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CASE REPORT | PEDIATRICS

Primary Mediastinal Large B-Cell Lymphoma Presenting as Acute Pancreatitis

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ABSTRACT

Pancreatitis is an inflammatory pancreatic disease; common etiologies include infection, anatomic abnormalities, biliary, inborn errors of metabolism, trauma, and rarely malignancy. Primary mediastinal large B-cell lymphoma commonly presents in younger women with principally mediastinal involvement. We report the first documented case of a pediatric patient presenting with acute pancreatitis secondary to metastatic primary mediastinal large B-cell lymphoma. Since diagnosis, the patient underwent a combination of chemotherapy and immunotherapy treatments, and the tumor burden had decreased significantly. Malignancy is a rare documented presentation of acute pancreatitis in the pediatric population, and it should be included in a broad differential diagnosis.

KEYWORDS: pediatric pancreatitis; pediatric malignancies; primary mediastinal large B-cell lymphoma

INTRODUCTION

Pancreatitis is an inflammatory disease of the pancreas, and common etiologies in pediatric patients include infection, drug-induced, anatomic abnormalities, biliary, inborn errors of metabolism, trauma, and rarely malignancy.[1] Primary mediastinal large B-cell lymphoma (PMBCL) is a rare and aggressive subtype of a non-Hodgkin B-cell lymphoma that originates in the mediastinum. It is often seen in female patients with a diagnosed median age of 35 years.[2] Diagnosis of PMBCL is established through histopathologic and cytopathologic testing.[3] Less than half of PMBCL arise from extranodal sites, including the gastrointestinal tract, skin, soft tissues, genitourinary tract, and rarely bone marrow.[4] In the gastrointestinal tract, PMBCL has been found to arise in the stomach, intestines, and liver.[4,5] Pancreatic involvement in PMBCL is very rare and has not been previously reported in pediatric populations.[6] Although there are several known cases of other non-Hodgkin lymphomas (Burkitt and B-cell) presenting as acute pancreatitis in children, our group presents the first documented case of a pediatric patient presenting with acute pancreatitis secondary to metastasis of PMBCL to the patient's pancreas.[7–10]

CASE REPORT

An otherwise healthy 16-year-old woman presented with 2 weeks of worsening epigastric pain and mild intermittent chest pressure. The patient denied fevers, coughs, jaundice, emesis, diarrhea, constipation, mental status changes, new signs of bruising, or weight loss. Clinical investigation demonstrated a leukocytosis (white blood cells 11.8 K/μL) and elevated lipase (2,524 U/L). At the time of presentation, an abdominal ultrasound was obtained due to suspected pancreatitis, which demonstrated a 5-cm mass of the pancreatic head and uncinate process, prompting further imaging through computed tomography abdomen/pelvis and transfer to Loyola University Medical Center for escalation of care. After the patient's admission to Loyola University Medical Center, the pancreatitis was managed conservatively with fluid resuscitation, antiemetics, analgesia, and bowel rest. Abdominal magnetic resonance imaging showed a 4.2-cm mixed solid/cystic pancreatic head mass with mild pancreatic duct dilation and a 2.6-cm hypoenhancing lobulated solid mass at the inferior pole of the spleen (Figure 1). Magnetic resonance cholangiopancreatography

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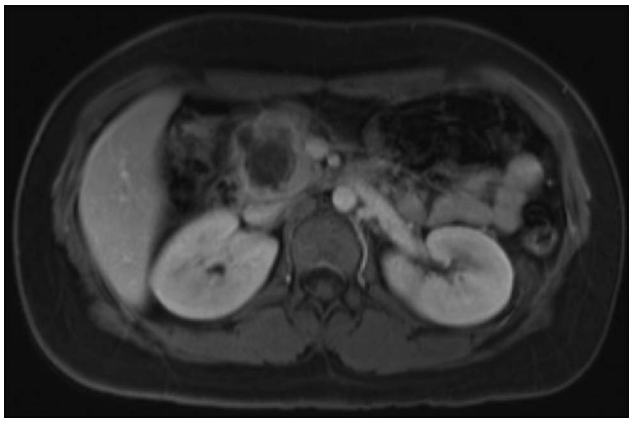

Figure 1. Abdominal magnetic resonance shows a mixed solid/cystic pancreatic head mass.

showed a necrotic pancreatic head mass, solid spleen mass, and an upper mediastinal mass. A repeat computed tomography abdomen/pelvis showed a 10.1-cm mediastinal mass with internal necrotic change (Figure 2). A transcatheter core needle biopsy of the mediastinal mass was consistent with PMBCL. The patient was initiated on a regimen of both chemotherapy and immunotherapy with etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and rituximab (DA-EPOCH-R) and nivolumab. At present, imaging shows full resolution of extrathoracic metastases with no evidence of disease recurrence at 6 months off of therapies.

DISCUSSION

Our patient is the first reported child who presented with acute pancreatitis secondary to PMBCL metastasis. PMBCL often presents with compressive symptoms secondary to a mediastinal mass. Patients are commonly dyspneic, complain of a persistent cough, and can present with superior vena cava syndrome.[11] Our patient did not present with any superior vena cava symptoms.

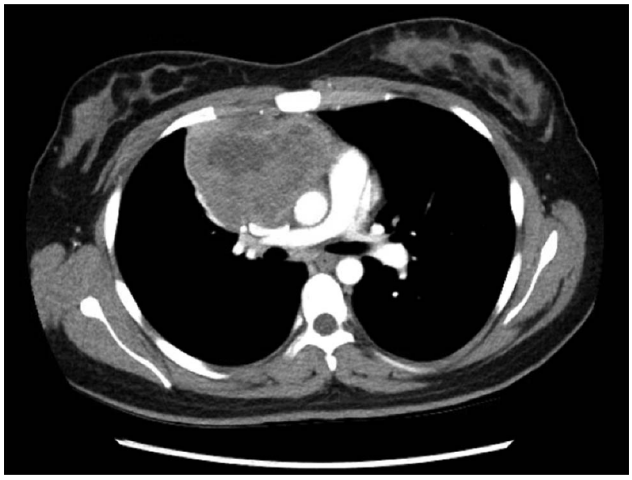

Figure 2. Abdominal computed tomography shows a mediastinal mass with internal necrotic change.

Instead, she presented with diagnostic features consistent with acute pancreatitis, including worsening epigastric pain and an elevated lipase. PMBCL commonly spreads to adjacent thoracic structures such as the pericardium, lungs, and chest wall.[12] Our patient's mediastinal mass had an unusual extrathoracic metastatic distribution to her pancreas and spleen.

According to the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition official guidelines for managing acute pancreatitis in the pediatric population, a pancreatitis diagnosis requires at least 2 of the following: abdominal pain, serum amylase, and/or lipase 3 times above the upper limits of normal and/or imaging findings consistent with acute pancreatitis.[1] Of note, imaging in the early phases of acute pancreatitis is not required if the patient's history, presenting symptoms, and chemical laboratory tests could be used to make a diagnosis.[1] Our patient exhibited abdominal pain and an elevated lipase; she had met 2 of the 3 criteria for acute pancreatitis, which could have resulted in an initial diagnosis of acute pancreatitis without pursuing further imaging, meaning her PMBCL diagnosis could have potentially been missed. Imaging was crucial for the diagnosis of malignancy in this patient.

Effective first-line treatments for PMBCL include chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone with radiation, or DA-EPOCH-R with or without radiation.[13] Remission rates in patients using EPOCH are 80% compared with 70% for cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP) patients; EPOCH patients, however, are more likely to experience treatment-related toxicity.[14–16]

We were able to enroll the patient in a Children's Oncology Group Clinical trial (COG ANHL1931), which incorporates 6 cycles of chemoimmunotherapy with nivolumab. The patient demonstrated a favorable clinical response, and no modifications to the protocol were required. We anticipated that there was lymphomatous involvement of the pancreas and that the patient's pancreatitis would improve after initiation of therapy. We continued to manage the patient medically for her pancreatitis during her chemotherapy treatments, which included routine monitoring of her pain, pain management, dietary modifications, and serial measurements of amylase and lipase. Her pain resolved after the first day of chemotherapy. We observed a peak of her amylase (207 U/L) and lipase (1,255 U/L) during her first cycle of chemotherapy. Amylase and lipase levels then normalized 3 weeks after her first cycle of chemotherapy was completed. We report that our patient tolerated treatment with only intermittent nausea and occasional fatigue. She is currently 9 months off therapy with no evidence of disease.

Although rare, malignancy is a documented precipitant of acute pancreatitis in the pediatric population, and it should be included in a broad differential diagnosis when evaluating similar constellations of symptoms. Pediatric patients in which pancreatitis is suspected should undergo ultrasonographic evaluation of the pancreas due to the cost-effective, accessible, and dependable nature of this imaging modality.

DISCLOSURES

Author contributions: K. Krawczyk, A. Kwak, C. Kujalowicz, and K. Truszkowska performed initial chart review and data collection. K. Krawczyk, A. Kwak, C. Kujalowicz, V. Biank, and N. Kamberos reviewed and interpreted the results. The manuscript was drafted by K. K. Krawczyk, A. Kwak, and C. Kujalowicz. The final manuscript was written and edited by K. K. Krawczyk, A. Kwak, C. Kujalowicz, V. Biank, and N. Kamberos. K. Krawczyk is the article guarantor. All authors have approved the final version of the manuscript.

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