

INFLAMMATORY MYOFIBROBLASTIC TUMOR OF THE PANCREATIC HEAD

L. Lacoste, Ch. Galant², J.-F. Gigot³, B. Lacoste¹, L. Annet¹

Inflammatory myofibroblastic tumors are rare, especially in the pancreas. It is sometimes difficult to obtain a definitive diagnosis with radiological imaging and there is not yet consensus about treatment. We report a case of a 56-year-old man with recurrent abdominal pain particularly in the right upper quadrant without other symptoms. The imaging results showed a pancreatic hypovascularized mass with stenosis of the main pancreatic duct and the common bile duct without metastasis. The FDG PET scanner showed two hypermetabolic foci in the head of the pancreas. The biopsies of the mass were not diagnostic. The therapy adopted was Whipple's pancreaticoduodenectomy with a histological diagnosis of the inflammatory myofibroblastic tumor.

Key-word: Pancreas, neoplasms.

Inflammatory myofibroblastic tumors (IMT), previously known as inflammatory pseudotumors, are rare lesions of the soft tissue, appearing most frequently in the young and for which the origin is unknown (1). They were initially observed in the lungs but they can also be found in the mesentery, the omentum, the abdominal organs, the bladder or the orbit.

Today, only 28 cases of pancreatic IMT have been reported in the literature (2). This pathology resembles both clinically and radiologically an adenocarcinoma of the pancreas which causes real problems in reaching a differential diagnosis. We present a new case of inflammatory myofibroblastic tumor of the pancreas covering all aspects from diagnosis to treatment.

Case report

A 56-year-old man was hospitalized several times in the gastroenterology department for recurrent localized abdominal pain particularly in the right upper quadrant, sometimes accompanied by nausea. There were no other symptoms and the clinical exam showed no particularities. Blood tests revealed an increase in transaminases, lipases, amylases and CA 19.9.

CT and MRI showed a heterogeneous solid hypervascularized mass that was slightly hypointense on T2, hypointense on T1, hypovascularized, in the head of the pancreas with stenosis of the cephalic Wirsung duct and of the intra-pancreatic portion of the common bile duct with upstream dilatation (Double Duct Sign) and visibility of the secondary

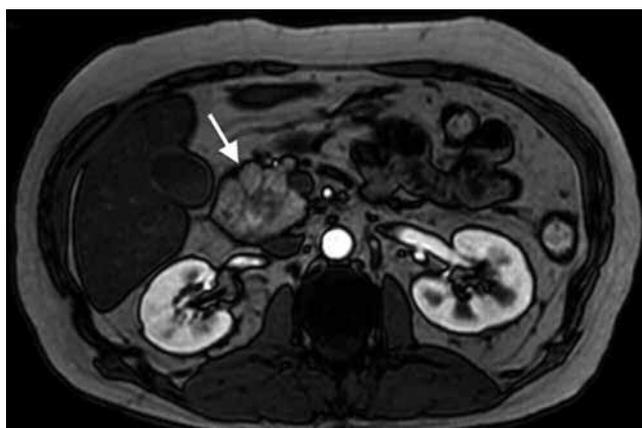


Fig. 1. — Axial T1-weighted gadolinium contrast-enhanced MR image shows a hypovascularized mass in the head of the pancreas (arrow).

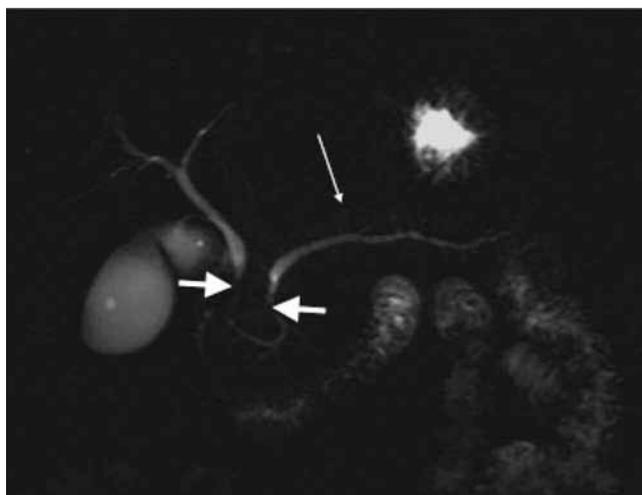


Fig. 2. — MR cholangiopancreaticography shows a stenosis of the main pancreatic duct and the common bile duct (thick arrows) «Double Duct Sign» with an upstream dilatation and visible secondary pancreatic ducts (thin arrow).

pancreatic ducts (Fig. 1, 2). There was no local adenopathy and no lesion of the liver to suggest metastasis.

The endoscopic ultrasonography showed a chronic pancreatopathy of the complete pancreas and con-

From: 1. Department of Medical Imaging, 2. Department of Anatomopathology, 3. Department of Surgery, UCL Saint Luc, Catholic University of Louvain, Brussels, Belgium.

Address for correspondence: Dr L. Lacoste, Department of Medical Imaging, UCL Saint Luc, Catholic University of Louvain, Avenue Hippocrate 10, B-1200 Brussels, Belgium.

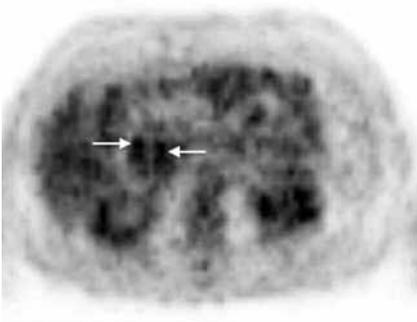


Fig. 3. — Axial FDG PET image shows a heterogeneous pancreatic head with two hypermetabolic foci (arrows).

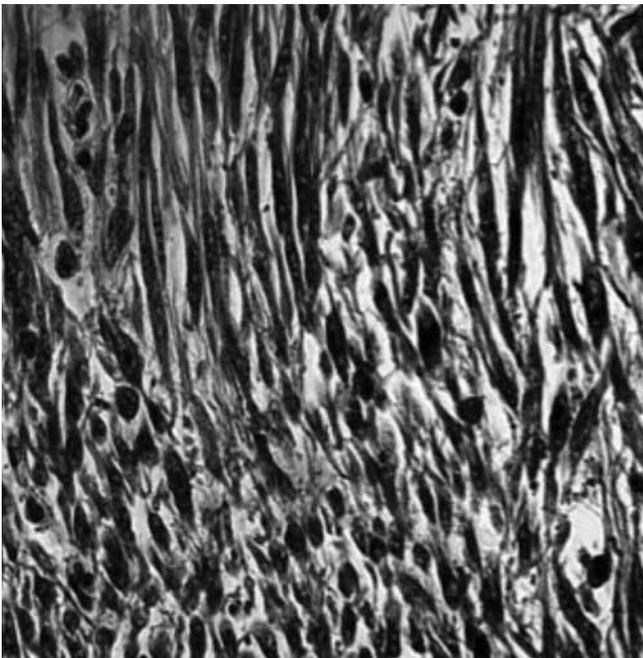


Fig. 4. — Tumor of the pancreatic head. Spindle cell proliferation made of myofibroblasts accompanied by a prominent inflammatory infiltrate consisting of plasma cells, lymphocytes, and eosinophils (original magnification $\times 20$).

firmed the presence of an expansive mass in the cephalic region of the pancreas. Biopsies were realized at the same time but were non conclusive.

A FDG PET scan was realized, showing a heterogeneous cephalic pancreatic region similar to what can be observed in chronic pancreatitis, but with the presence of two distinct foci of which the metabolic rates were comparable with active tumorous phenomena but also with acute inflammatory pathologies (Fig. 3).

Due to the difficulty in making an accurate diagnosis and eliminating a malignant tumor, it was decided in the multidisciplinary meeting to

undertake Whipple's pancreaticoduodenectomy. The operation went ahead without complications.

The macroscopic analysis of the specimen revealed a white solid mass with stenosis of the Wirsung duct. Histopathological analysis showed a proliferation of myofibroblasts accompanied by an important infiltration of chronic inflammatory cells (Fig. 4). The immunohistochemical analysis was positive for alpha smooth muscle actin (Fig. 5) and for desmin (Fig. 6).

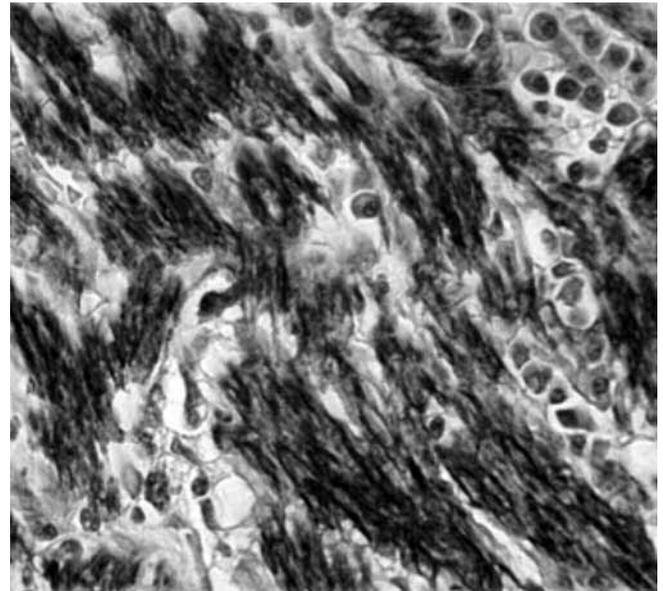


Fig. 5. — Strong immunoreactivity of most of the spindle cells for alpha smooth muscle actin (original magnification $\times 40$).

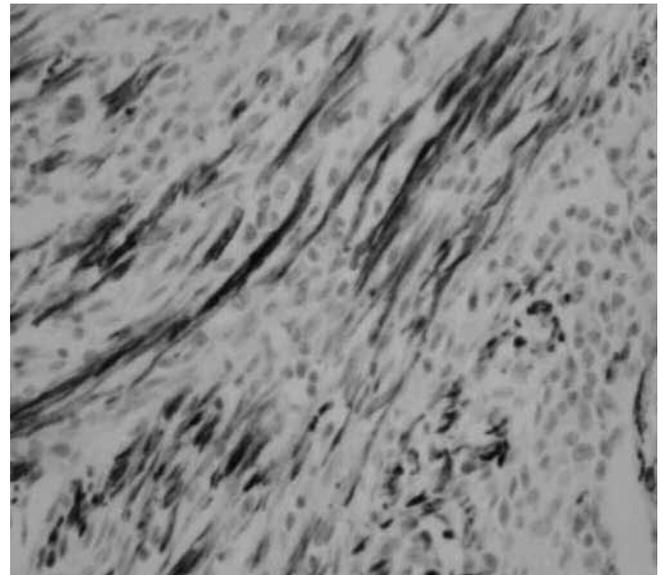


Fig. 6. — Strong immunoreactivity of some myofibroblasts for desmin (original magnification $\times 40$).

The surgical follow-up was uneventful and now the patient is doing well.

Discussion

The terminology of inflammatory myofibroblastic tumor is now generally accepted for the majority of lesions formerly named: inflammatory pseudotumor, plasma cell granuloma, omental-mesenteric myxoid hamartoma, and inflammatory fibrosarcoma (3, 4).

The inflammatory myofibroblastic tumors, often considered as a neoplastic entity, are part of the group of solid mesenchymal tumors characterized by the proliferation of myofibroblasts associated with a variety of inflammatory cells such as plasma cells, lymphocytes and eosinophils (3, 5).

They are rare tumors that can appear at any age but more frequently in the first two decades of life (1, 3-5) and in women (3).

They were initially observed in the lungs and can also be found in the mesentery, the omentum and in some rare cases in the abdominal organs, the bladder, the orbit, the head, the cervical region and the salivary glands (4-6).

The origin is still undetermined. Considered historically as reactionary inflammatory lesions following a trauma, a surgery, or an infection or considered as a paraneoplastic lesion, recent molecular and chromosomal studies lean more towards a neoplastic lesion rather than a reactionary lesion (1, 4).

IMTs of the pancreas are often incidentally diagnosed (5). When the tumors are symptomatic, one observes, as shown by Surakit et al. (1, 7), mainly abdominal pain (64%), weight loss (44%), jaundice (40%), a palpable mass (28%) and in rare cases, anemia, fatigue or sudden diabetes.

Imaging findings highlight a solid well-defined pancreatic mass, which can be multilobular or infiltrative, vascularized or not (5), of which the size is usually between five and ten centimeters (3).

Radiology also allows one to identify local complications and enables staging. However, imaging does not always allow to make a differential diagnosis with adenocarcinoma of the pancreas, endocrine tumors of the pancreas, auto-immune pancreatitis, metastasis and lymphoma.

Histologically, IMTs are essentially cellular, composed of an important

proliferation of fascicular myofibroblasts accompanied by a voluminous infiltration of chronic inflammatory cells particularly lymphoplasmocytary. The mitotic rates are variable and sometimes calcifications or necrosis can be found (3).

On immunohistochemistry, these lesions are generally positive for alpha smooth muscle actin as well as desmin and keratin. In 30 to 40% of the cases, and especially in the child, molecular genetic and cytogenetic analysis has revealed ALK gene rearrangements/translocations causing an overexpression of ALK protein which can be detected by specific immunohistochemistry (3).

Although, the risk of malignant transformation or metastasis has been described by certain teams, it remains low (< 5%) (1, 3, 5).

Given the difficulty in obtaining a definitive diagnosis based on biopsies and radiological exams, surgery with complete excision of the lesion associated with a clinical follow-up remains the primary therapeutic option (4, 5, 8), despite there being no real consensus regarding the treatment of IMT of the pancreas (1, 4). When the lesion cannot be excised, the patient can be treated with chemotherapy or radiotherapy and/or with medication but the latter is not clearly defined (1, 2).

Some authors obtain results either with corticoid medication or with an anti-inflammatory medical treatment, with or without radiotherapy (2, 4, 8).

The prognosis for this tumor is generally favorable (1, 6). There is a risk of up to 25% of recurrence post surgery, namely when the excision is incomplete (5).

Conclusion

Inflammatory myofibroblastic tumors of the pancreas are rare. The prognosis is usually favorable. However, there are currently no criteria for predicting the evolution.

Therefore, treatment must be decided case by case in function of the surgical risks.

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