Asymptomatic solid pancreatic hamartoma

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Pancreatic hamartomas occur extremely rarely. They may appear as solid masses or cystic forms and should be regarded as malformations rather than neoplasms [1]. Due to absence of characteristic differential features they may be clinically and radiologically mistaken for a malignancy [2]. Misinterpretation of hamartoma as a true neoplasm may result in unnecessarily aggressive surgery. The authors herein present a case report of solid pancreatic hamartoma and demonstrate its characteristics along with a review of the literature and discuss the problem of potential overtreatment of these patients.

A 69-year-old male patient was presented with a hypoechogenic, asymptomatic pancreatic body mass, incidentally revealed by ultrasonography. The patient gave no medical history of either chronic pancreatitis or other diseases of the pancreas. Pancreatitis and neoplasm (CEA, CA 19-9) markers were within the normal range; pancreatic exocrine and endocrine function was sufficient. Further endosonography scans revealed a 28 mm × 22 mm hypoechogenic tumour-like lesion within the body of the pancreas (Figure 1). Computed tomography scans confirmed a solid tumor with ill-demarcated margin (Figure 2). The patient was qualified for surgical treatment. Intraoperatively, the oval, pancreatic body mass of firm consistency was confirmed. Central pancreatic resection with Roux-en-Y pancreaticojejunostomy to the distal pancreatic remnant was performed. The postoperative period was complicated by leakage from the pancreatojejunostomy. Reoperation with re-suturing of the anastomosis was applied. The remaining postoperative time was uneventful. So far, the patient has been followed up for 55 months and has remained disease-free.

Macroscopically, a firm, solid, whitish, well-circumscribed, encapsulated mass measuring 3 cm in maximum diameter was confirmed. Histologically, the tumour was almost entirely composed of disorderly arranged, well-differentiated endocrine and exocrine pancreatic tissue (Figure 3). The spindle morphology of a small quantity of stroma cells and cystically dilated pancreatic ducts, filled with pancreatic juice, were observed (Figure 4). The tumour was surrounded by adjacent normal pancreatic parenchyma without significant fibrosis or features of chronic inflammation. Insulin, glucagon, chromogranin, somatostatin, amylase, CD34, CD117, S-100 and desmin immunostainings confirmed colocalization of the typi-

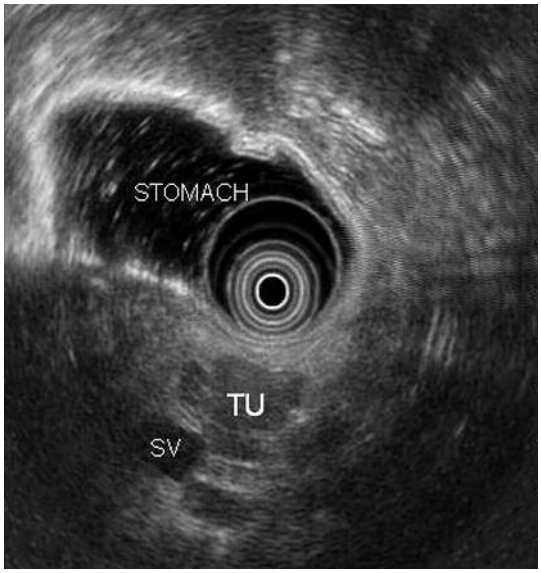

Figure 1. 7.5 MHz radial EUS view of a 28 mm × 22 mm pancreatic corpus tumour-like lesion
TU – tumour, SV – splenic vein

cal pancreatic cells. Surprisingly, immunohistochemical analysis of CD34 and CD117 showed a negative reaction in elongated stoma tumour cells. Therefore, the diagnosis of pancreatic hamartoma was established.

Hamartomas of the pancreas are rarely reported, since only a few cases have been described in the literature. Clinicopathological features of all noted cases are summarized in Table I [2-10]. Some authors have observed that this kind of lesion includes spindle-shaped cells which are immunoreactive for CD34 and CD117 [9]. The descriptive term of these tumours as cellular hamartomas resembling gastrointestinal stromal tumours was suggested. In our case, pathological changes in the cellular shape of stroma cells were observed, although immunohistochemical tests for CD 34 and CD117 were non-contributory, which excludes such a pathological characterization.

The histogenetic concept of hamartomas is enigmatic. Hamartomas of the pancreas were

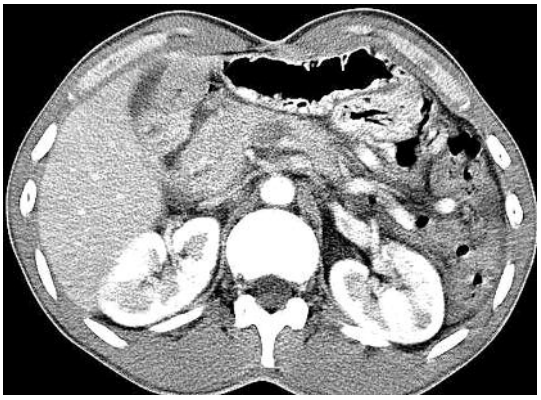

Figure 2. Computed tomography revealing solid tumour within the body of the pancreas

thought to coexist with chronic pancreatitis. On the other hand, Pauser *et al.* excluded such cases from this group. In the authors' opinion, the term of hamartoma should be reserved for asymptomatic patients. The hypothesis that inflammation is one of the factors that induce hamartoma development is not valid, since chronic pancreatitis may just mimic hamartoma lesions lacking acinar cells.

The natural course of hamartoma is not characteristic, as in our described case. Routine laboratory tests and imaging studies are non-contributory in establishment of the final diagnosis. Needle biopsy is suggested to be feasible in differential diagnosis. However, false negative results may occur frequently, and it may be associated with tumour cells seeding in malignancy. Thus, in the authors' opinion, patients with solitary incidentaloma such as hamartoma of the pancreas should not undergo routine pancreatic tumour biopsy.

Currently, preoperative differentiation between hamartomas or other benign tumours and malignancies is very difficult, if not in many cases impossible. As a consequence, it raises a question about treatment of patients with incidentally diagnosed solitary pancreatic tumour of unknown character. In view of the fact that pre-malignant and malig-

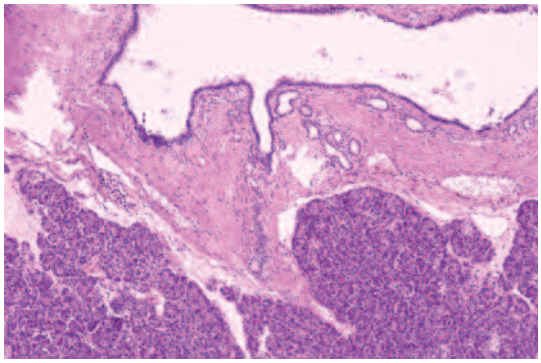

Figure 3. Exocrine part of the pancreas with islets and adipose tissue. HE, 120×

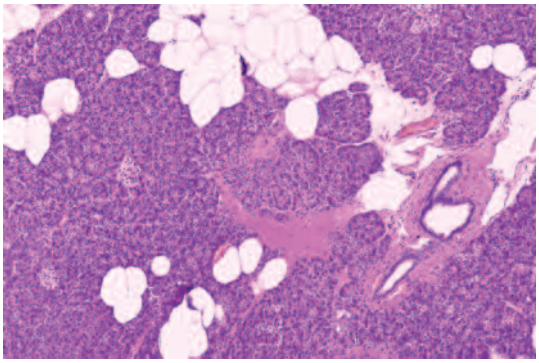

Figure 4. Dilated pancreatic ducts. HE, 120×

Table I. Characteristics of pancreatic hamartomas reported in the literature

Author	Age	Gender	Site	Size [cm]	Treatment
Anthony *et al.* [3]	46 years	M	Head	1.6	Pancreaticoduodenectomy
Anthony *et al.* [3]	35 years	M	Tail	Multiple	Local resection
Anthony *et al.* [3]	58 years	M	Head	1.0	Autopsy
Burt *et al.* [4]	34 weeks	F	Diffuse	11.5	Total pancreatectomy
Flaherty *et al.* [5]	20 months	F	Head	9.0	Local resection
Izbicki *et al.* [6]	25 years	M	Head	10.6	Pancreaticoduodenectomy
Wu *et al.* [7]	39 years	M	Head	8.0	Pancreaticoduodenectomy
McFaul *et al.* [8]	29 years	M	Head	1.0	Pancreaticoduodenectomy
McFaul *et al.* [8]	62 years	M	Head	3.5	Pancreaticoduodenectomy
Pauser *et al.* [2]	36 years	F	Head	7.0	Pancreaticoduodenectomy
Pauser *et al.* [2]	55 years	F	Neck	3.0	Distal pancreatectomy
Pauser *et al.* [9]	51 years	M	Tail	3.0	Local resection
Pauser *et al.* [9]	54 years	F	Body	2.0	Distal pancreatectomy
Nagata *et al.* [10]	58 years	F	Body	1.9	Distal pancreatectomy

nant histology of pancreatic asymptomatic incidentalomas is far more frequent, still surgical excision should be the treatment of choice, even though aggressive surgery may be overtreatment in patients among whom postoperative histopathological examination revealed hamartoma or other benign tumour.

In conclusion, pancreatic hamartoma is an extremely rare medical problem. Misinterpretation of asymptomatic hamartomas as a true neoplasms may result in unnecessarily aggressive surgery. Nevertheless, preoperative diagnostic difficulties may be finally resolved only with histopathological examination of the postoperative specimen.

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