11 Rare and Secondary Tumors of the Pancreas

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CONTENTS

11.1 Introduction 295
11.2 Histogenesis of Tumors of the Exocrine Pancreas 296
11.3 Rare Pancreatic Tumors of Ductal Cell Origin 296
11.3.1 Mucinous Carcinoma (Adenocarcinoma) 296
11.3.2 Adenosquamous Carcinoma 297
11.3.3 Pleomorphic Giant Cell Carcinoma 298
11.3.4 Giant Cell Carcinoma, Osteoclast-Type 298
11.4 Tumors of the Exocrine Pancreas with an Acinar Cell Origin 298
11.4.1 Acinar Cell Carcinoma 298
11.4.2 Pancreatoblastoma 298
11.5 Tumors of the Exocrine Pancreas of Uncertain Origin 299
11.5.1 Solid and Papillary Epithelial Neoplasm (Solid-Cystic Tumor, Papillary Cystic Tumor) 299
11.5.2 Other Tumors of the Pancreas of Uncertain Origin 300
11.6 Mixed Tumors 300
11.7 Intrapancreatic Lipoma 301
11.8 Pancreatic Plasmocytoma 301
11.9 Pancreatic Schwannoma 301
11.10 Pancreatic Hemangioma 302
11.11 Pancreatic Lymphoma 302
11.11.1 Imaging 303
11.12 Pancreatic Sarcoma 304
11.13 Intrapancreatic Metastases 305
11.13.1 Imaging 306
11.14 Conclusion 307
References 308

11.1 Introduction

The use of ultrasonography (US), helical computed tomography (CT), and magnetic resonance imaging (MRI) with fast sequences and contrast medium administration has improved the visualization of pancreatic tumors (Semelka et al. 1991; Kelekas and Semelka 1997). Indications for the use of endoscopic ultrasonography are now better defined and this technique provides detailed images of the pancreatic and peripancreatic area, and is most useful as a preoperative procedure to improve detection and staging of tumors (Botet and Lightdale 1992; Rösch et al. 1991; Kloppe and Maillet 1989). Although staging of pancreatic neoplasms has improved, and high accuracy for the detection and staging of tumors has been achieved, patient outcome remains uniformly poor (Baytor and Berg 1973). It seems that the main role of imaging is to prove inoperability of patients with tumors to prevent needless postoperative morbidity and mortality. Helical CT has decreased the role of other diagnostic tests like endoscopic retrograde cholangiopancreatography (ERCP) and angiography (Freeny and Lawson 1982; Friedman and Edmonds 1989). The origin of the tumoral cells are also more specifically defined by means of ultrastructural and immunohistochemical investigations, and it is now possible to describe imaging signs characteristic of most of the different histologic types of pancreatic tumor, including some rare ones (Cubilla and Fitzgerald 1979; Morohoshi et al. 1983, 1987). Percutaneous fine-needle biopsy under US or CT guidance enables preoperative diagnosis in numerous cases. Use of fine-needle biopsy can avoid useless laparotomy and reduce hospitalization (Freeny 1988).
11.2 Histogenesis of Tumors of the Exocrine Pancreas

Most (>95%) pancreatic tumors are of epithelial origin deriving from the ductal cell. A few originate from the acinar cell or have a mixed origin. The histogenesis of certain tumors (solid-cystic tumor, anaplastic tumor) remains uncertain. In addition to these tumors of epithelial origin, we give due consideration in this chapter to metastases, nonepithelial tumors, and malignant lymphomas. Cystic tumors and endocrine tumors are described in Chap. 9 (Cubilla and Fitzgerald 1984; Klöppel and Maillet 1989; Morohoshi et al. 1987).

11.3 Rare Pancreatic Tumors of Ductal Cell Origin

Ductal adenocarcinoma is by far the most frequent tumor. Rare ductal neoplasms include mucinous carcinoma, adenosquamous carcinoma, pleomorphic giant cell carcinoma, and giant cell carcinoma osteoclast-type.

11.3.1 Mucinous Carcinoma (Adenocarcinoma)

The clinical presentation is very similar to that of ductal adenocarcinoma. A unique feature of this tumor is that its density may be lower than that of ductal carcinoma; it may also be cystic (Cubilla and Fitzgerald 1984; Mathieu et al. 1989).

Mucinous carcinomas tend to be larger with broad cystic spaces filled with squamous substances bordered by malignant cells. Pseudocystic areas filled with mucin substances appear anechoic or hypoechoic on US, and hypodense on CT. Following chemotherapy, calcifications may appear within the tumor or its metastases. Cytodiagnosis is possible by means of fine-needle aspiration biopsy, but false-negatives are frequent owing to the relative rarity of malignant cells. The excessive mucin production may cause biliary obstruction and renal tubular obstruction in these patients. Pseudomyxoma peritonei can occur (Hertzana et al. 1989).

Mucinous adenocarcinomas are larger and softer than duct cell adenocarcinoma. The mucin-hyper-secreting carcinomas limited to the duct consist of flat or polypoid tumors confined to the main pancreatic duct and its side branches. These tumors have been reported to spread along the duct and frequently invade the parenchyma or extend outside the gland. The pancreatic ducts are dilated and filled with mucin produced by the tumor. Sonography of mucinous adenocarcinoma limited to the duct reveals a diffusely dilated main pancreatic duct filled with mucin of varying degrees of echogenicity. Contrast-enhanced CT may depict enhancing tumor nodules within the ducts. Otherwise, the mucin-filled ducts have a homogeneous or slightly inhomogeneous low attenuation. The parenchyma is generally atrophic (Fig. 11.1a,b).

Contrast-enhanced MRI may reveal large hypointense cystic areas on T1-weighted spin echo sequences. Polypoid nodules are enhanced after injection of gadolinium (Fig. 11.2a,b).

Endoscopic US, is remarkable for its ability to strikingly depict dilated main pancreatic ducts filled