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Biliary Obstruction Secondary to an Extramedullary Plasmacytoma of the Pancreas

CASE REPORT

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ABSTRACT

Extramedullary plasmacytoma (EMP) is an uncommon neoplastic disorder arising from B-cell lymphocytes and comprises 3% of the plasma cell neoplasms. Ninety percent of the cases are encountered in the head and neck regions. The involvement of the pancreas is rare and generally develops as secondary multiple myeloma (MM). To date, four cases of EMP of the pancreas, which developed as secondary solitary bone plasmacytoma, have been reported in the literature. We decided to present this rare case, which was diagnosed and treated at our clinic.

Keywords: Extramedullary plasmacytoma, pancreas, obstructive jaundice

INTRODUCTION

Extramedullary plasmacytoma (EMP) represent an uncommon variant of the plasma cell tumor involving organs outside the bone marrow. They are typically identified after the diagnosis of multiple myeloma (MM) (1). The most common sites for EMP are the upper respiratory tract, followed by the spleen, lymph nodes, and liver. The pancreas is rarely infiltrated by plasma cells as an EMP, and obstructive jaundice is also rarely diagnosed as a result of plasma cell neoplasms (2). Only 25 cases have been reported in the English literature to date (3). We reported a case of pancreatic plasmacytoma presenting with obstructive jaundice. The patient had a previous solitary bone plasmacytoma. Following this, the patient presented with obstructive jaundice and a pancreatic mass. Abdominal computed tomography (CT) revealed a 9 cm mass in the head of the pancreas, and histological examination showed multiple sheets of atypical plasma cells. Concomitant external radiation and steroid therapy was performed after the diagnosis of pancreatic plasmacytoma. When a pancreatic mass is observed in patients with a history of plasmacytoma, secondary extramedullary plasmacytoma of the pancreas should be considered as a differential diagnosis.

CASE REPORT

A 68-year-old man was diagnosed with a solitary bone plasmocytoma of the sternum in May 2010. The patient was successfully treated with radiotherapy (total dose 50 Gy) and a six course regime of chemotherapy (vincristine, adriablastin, and dexamethasone), leading to complete remission. Post-therapy positron emission tomography (PET)-CT showed a markedly reduced mass in the sternum. He was admitted because of jaundice, nausea, and vomiting 6 months after chemoradiotherapy. On admission, physical examination revealed icteric sclerae and mild epigastric pain. Laboratory data were as follows: aspartate aminotransferase (AST), 72 U/L (normal range, 0–35); alanine transferase (ALT), 65 U/L (10–49); alkaline phosphatase, 190 U/L (45–129); gamma-glutamyl transferase, 292 (0–73); total bilirubin, 22 mg/dL (0–1.1); direct bilirubin, 16.4 mg/dL (0–0.4); total protein, 6.9 g/dL; albumin, 3.8 g/dL; calcium, 8.5 g/dL; Ig A, 180 mg/dL (45–380); Ig G, 781 mg/dL (700–1600); and Ig M, 40 mg/dL (50–301). Contrast-enhanced abdominal CT showed a 9 cm sized, well-defined, hypodense mass in the head of the pancreas and intra- and extrahepatic bile duct dilatation (Figure 1). Repeated PET-CT revealed a mass of intense fluorodeoxyglucose (FDG) uptake in the head of the pancreas and an osteosclerotic lesion of the sternum (Figure 2).

Percutaneous transhepatic cholangiogram confirmed stenosis of the common bile duct by an extrinsic mass. For the relief of obstruction symptoms, external and internal biliary drainage was inserted. Fine-needle pancreatic mass biopsy under ultrasonography (US) guidance was performed and it was diagnosed as plasmacytoma (Figure 3). Repeat bone marrow aspiration and biopsy results were normal. Serum and urine immunofixation results were normal. The results of the test to detect urine Bence Jones protein were negative. The final diagnosis of EMP of pancreas secondary SBP was made. Although biliary stent placement gradually increased direct

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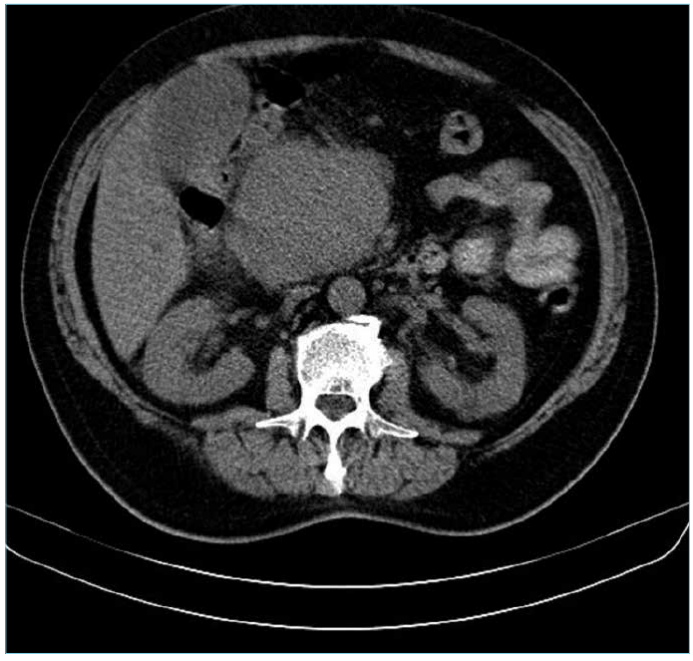

Figure 1. Contrast-enhanced CT image demonstrates a large homogeneously enhancing mass in the region of the pancreatic head

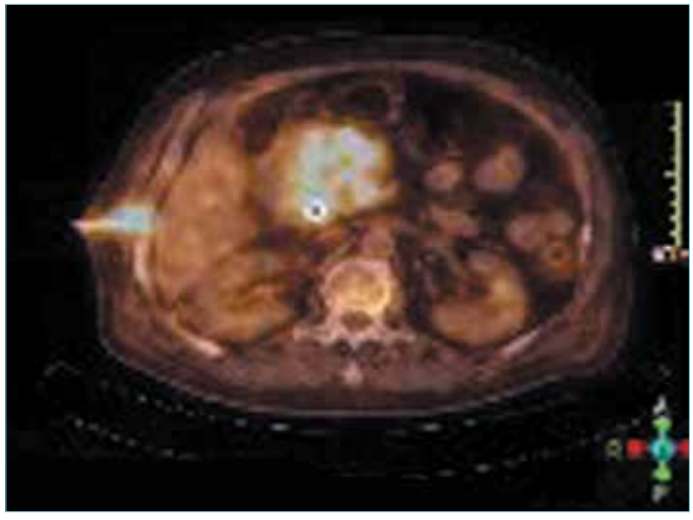

Figure 2. The PET/CT scan demonstrates a mass of intense FDG uptake in the head of the pancreas

bilirubins (up to 34 mg/dL). Therefore, treatment with concomitant radiotherapy and 40 mg/day on 1-4. day dexamethasone was urgently started. Obstructive jaundice disappeared after 2 weeks of chemoradiotherapy. After 3 months, the levels of his liver enzymes remained normal and a control CT scan revealed complete disappearence of the pancreatic plasmacytoma. Written informed consent was obtained from the patient.

DISCUSSION

A solitary plasmacytoma (SP) is defined as a neoplastic plasma cell tumor that may occupy extramedullary sites without an evidence of MM (1). SPs are categorized into two groups: SPs of the bone (SBP) and EMP of the soft tissue. SPs are uncommon

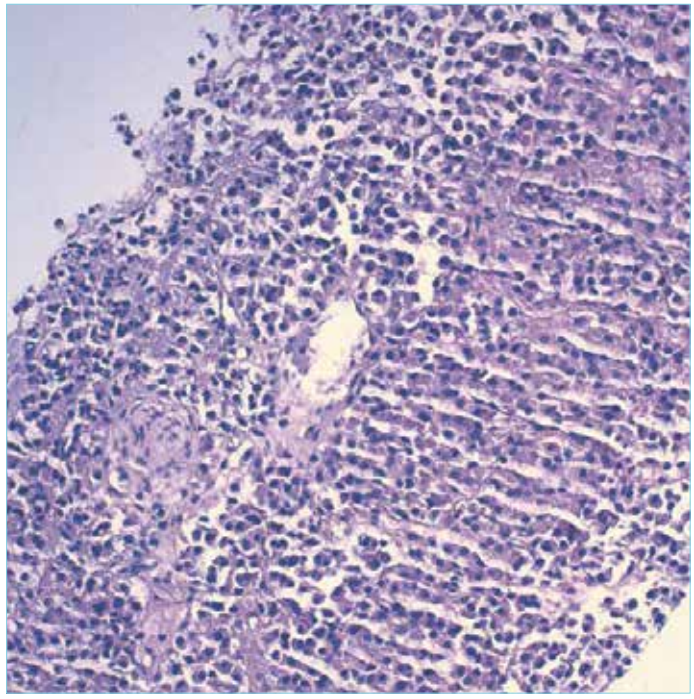

Figure 3. A US-guided fine needle biopsy of the demonstrating sheets of atypical plasma cells which were positive with monoclonal lambda light chain staining by immunoperoxidase stains. No staining was seen for kappa light chains (H & E stain, 20× magnification)

and constitute 2%–5% and 3%, respectively, of all plasma cell neoplasms (4). SBP are most often found in the vertebra and skull, whereas EMPs almost arise in the upper respiratory tract, including the oropharynx, nasal cavity, paranasal sinus, and larynx. Gastrointestinal tract involvement is uncommon, with the most common organs affected being the liver, spleen, and lymph nodes (5, 6). The pancreas is rarely infiltrated by plasmacytes. Only 25 cases of EMP of the pancreas have been reported in the English literature to date. Hayes et al. (6) reported 182 autopsies of patients who died of MM and observed only involved pancreas in seven patients (3%).

EMPs may be primary or secondary, but most EMPs generally occur secondary to MM or another EMP. According to the English literature, in our patient, the primary lesion was an SBP of the sternum, which had been successfully treated by radiochemotherapy. The recurrence of SBP converting to an EMP without systemic spread is rarely reported in the English literature. In our investigation, we found four cases with SBP converting to an EMP; none of them showed evidence of progression to MM (7).

The typical presentation of EMP of the pancreas includes jaundice and abdominal pain, which is often related to the obstruction of the biliary tree. The radiologic features are non-specific. Ultrasound demonstrates a hypoechoic heterogeneous focal mass. It is most often located in the head of the pancreas (8). The CT appearance of pancreatic plasmacytoma is well established and typically described as a multilobular, hypodense solid tumor (9). Other possible diagnostic modalities such as endoscopic US, magnetic resonance imaging, and PET can also detect the pancreatic mass. Diagnosis

is generally made by CT/US-guided percutaneous fine needle biopsy, as in our case (10).

There appears to be no standardized treatment for EMP of the pancreas. External beam radiotherapy, chemotherapy, and bypass procedures, alone or in combination, have been described as treatment methods. Because of the highly radiosensitive nature of plasma cell tumors, radiation therapy has been suggested as the treatment of choice (4, 8). Surgical procedures other than distal pancreatectomy for isolated pancreatic tail involvement are not commonly performed because of the common systemic nature of the disease and the radical nature of these surgical procedures. Chemotherapeutic agents are commonly used, particularly when plasmacytomas are secondary in nature (1). In the patients who underwent radiotherapy or chemotherapy, the resolution of biliary obstruction was noted in the majority, thus avoiding the need for surgical bypass or stenting (1, 10). In our patient, the resolution of obstructive jaundice was achieved with the use of radiotherapy and chemotherapy.

CONCLUSION

We believe that whenever a patient with a clinical history of plasma cell neoplasm has a pancreatic mass, EMP of the pancreas should be considered as a differential diagnosis. Because of the high radiosensitivity of EMP, radiation is the first treatment of choice, and unnecessary surgical resection should be avoided.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Authors' contributions: Conceived and designed the experiments or case: DOC. Performed the experiments or case: DOC. Analyzed the data: AY, LK. Wrote the paper: DOC. All authors have read and approved the final manuscript.

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