Plasmacytoma of the Pancreas

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Extramedullary plasmacytomas are plasma cell tumors consisting of neoplastic plasma cell proliferation that occurs outside the bone marrow. We describe a rare case of pancreatic plasmacytoma that was incidentally diagnosed in a patient with known multiple myelomas and newly discovered gastric lymphoma. The purpose of this report is to highlight this rare manifestation of multiple myeloma in the light of previous reports of the same lesion.

Patient Description

An 82 year old woman with an 8 year history of immunoglobulin G K-type gammapathy presented with nausea and fatigue. Her disease manifested as sacral and base of skull plasmacytomas. Bone marrow aspiration from the sternum revealed 5% plasma cells. She was treated with local radiotherapy followed by five courses of the M2 protocol (carmustine, cyclophosphamide, vincristin, melphalan and prednisone). Her bony disease progressed gradually, but she was lost to follow-up for 2 years.

Physical examination revealed a thin pale woman. On palpation an epigastric mass was encountered. Stool examination was positive for occult blood and laboratory tests revealed iron deficiency anemia (hemoglobin 7.5 g/dl, mean cell volume 66 fl). Serum bilirubin level and liver function tests were within normal limits. Serum IgG level was 977 mg/dl. Kappa light chain levels in urine were elevated. Repeated bone marrow aspiration at that stage demonstrated 25–30 plasma cells.

Endoscopy revealed a large ulcerated mass in the fundus and anterior wall of the stomach, which was shown by biopsy and histologic examination to be an immunoblastic large cell lymphoma. Contrast-enhanced computed tomography examination of the abdomen was performed as part of the evaluation and demonstrated in addition to the gastric tumor a hypodense homogenous mass in the head of the pancreas (Figure). The tumor measured 5 x 7 cm in diameter and had smooth well-defined margins. The peripancreatic fat was intact with absence of bile duct dilatation. CT-guided percutaneous transgastric core needle aspiration biopsy of the pancreatic mass revealed monodonal kappa light chain plasmacytoma. The pancreatic mass disappeared following radiotherapy. The gastric mass resolved after CHOP chemotherapy (cyclophosphamide, vincristin, Adriamycin and prednisone) and the gastrointestinal symptoms subsided. Eight months later the patient died of septic shock.

Comment

Multiple myelomas may present as diffuse myelomatosis, solitary myeloma of bone, or as an extramedullary plasmacytoma. The disease comprises 10–15% of hematologic malignancies. Extraskeletal plasmacytomas are uncommon, occurring in less than 5% of plasma cell neoplasms, and are usually associated with the more aggressive anaplastic form of the disease. Although extraneous lesions can involve any tissue or organ, the most common sites for extramedullary myelomatous infiltration are the submucosal lymphoid tissue of the upper respiratory tract, followed by the spleen, lymph nodes, liver and kidney [1]. Pancreatic involvement is rare, being found in approximately 2.3% of autopsies.

Our patient had concomitant plasmacytoma of the pancreas and B cell lymphoma of the stomach. Lymphoma and multiple myeloma are both B cell disorders. Plasmacytoma is a tumor consisting of monomorphic proliferation of morphologically mature-appearing plasma cells. In contrast, B cell lymphoma is an undifferentiated B cell neoplasm. In cases of gastric B cell lymphoma with plasma cell differentiation, the tumor consists of a proliferation of lymphoid cell components and mature and immature plasma cell components [2]. This was not the pathologic appearance in our case.

Reports of pancreatic plasmacytomas are sparse, and to date only 17 well-documented cases with imaging description of the pancreatic mass have been described in the literature [3–5]. In all these cases the pancreatic plasmacytoma...
was associated with myelomatosis to other extra-osseous or bony sites. Of these 17 cases, 11 presented with obstructive jaundice secondary to the mass in the head of the pancreas [3-5]. The CT appearance in most of the previously reported cases was of a focal mass usually in the head of the pancreas. In one case a diffuse pancreatic infiltration was demonstrated, and in another, a mass in the pancreatic tail [3]. In one patient two concomitant pancreatic plasmacytomas were found [5].

CT is the ideal method for demonstrating pancreatic masses. The CT appearance of pancreatic plasmacytoma is well established and is typically described as a multilobular homogenous solid tumor that is hypodense as compared to the pancreatic parenchyma. However, these CT features are not specific since they resemble typical findings in other pancreatic neoplasms, including carcinoma, islet cell tumors, lymphoma and metastases. Other possible diagnostic modalities, such as ultrasonography, endoscopic ultrasonography, magnetic resonance imaging and positron-emission tomography CT, can also demonstrate the pancreatic mass but are not able to determine its exact nature. Diagnosis is usually and easily made by CT-guided percutaneous fine needle aspiration biopsy [1], as in our case.

In conclusion, in a patient with multiple myeloma and a pancreatic mass, pancreatic plasmacytoma should be considered. A correct diagnosis obviates unnecessary surgery, as this neoplasm usually responds well to local radiotherapy.

References

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Giant Hemangioma of the Adrenal

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Key words: giant hemangioma, adrenal

Adrenal hemangiomas are a well-known entity, but giant adrenal hemangiomas are uncommon. We report on a patient with a symptomatic giant right adrenal hemangioma that was initially presumed to originate from the liver. The clinical presentation and management are described.

Patient Description
A 45 year old healthy woman complained of early satiety, right upper quadrant discomfort and fullness of 6 months duration. She denied fever, weight loss, jaundice or other systemic complaints. Initial workup revealed a mild elevation in liver enzymes, and a subsequent computed tomography scan of the abdomen revealed what was believed to be a giant liver hemangioma (Figure). Because of continuing abdominal discomfort the patient was taken to the operating room for exploration. Intraoperatively, a large 10 cm firm cystic mass was identified under the right liver lobe. After mobilization of the right liver lobe, it became clear that the mass did not violate the liver capsule but instead arose from the right adrenal gland. A right adrenalectomy was performed en bloc with removal of the mass.

Gross pathologic examination showed a 10.8 cm ovoid mass with multiple cystic spaces filled with fresh organizing thrombus arising from the adrenal gland. Histologic examination showed large hemangiomatous pockets lined with flattened endothelial cells. The tumor compressed the adrenal remnant, which showed normal histology. The findings were consistent with an adrenal hemangioma. The patient had an uneventful recovery with no further symptomatology.

Comment
Hemangiomas of the adrenal gland are rare. The first clinical case was reported in 1955 by Johnson and Jeppesen [1]. Since then, approximately 50 cases have been described, with fewer than 10 having a diameter larger than 10 cm.