Pancreatic Mass is not always Adenocarcinoma

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Introduction

Ductal adenocarcinoma accounts for 85% to 90% of pancreatic tumors.[1] Non-Hodgkin's lymphoma (NHL) frequently arises in extra-nodal sites, with about 50% of patients having extra-nodal involvement. Extra-nodal involvement is seen most commonly in the gastrointestinal tract and bone marrow. Involvement of the pancreas by NHL has been reported infrequently.[2,3] Only about 0.2 - 2% of patients with NHL have pancreatic involvement at the time of presentation.[1,3,4] Histopathological examination is usually necessary to obtain a definitive diagnosis since symptoms and radiological features are similar to those of other pancreatic masses. We present a case of pancreatic lymphoma presenting as abdominal pain. The patient was diagnosed with a CT-guided biopsy and responded very well to chemotherapy.

Case Report

A 74-year-old Caucasian female patient presented with a past medical history significant for hypertension and arthritis. Her chief complaints were abdominal pain and weight loss. Pain was located in the epigastrium, sharp in nature, constant, and severe. Her symptoms began three months prior but worsened over time. She denied any fever or chills.

On examination, the patient had diffuse abdominal tenderness greatest in the epigastric region. Laboratory data showed iron deficiency anemia, sodium of 127 mEq/L, potassium of 2.9 mEq/L, and magnesium of 1.6 mEq/L. Liver function tests and lipase were normal. She underwent an abdominal ultrasound which showed a hypoechoic area in the spleen measuring 3.2 x 3.7 x 2.7 cm and was thought to be a cyst. She had worsening gastrointestinal symptoms as well as 20 pound weight loss.

A CT scan of the abdomen showed a soft tissue mass near the spleen originating from the tail of the pancreas with extension to and encasement of the superior mesenteric artery and celiac artery. At that point, unresectable adenocarcinoma of the pancreas was very high in the differential diagnosis and her options seemed to be limited. Her workup included a cancer antigen (CA) 19-9, which was 8, within the normal limit. Endoscopic ultrasound was performed with fine-needle aspiration (FNA) of the mass and sent for pathology. Her FNA was inconclusive and a CT-guided biopsy revealed a diffuse large B cell lymphoma, nongerminal center subtype with an immunoprofile positive for CD20, BCL-6, BCL-2, and negative for CD10. A positron emission tomography (PET) was negative in the neck, chest, and pelvis with no osseous or hepatic involvement of the metabolically-active neoplasm.

The patient was started on chemotherapy with R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) for six cycles. She had an excellent response to her first two cycles of chemotherapy and her repeat PET scan was

consistent with complete response. After six cycles of chemotherapy, her symptoms resolved completely.

Discussion

Diffuse large B-cell lymphoma infrequently involves the pancreas.[5,6] The presenting symptoms of pancreatic lymphoma are usually non-specific and include abdominal pain (83%), abdominal mass (58%), weight loss (50%), jaundice (37%), acute pancreatitis (12%), small bowel obstruction (12%), and diarrhea (12%).[5,6] Clues suggesting the possibility of a primary pancreatic lymphoma include a lack of jaundice, constitutional symptoms (weight loss, fever, and night sweats), an elevated serum lactate dehydrogenase (LDH) or beta-2 microglobulin level, and a normal serum CA 19-9.

Imaging is sometimes important in the diagnosis and staging of pancreatic lymphoma. CT scan is the modality commonly used for the detection of pancreatic lymphoma.[7,8] It can present either as a tumor-like, well-circumscribed hypo-echoic mass or as a diffuse enlargement infiltrating the pancreas.

Pancreatic lymphoma might be distinguished from pancreatic adeno-carcinoma by the absence of pancreatic duct involvement and the presence of surrounding lymphadenopathy.[7,8] Primary pancreatic lymphomas typically are larger than 6 cm, and surrounding lymph-adenopathy is common as with any lymphoma. Neither of these features, however, would exclude adenocarcinoma. It is unlikely to have a pancreatic adeno-carcinoma above 10 cm in size; about 60% of pancreatic lymphomas are greater than 6 cm in diameter.[9]

The location of the tumor in the pancreas does not appear to be helpful in determining whether the mass is adenocarcinoma or lymphoma. Ultrasound or CT-guided fine needle biopsy of the pancreatic mass can help in the diagnosis of pancreatic lymphoma.[7] In the absence of any pathognomonic clinical or radiological features, the diagnosis is established only on histopathological examination.

Our patient was found to have a mass on CT scan. Endoscopic ultrasound-guided (EUS) biopsy did not help in diagnosis, so CT-guided biopsy was done and pancreatic lymphoma was diagnosed. A PET scan did not show any metastasis. The patient responded very well to chemotherapy with resolution of symptoms and clearing of tumor burden on imaging.

Treatment and prognosis of pancreatic lymphoma are significantly different from those for pancreatic adenocarcinoma.[7] Anthracycline-based chemotherapy is the standard treatment for many types of NHL, and includes six to eight cycles of R-CHOP for patients of all ages.

Diffuse large B-cell lymphoma rarely presents with synchronous pancreatic and splenic localizations. On literature review, no other cases of primary lymphoma of the pancreas confounded with a splenic involvement were found. Although the splenic mass was not biopsied, it resolved after treatment with chemotherapy. Primary pancreatic lymphoma should be considered in the differential diagnosis of pancreatic tumors and an attempt to obtain a tissue diagnosis is always necessary before proceeding to radical surgery, especially on young patients.

Conclusion

Pancreatic B-cell lymphoma is a rare tumor. Most of the tumors of the pancreas are adenocarcinoma. Pathologic diagnosis is important in distinguishing both of them. It is crucial to diagnose pancreatic lymphoma because the prognosis and treatment are different. Most cases of pancreatic lym-phoma respond very well to chemotherapy.

EUS with fine needle aspiration is usually the diagnostic method used for the diagnosis of pancreatic malignancies; however, it has not been well studied in primary pancreatic lymphoma due to the rarity of the disease.

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