Isolated Pancreatic Metastasis of Extremity Myxoid Liposarcoma: Report of a Case

Fabio Carboni1, Giuseppe Maria Ettorre1, Riccardo Lorusso1, Pasquale Lepiane1, Roberto Santoro1, Pietro Mancini1, Francesco Maria Di Matteo2 and Eugenio Santoro1

1Department of Digestive Surgery and Liver Transplantation, Regina Elena Cancer Institute, Rome and 2Department of Digestive Disease, Campus Biomedico University, Rome, Italy

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Myxoid liposarcoma (ML) has a high predilection for extrapulmonary sites of metastases, including intra-abdominal metastases, but pancreatic involvement is extremely rare. Here, we report the case of a 66-year-old male patient, who underwent pancreaticoduodenectomy for isolated pancreatic metastasis of ML of the left lower extremity that had been excised 6 years before. Completion pancreatectomy was necessitated afterwards for a delayed haemorrhage associated with pancreatic fistula. Currently the patient is alive with no evidence of disease. Highly selected patients with isolated pancreatic metastasis of soft tissue sarcoma may benefit from a curative surgical resection.

Key words: myxoid liposarcoma – pancreatic metastasis – diagnosis – surgery

INTRODUCTION

Even though myxoid liposarcoma (ML) has a propensity to metastasize to extrapulmonary sites, pancreatic metastases are extremely rare (1–5). In such cases, an accurate diagnosis is mandatory to select the appropriate treatment, and surgical resection may increase the survival rate in highly selected patients (6–8). We report the case of a patient submitted to pancreaticoduodenectomy (PD) for isolated pancreatic metastasis of ML of the left lower extremity that had been excised 6 years before.

CASE REPORT

A 66-year-old male was admitted to our department for a pancreatic head lesion in August 2005. At the time of admission, he was completely asymptomatic. History revealed that 6 years before he underwent surgical excision of an ML of the left lower knee, without any adjuvant treatment. Tumour size was 6 cm, deeply situated (T2b), no regional lymph node involvement (N0), intermediate grade (G2), stage IIA, according to the AJCC staging (9). Histological tumour grade evaluated according to the FNCLCC system (10) also showed a G2 lesion. Three years later he developed a local recurrence of the disease and underwent re-excision with adjuvant radiotherapy at a total dose of 61.4 Gy. During the follow-up he had no evidence of recurrent disease.

A helical computed tomography (CT) scan showed in the pancreatic head a 3 cm hypodense mass (Fig. 1). Magnetic resonance cholangiopancreatography (MRCP) showed a 2.9 cm mass hypointense in T1 and hyperintense in T2-weighted sequences in the pancreatic head, with normal common bile duct (Fig. 2). Endoscopic ultrasonography (EUS) revealed an hypoechoic lesion (17.2 × 30 mm) in the pancreatic head without abnormalities in the Wirsung’s duct and the common bile duct, and no enlarged nodes in the pancreas. During the procedure, a fine-needle aspiration (FNA) was performed, and cytology revealed a lipomatous neoplasm. Serum tumour marker levels (CEA, Ca 19-9, Ca 72-4) were within the normal range. An 18-fluorodeoxyglucose positron emission tomography (18-FDG PET) was negative.

The presumed diagnosis was isolated pancreatic metastasis from liposarcoma, and a surgical resection was scheduled. At laparotomy, a pancreatic head lesion was found, with no evidence of liver or peritoneal involvement, and therefore the patient underwent standard PD. Histological examination showed an ML grade 2 thoroughly contained within the pancreas (Fig. 3). Immunohistochemical analysis was positive...
for S-100 protein, and no round-cell components were found. Slides of the extremity ML previously excised were carefully reviewed, and cytological features of the primary lesion were found to be identical with the pancreatic metastasis.

On the fourth postoperative day a leakage of the pancreatico-gastric anastomosis was discovered, which was conservatively treated. On the 25th postoperative day a delayed haemorrhage (DH) occurred. Since the patient was haemodynamically stable, a transarterial selective embolization of the splenic artery was performed, following the protocol described previously (11). Three days later, however, a new massive DH occurred and an emergency operation was performed with completion pancreatectomy, toileting of the abdominal cavity and suture of the gastric defect. The patient was discharged 1 month later with insulin therapy for diabetes and pancreatic enzyme supplementation. Currently, he is alive with no evidence of disease after 6 months.

DISCUSSION

Liposarcomas are one of the most common histologic types of soft tissue sarcoma of the extremities (1–4). Several subtypes have been identified, of which the myxoid one is among the most prevalent. ML is described as being of low grade with low metastatic potential, similar to well-differentiated forms and give metastatic disease in 29–32% of cases, with a median time to recurrence of 24 months (1–3). Moreover, they show different patterns of metastatic spread, in comparison with other sarcomas, with a propensity for extrapulmonary sites, including retroperitoneum, abdominal wall and cavity (1–5). According to most authors, patients with a history of resected ML should undergo abdomino-pelvic CT scan along with chest radiographs as part of their follow-up evaluation (1–5). Multimodality treatment, including surgical resection when feasible, may improve the prognosis in these patients. In a recently published series, abdominal metastasectomy was associated with slight improved median survival (33 versus 8 months for unresected patients) (4). However, long survival rates can be achieved if the patients are submitted to curative (R0) resection only (1,2,4,5). Pancreatic metastases are extremely rare but an aggressive surgical approach may be recommended for selected patients with isolated lesions because it could be the only chance of cure. To our knowledge, this is the first report of resected isolated pancreatic metastasis from extremity ML.

Autopitc studies have reported an incidence of metastases to the pancreas between 3 and 11% (8). Metastatic involvement of the pancreas can be limited or diffuse to the whole gland. In a
recent review, 64.7% of the lesions were solitary and mostly located in the head of the gland (8). They are potentially resectable in 0.4–3% of patients, and PD represents a very rare indication, accounting for <2% of all pancreatic malignant tumours surgically treated (7,8).

In most cases, differential diagnosis with primary tumours is difficult. Presenting symptoms and signs are similar, which include jaundice, pancreatitis, pain, weight loss, duodenal obstruction and upper gastrointestinal bleeding (6–8,12). However, the lesion may also be completely asymptomatic and incidentally discovered in a high percentage of patients (6,7), as in our case. Although rare, metastatic disease should always be considered in patients with isolated pancreatic lesions and a history of previous malignancy, including soft tissue sarcomas, regardless of the disease-free interval (4–7,12). An accurate diagnosis is mandatory, because surgical resection may offer long-term survival in highly selected patients.

As far as diagnosis is concerned, at CT scan large metastatic lesions appear hypodense but small lesions may appear isodense. After contrast injection, there is a peripheral rim of enhancement and a central area of attenuation is visible (8,13,14). At MRI they usually appear hypointense on T1 images and hyperintense in T2 (13). At EUS the tumour can appear hypo-isoechoic with some Wirsung abnormalities. Eighty-eight per cent of metastases are solid, hypoechoic with well-defined borders, and up to 17% can be discovered by EUS after negative CT scan (15). Since in the presence of a solitary pancreatic lesion imaging alone is not able to accurately differentiate between metastases and other lesions, EUS-FNA biopsy is a safe and non-invasive diagnostic method with high accuracy rates, which may be helpful in confirming the diagnosis (15).

Indications for pancreatic resections of metastases have not been clearly defined. As for primary tumours, resections performed in high volume centres are safe and PD is associated with low postoperative mortality and acceptable morbidity rates (7,12,16). Long survival periods are reported after curative resection in selected cases, especially after long disease-free interval following the primary operation, and patients show a good quality of life (6–8).

In conclusion, highly selected patients with isolated pancreatic metastasis of soft tissue sarcoma may benefit from a curative surgical resection.

References