatic lipoma. This 67-year-old man had lung cancer and brain and skeletal metastases. A small round lesion with homogeneous fatty density (–70 HU) is seen in the pancreatic head (arrow) without direct connection with peripancreatic fat. This lipoma was confirmed at autopsy.

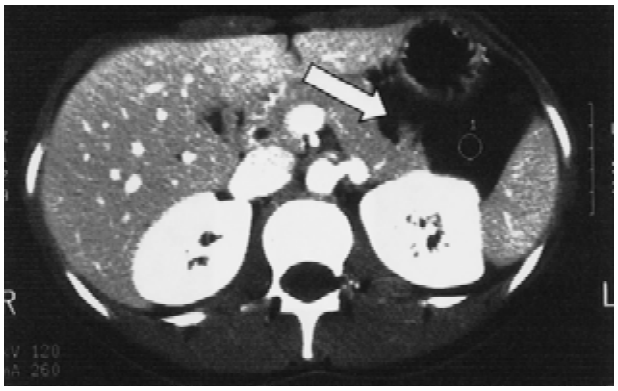

FIG. 2. Cystic lymphangioma of the pancreas. This 28-year-old woman had left upper quadrant abdominal pain. Spiral CT shows a large mass with homogeneous water density [region of interest], thin walls, and a pedicle (arrow) arising from the pancreatic tail.

TERATOMA

Pancreatic teratoma is very rare. The tumor develops from pluripotential cells of embryonic remnants of the ectodermal line and usually contains a combination of both cystic and solid elements, including hair, teeth, calcifications, cartilage, and dermal appendages such as hair follicles, sweat glands, and sebaceous material (9). No cases of malignant degeneration have been described in the pancreas. The CT findings depend on the relative proportions of various tissues composing the lesion with a fatty component (homogeneous negative Hounsfield unit numbers), fatty/fluid levels, hair/fluid interfaces, or a more complex appearance with solid, cystic, and calcified components (10) (Fig. 4).

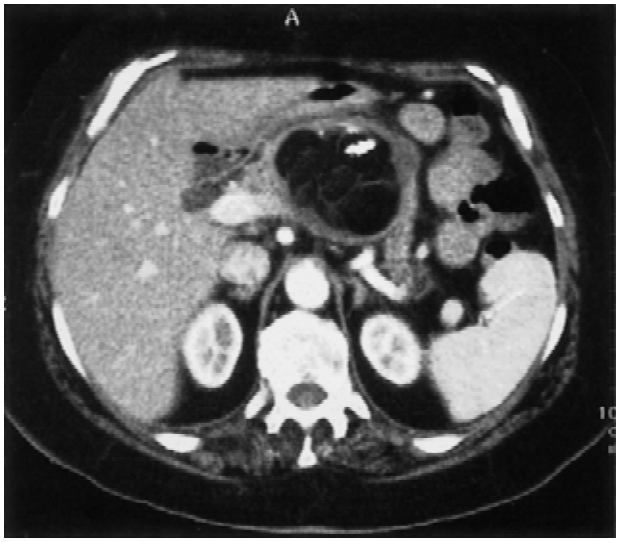

FIG. 4. Mature teratoma of the pancreas. This 39-year-old woman had dyspepsia and digestive troubles. Spiral CT shows a large heterogeneous density mass in the pancreatic body. The predominant component is fatty; calcifications and mucinous (30 HU) components are also documented.

The differential diagnosis includes foregut-derived cyst, which, when containing fat, may show similar CT appearance, and liposarcoma (7,11).

PANCREATOBLASTOMA

Pancreatoblastoma is a rare tumor that occurs almost exclusively during childhood. It is an embryonal neoplasm arising from multipotential stem cells that mimic the embryogenesis of the pancreas with both epithelial and mesenchymal components (12). Areas characterized by functioning or nonfunctioning neuroendocrine tissue are also present (13). Typically, these tumors are located in the cephalic region of the pancreas. In most cases, the tumor is surrounded by a firm capsule that facilitates the surgical approach. If present, metastases are more frequently seen in lymph nodes, liver, and lung (12).

The CT appearance is nonspecific, showing a solid and hypovascular or a complex multiloculated cystic mass (13) (Fig. 5). Because of its large size, the tumor is often misdiagnosed as neuroblastoma.

SCHWANNOMA

Visceral schwannomas, arising from the Schwann cells enveloping the sympathetic and parasympathetic nervous fibers, are rare lesions. The pancreas is an extremely unusual site of origin for this tumor, with only a few cases of benign schwannoma described in the literature (14).

The tumor is usually encapsulated. Pathologically it consists of two components, the quantitative distribution of which is highly variable from tumor to tumor: an organized cellular component (Antoni type A area) and a loose hypocellular component (Antoni type B area) (15).

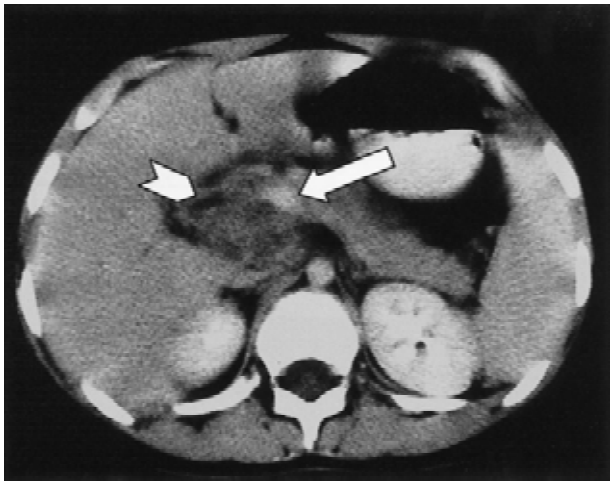

FIG. 5. Pancreatoblastoma. This 10-year-old girl had fever, nausea, abdominal pain, and weight loss. CT scan shows an inhomogeneous mass with necrotic components (arrowhead) in the pancreatic head. The portal vein (arrow) appears to be encased by the tumor.

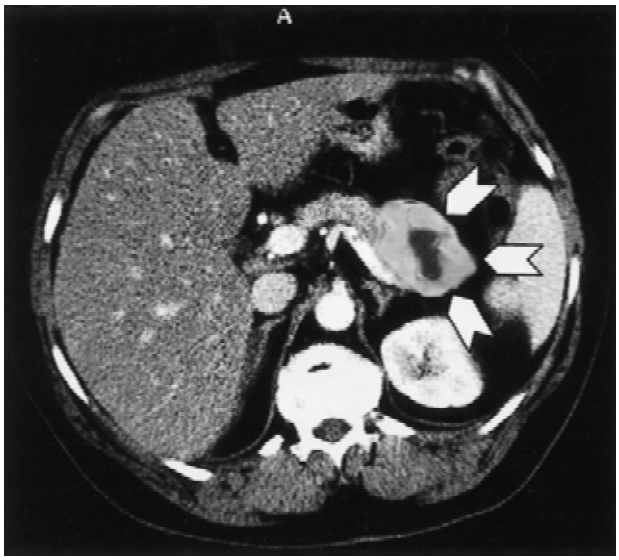

FIG. 6. Benign pancreatic schwannoma. This 58-year-old woman had nausea, dyspepsia, and vague abdominal pain. Spiral CT shows a large enhancing mass in the pancreatic tail (arrowhead) without infiltration of the peripancreatic fat. A hypodense water density central core is documented. Pathologic analysis demonstrated a nearly exclusive representation of Antoni type A tissue.

These features correlate quite well with the CT findings (16). In forms with a nearly exclusive or a prevalent representation of Antoni type A tissue, CT shows solid highly enhancing masses with coarse central necrosis (Fig. 6) or inhomogeneous hypodense lesions (Fig. 7). In the latter case, the near-fluid or <0 HU values are due to the high lipidic content of the tumor, and the reticular vascular component gives rise to the contrast enhancement and complex appearance of these lesions, which may occasionally appear septated (15,16). In forms with a high prevalence of Antoni type B tissue, the loose architecture, with poor cellularity and no significant fibrillar component of the stroma, is often associated with xantomatous regression, containing aggregates of lipid-laden cells, and/or pseudocystic degeneration, most likely secondary to vascular thrombosis (17). These phenomena may cause a pseudocystic appearance, with homogeneous fluid Hounsfield unit readings, without significant contrast enhancement (Fig. 8).

In the case of solid and hypervascular radiological patterns, the differential diagnosis includes endocrine tumors. In the case of cystic/pseudocystic patterns, the differential diagnosis includes epithelial tumors (microcystic serous adenoma, solid and papillary neoplasm, and mucinous macrocystic tumor) (16).

NEUROFIBROMA

The neurofibroma, and in particular the plexiform variant, is the hallmark of von Recklinghausen disease. The structure of the tumor consists of fibroblasts, Schwann cells, and neural elements that expand and infiltrate the nerve diffusely.

The pancreatic localization is exceptional. A regional

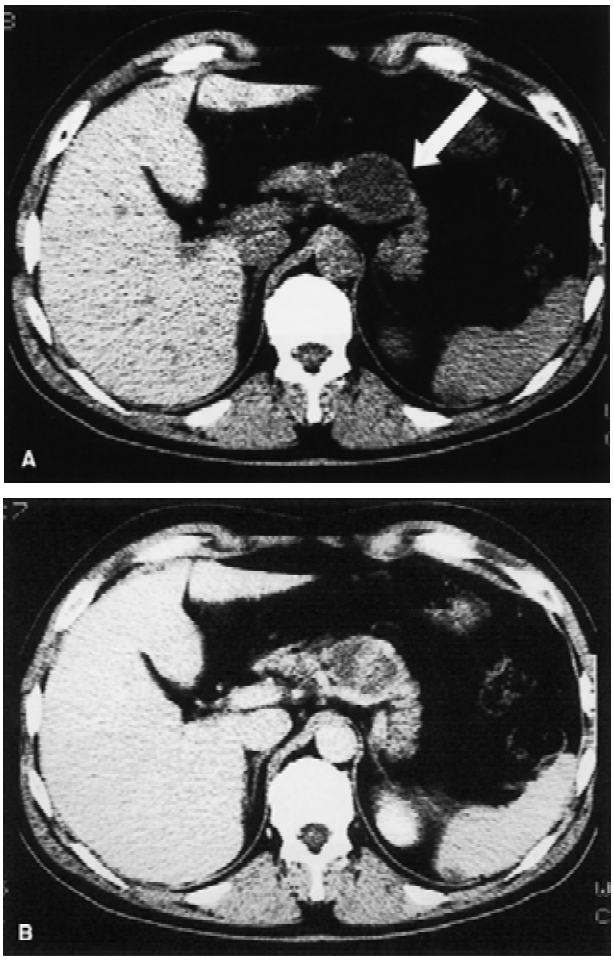

FIG. 7. Benign pancreatic schwannoma. This 47-year-old man had dyspepsia and recurrent right upper quadrant pain. Unenhanced CT scan shows a hypodense round lesion in the body of the pancreas (arrow) with attenuation values between 0 and 10 (**A**). After contrast medium administration, mild to moderate enhancement of the pseudocapsule and of internal septa is seen (**B**). Pathologic examination demonstrated both Antoni type A and type B tissue.

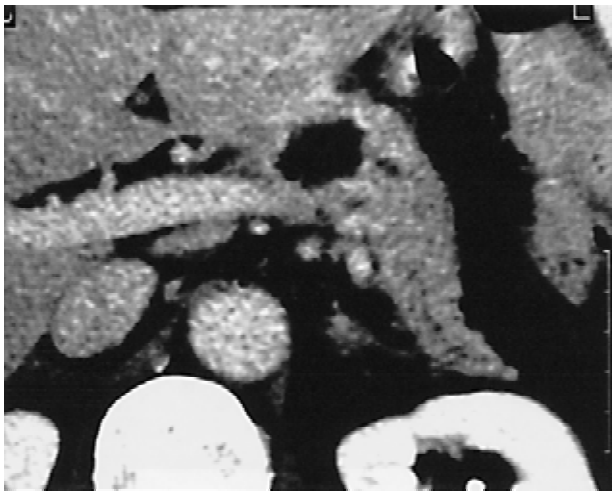

FIG. 8. Benign pancreatic schwannoma. This 63-year-old man had a peptic ulcer medically treated for 2 years and nonspecific upper abdominal symptoms. There is a cystic lesion in the body of the pancreas, with mild contrast enhancement of its borders. No significant enhancement of the central portion is identified. At pathology, the lesion demonstrated a nearly exclusive representation of Antoni type B tissue.

diffusion from celiac plexus involvement in type 1 neurofibromatosis is the most common finding (18) (Fig. 9).

The characteristic CT features of neurofibromas (marked and homogeneous hypodensity, 20–25 HU at baseline, and mild contrast enhancement) have been variously attributed to the high lipid content of Schwann cells, to entrapment of perineural adipose tissue, to adipose metaplasia of fibroblastic cells, to cystic degeneration.

LYMPHOMA

Non-Hodgkin lymphoma (NHL) ensues as an extranodal disease in 20% of cases. The pancreatic localization is rare (<1% of NHL), and the pancreas can be

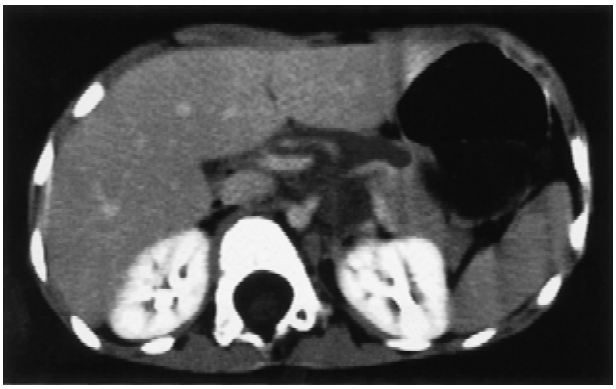

FIG. 9. Plexiform neurofibroma. This 17-year-old child had type 1 neurofibromatosis. A large retroperitoneal mass encasing the celiac axis and involving the body of the pancreas is documented. The pancreatic involvement was confirmed at surgery.

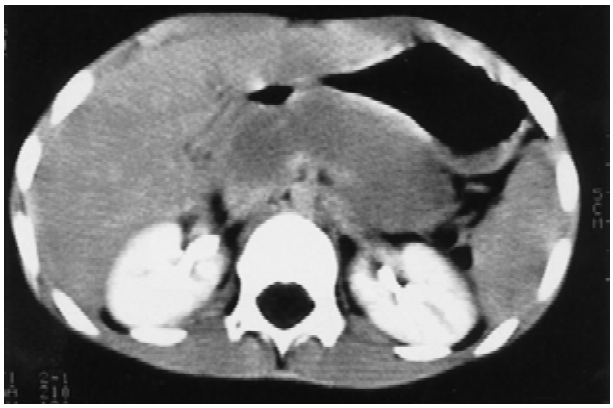

FIG. 10. Burkitt non-Hodgkin lymphoma. In this 12-year-old child, the pancreas is globally enlarged, homogeneous in structure, with mild contrast enhancement. No necrotic changes are documented.

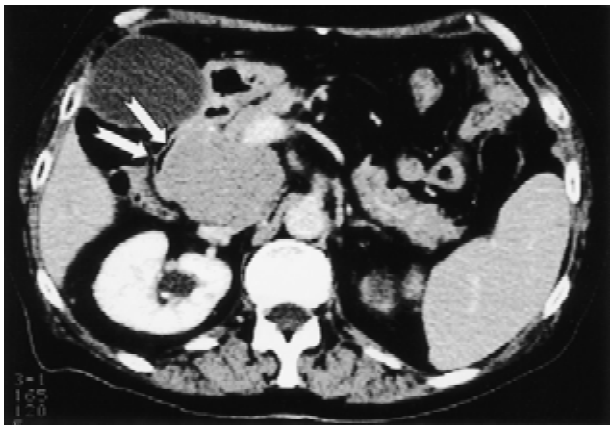

FIG. 11. Immunoblastic primary non-Hodgkin lymphoma of the pancreas. In this 75-year-old man, a large, homogeneous in structure, mildly enhancing mass in the pancreatic head is demonstrated. No necrosis or calcifications are documented. No fat planes are seen with the duodenum (arrows).

involved by systemic lymphoma or rarely may be the primary or predominant site of involvement (19). AIDS-related NHL involves the pancreas more frequently than in the general population.

Two patterns of involvement are described at CT: diffuse, infiltrating lesions that diffusely involve the pancreas, which appears enlarged (Fig. 10), or alternatively focal, well delineated masses (20) (Fig. 11). The lymphomatous structure is typical: homogeneous, hypodense, and hypovascular (19). Necrotic or hemorrhagic changes are rare but less prominent than seen in solid neoplasms. Calcifications are usually rare. Lymphadenopathy (in the retroperitoneal area, particularly below the level of the renal veins) may or may not present

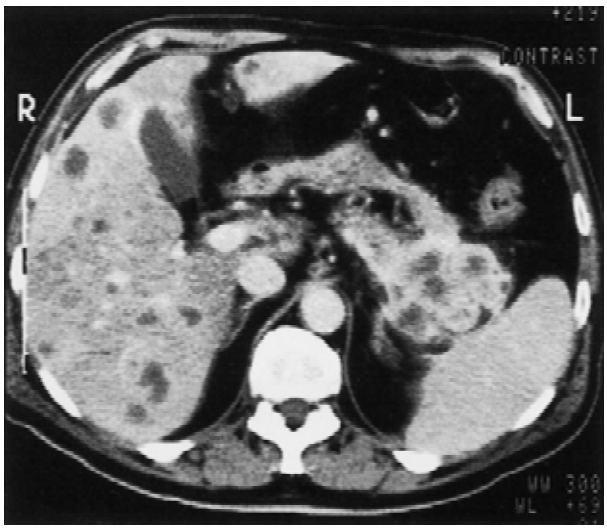

FIG. 13. Pancreatic leiomyosarcoma. This 67-year-old woman had weight loss, abdominal pain, nausea, and occasional vomiting. A large heterogeneous mass is seen in the pancreatic tail. Its structure is hypervascular with multiple necrotic foci. Multiple hepatic metastases with a peripheral rim of contrast enhancement are also documented.

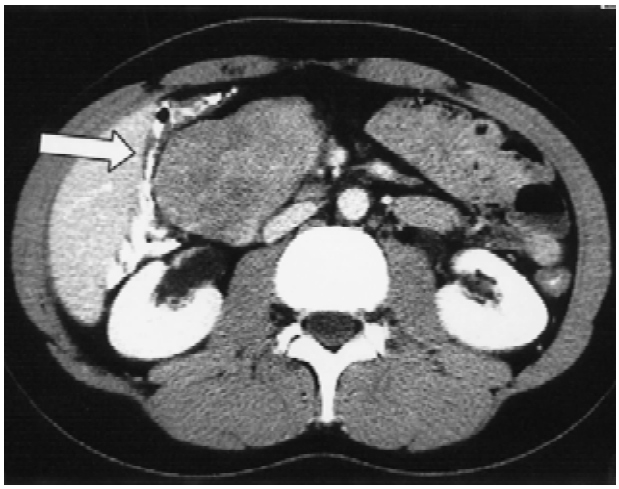

FIG. 12. Pancreatic leiomyosarcoma. This 30-year-old man had abdominal pain, fatigue, dyspepsia, and weight loss. A large roundish mass is identified in the head of the pancreas. The structure is finely heterogeneous, but no significant necrotic component is documented. The duodenum (arrow) is displaced but not infiltrated.

(20). Biliary ductal dilatation, if the lymphoma involves the cephalic area, is a less common complication than in epithelial neoplasms. Secondary pancreatitis is uncommon (20).

Metastatic disease can produce similar findings in case of diffuse involvement, whereas the focal pattern can be seen in several other tumors (19).

SARCOMA

Primary sarcomas of the pancreas are very rare, accounting for a small fraction (~0.6%) of pancreatic malignancies (21). In fact, most malignant tumors of the pancreas with a sarcomatous appearance are represented by anaplastic carcinomas with sarcomatoid differentiation or pancreatic extension of retroperitoneal sarcomas. However, primary undifferentiated sarcomas, fibrosarcomas, malignant fibrous histiocytomas, Kaposi sarcomas, and leiomyosarcomas have been described (22).

Leiomyosarcoma is the histotype least commonly described. It has been postulated that this tumor arises from either the pancreatic duct or blood vessels within the pancreas (21). Sarcomas are highly malignant neoplasms, frequently showing hypervascularity and early metastatic diffusion, particularly to the liver (22).

CT findings can range from a finely heterogeneous solid mass (Fig. 12) to a highly inhomogeneous enhancing lesion with peripheral vascularization and necrotic components (Fig. 13). No significant CT findings permit the differential diagnosis from an adenocarcinoma.

OTHER TUMORS

Sporadic cases of other mesenchymal tumors of the pancreas, both benign (fibroma, leiomyoma, desmoid,

fibrous histiocytoma, hemangioma, and hemangioendothelioma) and malignant (fibrous histiocytoma, hemangiopericytoma, peripheral nerve sheath or neuroectodermal neoplasms, etc.) have been described in literature, but no typical CT findings have been reported.

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REFERENCES

1. Kloppel G, Maillet B. Classification and staging of pancreatic nonendocrine tumors. *Radiol Clin North Am* 1989;27:105–19.
2. Abe H, Kubota K, Noie T, Bandai Y, Makuuchi M. Cystic lymphangioma of the pancreas: a case report with special reference to embryological development. *Am J Gastroenterol* 1997;92:1566–7.
3. Paal E, Thompson LD, Heffes CS. A clinicopathologic and immunohistochemical study of ten pancreatic lymphangiomas and review of the literature. *Cancer* 1998;82:2150–8.
4. Hanelin LG, Schimmel DH. Lymphangioma of the pancreas exhibiting an unusual pattern of calcification. *Radiology* 1977;122: 636.
5. Pandolfo I, Scribano E, Gaeta M, Fiumara F, Longo M. Cystic lymphangioma of the pancreas: CT demonstration. *J Comput Assist Tomogr* 1985;9:209–10.
6. Katz DS, Nardi PM, Hines J, et al. Lipomas of the pancreas. *AJR* 1998;170:1485–7.
7. Katz DS, Hines J, Math KR, et al. Using CT to reveal fat-containing abnormalities of the pancreas. *AJR* 1999;172:393–6.
8. Di Maggio EA, Solcia M, Dore R, et al. Intrapancreatic lipoma: first case diagnosed with CT. *AJR* 1996;167:56–7.
9. Fernandez-Cebrian JM, Carda P, Morales V, Galindo J. Dermoid cyst of the pancreas: a rare neoplasm. *Hepatogastroenterology* 1998;45:1874–5.
10. Jacobs JE, Dinsmore BJ. Mature cystic teratoma of the pancreas: sonographic and CT findings. *AJR* 1993;160:523–4.
11. Kohzaki S, Fukuda T, Fujimoto T, et al. Case report: ciliated foregut cyst of the pancreas mimicking teratomatous tumour. *Br J Radiol* 1994;67:601–4.
12. Hua C, Shu XK, Lei C. Pancreatoblastoma: a histochemical and immunohistochemical analysis. *J Clin Pathol* 1996;49:952–4.
13. Willnow U, Willberg B, Schwamborn D, Korholz D, Gobel U. Pancreatoblastoma in children. Case report and review of the literature. *Eur J Pediatr Surg* 1996;6:369–72.
14. Lee JY, Kim I. CT and US findings of pancreatoblastoma. *J Comput Assist Tomogr* 1996;20:370–4.
15. Feldman L, Philpotts LE, Reinhold C, Duguid WP, Rosenberg L. Pancreatic schwannoma: report of two cases and review of the literature. *Pancreas* 1997;15:99–105.
16. Ferrozzi F, Bova D, Garlaschi G. Pancreatic schwannoma: report of three cases. *Clin Radiol* 1995;50:492–5.
17. Burd DA, Tyagi G, Bader DA. Benign schwannoma of the pancreas. *AJR* 1992;159:675.
18. Imai H, Kobayashi J, Manabe R, Namio H, Ichinona T. A case of neurofibroma located in the retroperitoneum involving the uncinate process of the pancreas. *Gastroenterol Jpn* 1989;24:421–4.
19. Ferrozzi F, Campani R, Garlaschi G, Campodonico F. Extranodal lymphoma: CT findings and differential diagnosis. *Radiol Med* 1997;93:429–41.
20. Van Beers B, Lalonde L, Soyer P, et al. Dynamic CT in pancreatic lymphoma. *J Comput Assist Tomogr* 1993;17:94–7.
21. Zalatnai A, Kovacs M, Flautner L, et al. Pancreatic leiomyosarcoma. Case report with immunohistochemical and flow cytometric studies. *Virchows Arch* 1998;432:469–72.
22. Paciorek ML, Ross GJ. MR imaging of primary pancreatic leiomyosarcoma. *Br J Radiol* 1998;71:561–3.