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Case report

Leiomyosarcoma of the pancreas

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Abstract. We report a case of leiomyosarcoma of the pancreas. A spindle cell pattern at light microscopy, immunocytochemical reactivity with desmin, alpha-1-antitrypsin, vimentin and actin and ultrastructural features of smooth muscle differentiation help to establish the diagnosis.

Key words: Pancreas – Leiomyosarcoma

Introduction

Leiomyosarcoma of the pancreas is a very uncommon tumour. As far as we are aware, only 14 cases have been reported some of which were unirresectable. We report a further case, which was completely excised.

Case report

A 71-year-old man presented with a 2 month history of left upper abdominal pain and weight loss (6 kg in the last 2 months). The abdominal CT scan revealed a 4 cm well-defined, solid mass located in the pancreatic body. Coeliac angiography showed that the mass was supplied by branches of the main pancreatic artery. A clinical diagnosis of a tumour of probable endocrine origin was made. Fine-needle aspiration biopsy was performed, but did not help in the diagnosis. In the surgical procedure, the body and the tail of the pancreas were removed.

Pathological findings

Macroscopically a round, whitish, well-demarcated mass, 3.6 cm in diameter was found in the body completely surrounded by pancreatic tissue (Fig. 1).

Microscopically the tumour was fairly cellular and composed of spindle cells of uniform size, arranged in interlacing bundles. The nuclei were elongated, with

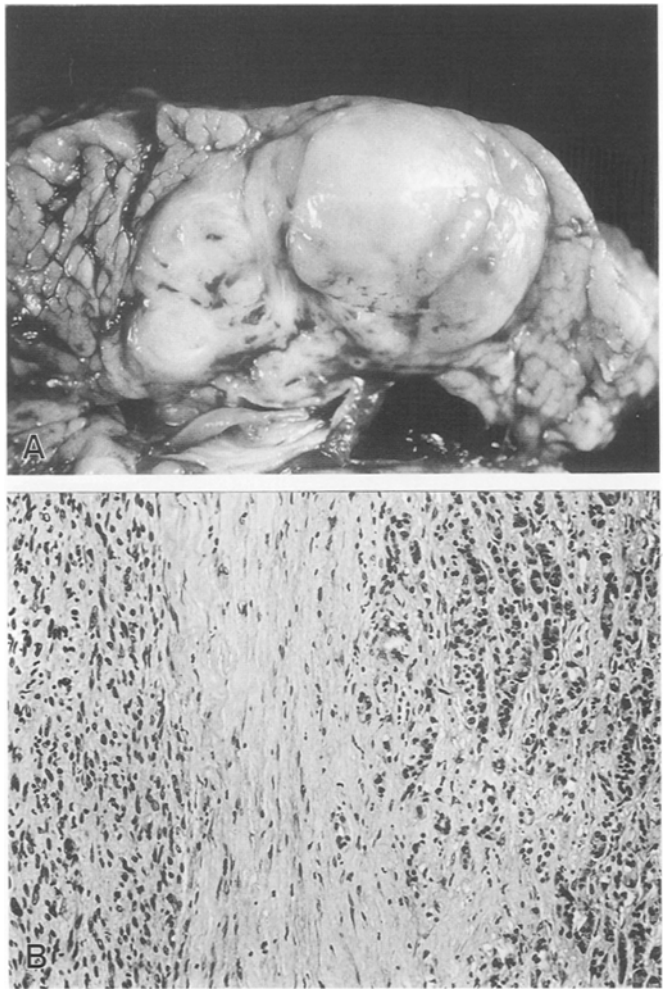

Fig. 1. A Macrograph showing a well-demarcated tumour surrounded by normal appearing pancreas. **B** Micrograph of the edge of the tumour. A pseudocapsule intervenes between the tumour and the atrophic pancreatic tissue. H & E ×150

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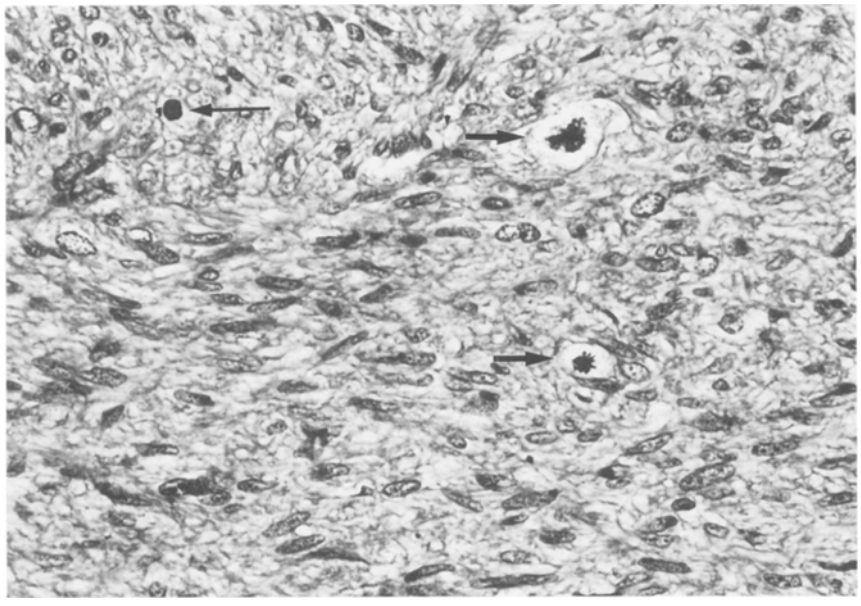

Fig. 2. Tumour field showing marked cellularity and atypia. Three mitotic figures are seen (*arrows*). H & E ×400

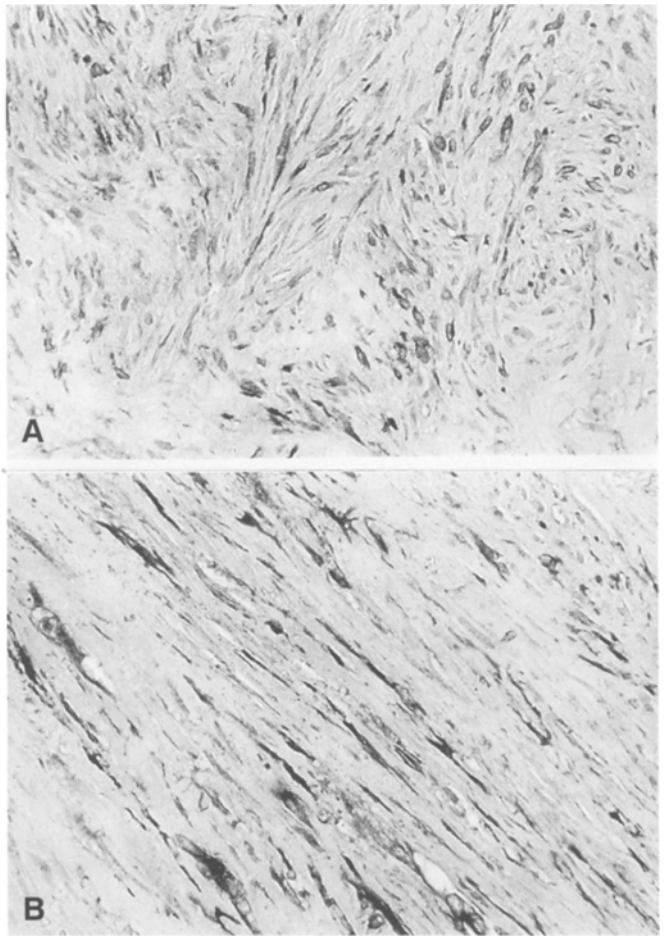

Fig. 3A, B. Tumour cells showing cytoplasmic reactivity to desmin (**A**, ×400), and to vimentin (**B**, ×400)

blunt ends and sharply outlined by an eosinophilic staining cytoplasm in which peripheral myofibrils could be shown up by a phosphotungstic acid haematoxylin stain. Rather poorly differentiated areas with ovoid and polygonal cells of varying size and occasional giant cells were observed. Mitotic figures were frequent (10–14 per 10 HPF), (Fig. 2). Some areas showed concentric arrangements of spindle cells, suggesting a small vessel origin. Small areas of pancreatic tissue composed of atrophic exocrine acini and normal islets of Langerhans were seen trapped inside the tumour. Immunocytochemical studies showed reactivity with desmin (Fig. 3A) (Dako, Dakopatts A/S, 1/200), alpha-1-antitrypsin (Dako, 1/900), vimentin (Fig. 3B) (BioGenex, 1/900) antisera, and focally to muscle specific actin (Dako, 1/500). There was no reactivity to AE3/AE1 keratins (BioGenex, 1/200) nor to CAM 5.2 keratin (BioGenex, prediluted).

Ultrastructurally, the tumour consisted of small cells completely surrounded by a distinct basement membrane. The nucleus was often folded and cytoplasm contained bundles of thin filaments with focal densities along their course. Subsarcolemmal densities in which filaments appeared to be anchored were also seen (Fig. 4). The spleen and lymph nodes were normal. A diagnosis of leiomyosarcoma was established.

Discussion

Leiomyosarcoma of the pancreas is a rare tumour. Fourteen cases have been reported (Table 1), usually as single communications (Becker et al. 1963; Berman and Levine 1956; Carda et al. 1976; Ishikawa et al. 1981; Murata et al. 1990; Nordback et al. 1990; Oyamada et al. 1970; Rodl and Hoffman-Preiss 1988; Ross 1951; and Tulha et al. 1982), although Baylor and Berg (1973) collected

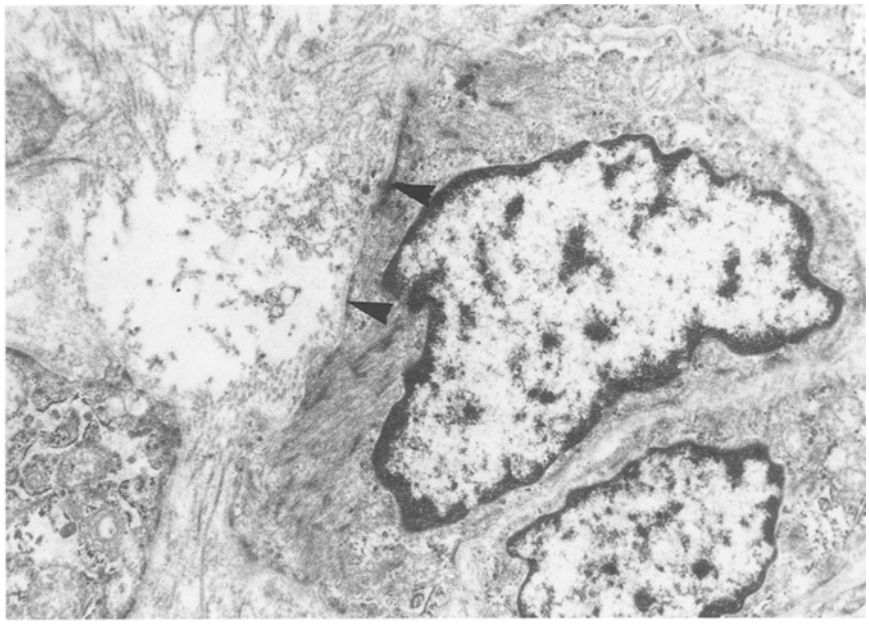

Fig. 4. Electron micrograph of a neoplastic cell surrounded by a basement membrane and showing an indented nucleus. The cytoplasm contains bundles of myofilaments and focal and subsarcolemmal densities (*arrowheads*). ×11375

Table 1. Reported cases of leiomyosarcoma of the pancreas

Author	Year	Age (years)	Sex	Pathology	Treatment
Ross	1951	80	M	Whole pancreas. Unresectable	Autopsy case
Berman	1956	47	M	Head of the pancreas. 5.5 cm	Pancreatoduodenectomy
Feinberg	1957	14	M	Head of the pancreas. 11 cm. Cystic change.	Pancreatoduodenectomy
Becker	1963	–	–	Cystic change	–
Oyamada	1970	47	M	15 cm. Cystic change.	Unresectable
Baylor	1973	51(mean)	M:3, F:2	Local: 1, Regional: 1, Metastatic: 3	–
Carda	1976	56	F	Whole pancreas. Cystic change.	Palliative surgery
Ishikawa	1981	44	M	Metastasis in the liver.	Pancreatoduodenectomy
Tulha	1982	–	–	–	–
Murata	1990	55	F	–	Caudal pancreatectomy
Present case	1992	71	M	Body of the pancreas. 3.6 cm.	Pancreatectomy

a series of 5 cases among 5,057 pancreatic malignant tumours.

Gastric, duodenal, vena cava, and splenic vein leiomyosarcomas have been described involving the pancreas and resembling a primary pancreatic tumour (Nordback et al. 1990). In our case the tumour was completely surrounded by normal pancreatic tissue which strengthens the view that it was of pancreatic origin. The finding of cellular whorls suggests that it might have originated in the smooth muscle of the walls of pancreatic vessels.

Table 1 shows the clinicopathological data of the 14 reported cases of leiomyosarcoma of the pancreas. Most patients were middle-aged and the tumour was more frequent in males than in females. At the time of diagnosis the tumours were large and usually cystic. The smallest was of 5.5 cm diameter and showed no cystic change, which suggests that this occurs late in development (Berman and Levine 1956). The tumour we described is even smaller, suggesting that it was diagnosed at an early stage.

To lay down criteria for malignancy in smooth muscle tumours is difficult (Enzinger and Weiss 1988). Although some features such as size, cellularity, atypia, necrosis, and the mitotic count have been proposed, only the latter is accurate and reproducible in predicting metastasis (Enzinger and Weiss 1988). Ranchod and Kempson (1977) considers that a mitotic count above 5 per 10 HPF in a retroperitoneal leiomyosarcoma should be considered malignant, since most such patients die of the tumour (Enzinger and Weiss 1988). Nevertheless, it may be considered that extra-uterine leiomyosarcomas may exhibit very few mitosis (Purdy Stout and Lattes 1967). In our case, the tumour has not metastatized to date, although its mitotic count suggest a malignant nature.

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