Case reports of interest

Secondary extramedullary plasmacytoma involving the pancreas

Shizuhiro Hirata1, Koji Yamaguchi1, Sachiko Bandai1, Akihiko Izumo2, Kazuo Chiiwa1 and Masao Tanaka1

1 Department of Surgery and Oncology, Graduate School of Medical Sciences, Kyushu University, Fukuoka 812-8582, Japan
2 Department of Anatomic Pathology and Pathological Sciences, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

Abstract Extramedullary plasmacytoma is a rare variant of plasma cell tumor involving organs outside the bone marrow. The vast majority of extramedullary plasmacytomas present as a secondary tumor of systemic myelomatosis of the bone marrow. We experienced a patient with extramedullary plasmacytomas of the head and tail of the pancreas presenting as secondary masses from extramedullary plasmacytoma of the maxillary sinus that had been treated 5 years previously. A 38-year-old Japanese man had undergone radiation therapy for an extramedullary plasmacytoma of the maxillary sinus 5 years before the current presentation. He experienced severe upper abdominal pain in November 1999, when laboratory data showed elevation of the serum amylase level. Computed tomography showed two isodensity masses, in the head and tail of the pancreas. Angiography showed two hypervascular masses, one in the head and the other in the tail of the pancreas, and encasement of the portal vein trunk junction. Laparotomy was performed, with the tentative diagnosis of extramedullary plasmacytoma of the pancreas, in order to obtain a definite diagnosis. Intraoperative biopsy revealed that the two pancreatic masses were extramedullary plasmacytomas. External radiation therapy was performed after the operation. When a pancreatic mass is noticed in patients with a history of plasmacytoma, secondary extramedullary plasmacytoma of the pancreas should be considered as a differential diagnosis.

Key words Extramedullary plasmacytoma · Pancreas

Introduction

Plasmacytoma usually affects the bone marrow, but extramedullary plasmacytoma involves organs outside the bone marrow, the incidence being 5% of all plasma cell neoplasms. Extramedullary plasmacytoma is mainly a secondary tumor of multiple myeloma of the bone marrow, and primary extramedullary plasmacytoma is rarely seen. The most common site of primary extramedullary plasmacytoma is the nasal cavity. According to an English-language literature review of 18 cases of extramedullary plasmacytoma of the pancreas, the vast majority of pancreatic plasmacytomas present as a mass secondary to plasmacytoma of the bone marrow or as an extramedullary plasmacytoma of the nasal cavity.1–3 We, herein, report a patient with secondary extramedullary plasmacytoma of the pancreas that occurred 5 years after complete remission of extramedullary plasmacytoma of the nasal cavity by radiation therapy, and we briefly discuss the clinical implications.

Case report

A 33-year-old Japanese man noticed a maxillary sinus tumor in September 1994. The diagnosis of a biopsy specimen of the tumor was extramedullary plasmacytoma. Whole body examinations, including bone scintigraphy and bone marrow tap, were negative for plasmacytoma. He was given 4000 rads of local irradiation to the maxillary area, and the tumor disappeared. He had been doing well until November 1999, when he experienced severe upper abdominal pain and laboratory data showed elevation of the serum amylase level. Computed tomography showed two isodensity masses, in the head and tail of the pancreas. Angiography showed two hypervascular masses, one in the head and the other in the tail of the pancreas, and encasement of the portal vein trunk junction. Laparotomy was performed, with the tentative diagnosis of extramedullary plasmacytoma of the pancreas, in order to obtain a definite diagnosis. Intraoperative biopsy revealed that the two pancreatic masses were extramedullary plasmacytomas. External radiation therapy was performed after the operation. When a pancreatic mass is noticed in patients with a history of plasmacytoma, secondary extramedullary plasmacytoma of the pancreas should be considered as a differential diagnosis.

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chemistry was within normal limits, except for mild elevation of Ig-G (1541 mg/dl) and gamma globulin (20.2%), showing a monoclonal spike of the Ig-G-k. Bone marrow tap did not show proliferation of plasma cells and bone scintigraphy showed no abnormal accumulation. Computed tomography (CT) revealed the presence of two homogeneous masses, in the head and the tail of the pancreas. The pancreatic duct was moderately dilated and the biliary tract was not dilated. The duodenal loop was displaced to the right by the cystic mass in the head of the pancreas (Fig. 1). Hypotonic duodenography showed a widening of the duodenal loop, and a submucosal mass with a smooth surface protruding from the medial aspect of the second portion of the duodenum (Fig. 2). Angiography showed two hypervascular masses, one in the head and one in the tail of the pancreas (Fig. 3), and encasement of the portal vein trunk. Endoscopic retrograde cholangiopancreatography showed displacement and stenosis of the pancreatic duct in the head of the pancreas (Fig. 4). Laparotomy was performed, with the tentative diagnosis of extramedullary plasmacytoma of the pancreas, to obtain a definite diagnosis. The large mass almost replaced the head of the pancreas, and the other was located in the tail of pancreas. Needle biopsy specimens of the two pancreatic masses revealed sheets of atypical plasma cells of various sizes and shapes, showing binucleation and abnormal mitotic figures (Fig. 5). The diagnosis was extramedullary plasmacytomas of the pancreas. The patient underwent postoperative external radiation therapy to the two pancreatic masses. After the external radiation therapy (50 Gy), CT showed shrinkage of the pancreatic masses (Fig. 6). The patient is doing well 1 year after the treatment without any signs of exacerbation or recurrence.

**Discussion**

Extradomedullary plasmacytoma is a rare plasma cell malignancy, the incidence being only 3%–4% of plasma cell malignancies. The diagnosis of primary extradomedullary plasmacytoma requires the demonstration of an extraosseous myelomatous mass without involvement of the bone marrow (plasma cells, fewer than 5%) and...