

Primary Pancreatic Lymphoma Simulating Acute Cholestatic Hepatitis in a 7-Year-Old Child

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Abstract

Primary pancreatic lymphoma in children has been described infrequently in literature, and its acute presentation as cholestatic hepatitis is similarly rare. We report a case of a 7-year-old child with primary pancreatic lymphoma presenting as acute infective hepatitis, leading to delay in correct diagnosis and management.

Introduction

Though seen commonly in adults, pancreatic malignancies are an exceedingly rare entity in the pediatric population.¹⁻³ Lymphomatous involvement of pancreas can occur primarily, or, more commonly, as a secondary phenomenon of metastasis.¹ Primary pancreatic lymphoma (PPL) in children has been described infrequently in literature, constituting less than 2% of all extranodal non-Hodgkin's lymphoma, and its acute presentation as cholestatic hepatitis is similarly rare.⁴⁻⁹ In a recent 10-year retrospective study of malignant pancreatic tumors in children involving 2 centers, no cases of PPL were found.²

Case Report

A 7-year-old child was admitted with complaints of yellow discoloration of eyes with dark-colored urine for 20 days. Jaundice was painless and gradually progressive, with development of pale stools and pruritus. It was preceded by low-grade fever for 1 day with occasional episodes of nausea and non-bilious vomiting. There was no associated pain in the abdomen, night sweats, weight loss, loss of appetite, bone tenderness, fatigue, skin or mucosal bleeds, drug intake, abdominal distension, edema, delirium, symptoms of fat-soluble vitamin deficiency, or steatorrhea.

On examination, the child was well-developed with stable hemodynamics. Physical examination revealed pallor, icterus, and bilateral mild corneal xerosis. Abdominal examination was remarkable for a palpable liver 2 cm below right costal margin in midclavicular line, with a total span of 7.5 cm, soft in consistency, non-tender with smooth surface, and regular margins. We suspected acute infective hepatitis, which was initially managed conservatively with nutritional supplements.

Laboratory parameters showed evidence of conjugated hyperbilirubinemia and high cholestatic enzymes, suggestive of obstructive etiology. Abdominal ultrasound showed a well-defined, hypoechoic mass (6.4 x 4.5 cm) with internal vascularity arising from head of pancreas reaching to porta, encasing the common bile duct

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Figure 1. (A) Abdominal ultrasound showing hypoechoic ill-defined mass in pancreatic head (short arrows) with encasement of common bile duct (red arrow) and main pancreatic duct (not shown in figure). Note the echogenic wall (white arrow) of the common bile duct coursing through the lesion, which is well appreciated in grayscale ultrasound. (B) Axial contrast-enhanced abdominal CT revealing poorly marginated hypodense homogenous hypoechoic mass in pancreatic head (short arrows) encasing spleno-portal confluence, entire main portal vein (white arrow), and celiac axis (red arrow). (C) Coronal contrast-enhanced abdominal CT with minimum intensity projection reformation revealing gross dilatation of the main pancreatic duct (white arrow) and common bile duct (red arrow) with bilobar intrahepatic duct dilatation.

(CBD) with upstream biliary dilation (Figure 1). Contrast-enhanced abdominal computed tomography (CT) revealed a large homogeneously hypoechoic lobulated mass (4.8 x 5.4 x 5.4 cm isodense to pancreas in plain scan) in the pancreatic head encasing the common hepatic artery and main portal vein (Figure 1). The mass was obstructing the distal CBD and main pancreatic duct (MPD) with associated upstream ductal dilation (CBD 13 mm; MPD 4 mm) and bilobar gross intrahepatic biliary dilatation, consistent with double duct sign. Posteriorly, the mass encased the left renal vein (Figure 1). There was no evidence of calcification/necrosis noted within the mass. Tumor markers showed elevated alpha-fetoprotein (AFP) of 4.9 ng/mL, CA-125 of 57.5 U/mL, and CA19-9 of 783 U/mL.

Fine-needle aspiration cytology/cell block from the mass showed atypical large lymphoid cells with enlarged, hyperchromatic nuclei with high nucleus: cytoplasm ratio, conspicuous nucleoli, and scanty rim of deep basophilic cytoplasm suggestive of diffuse large B-cell lymphoma (CD20/CD45 positive and CD3/CD30 negative on immunohistochemistry; Figure 2). Contrast-enhanced chest

CT, bone marrow aspiration/biopsy, and cerebrospinal fluid examination done for staging showed abnormal malignant lymphoid cells in bone marrow, but none in cerebrospinal fluid, and a prominent thymus in chest. Ann Arbor staging was IVEA.¹⁰ The child was referred to a pediatric oncology center for management, including chemotherapy.

Discussion

PPL is extremely rare, both in adults and children, with only limited case reports or small case series available in world literature; it accounts for <1% of extranodal lymphomas and 0.7% of all pancreatic malignancies.^{4-9,11} PPL is an extranodal lymphoma arising in the pancreas, with bulk of the tumor localized to the pancreas.⁴ Standard diagnostic criteria include: 1) neither superficial lymphadenopathy nor enlargement of mediastinal lymph nodes on chest radiography, 2) a normal leukocyte count in peripheral blood, 3) main mass in the pancreas with lymph nodal involvement confined to peripancreatic region, and 4) no hepatic or splenic involvement.¹² Though no pediatric-specific definition is available, our case fulfilled the standard definition.

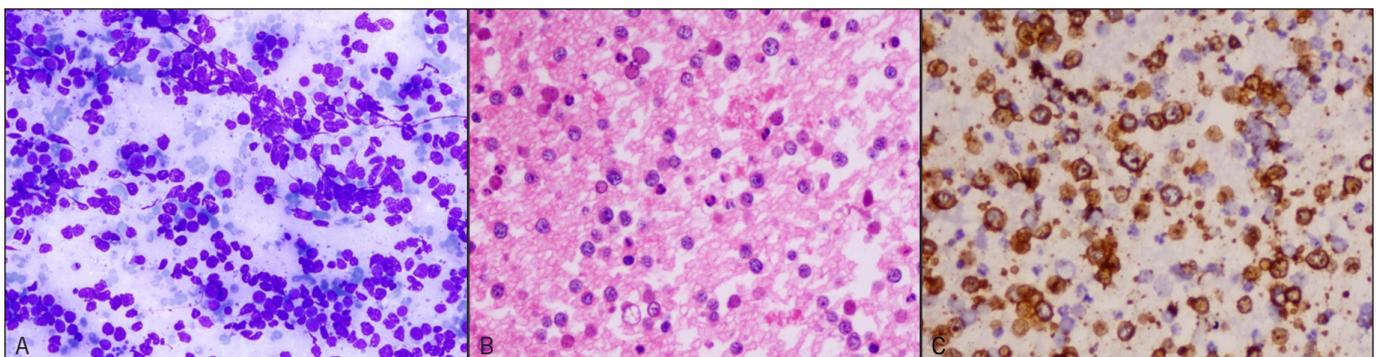


Figure 2. Cytology smear from pancreatic mass showing (A) Geimsa stain of large malignant lymphoid cells (200x), (B) H&E stain of scattered malignant lymphoid cells (200x), and (C) CD20 positivity on immunohistochemistry.

There are no specific clinical indications, but PPL usually presents with abdominal pain, abdominal mass, weight loss, jaundice, or acute pancreatitis.^{5,9} Presentation as obstructive jaundice is rare, seen in only about one-third of adult cases, and is much less common than in adenocarcinoma.^{5,7} Our patient presented with features of obstructive jaundice leading to a false first impression of acute cholestatic hepatitis. Diagnosis by ultrasound, CT, or endoscopic ultrasound is suggested, but requires mandatory histological confirmation with tissue biopsy. Histologically, most of PPL are B-cell type, with diffuse large-cell lymphoma as the predominant histotype.⁹

Classical imaging findings include well-defined, sometimes bulky and infiltrating, homogeneous low-attenuation masses relative to the enhancing pancreatic parenchyma, with only mild enhancement on contrast-enhanced CT and the presence of large lymph nodal masses.^{1,3,13} Lesion may appear as a solitary hypoenhancing lesion, multiple non-contiguous lesions, or as a diffusely enlarged gland.^{1,3,13} The head of the pancreas is the most common location of PPL.⁹ Vascular encasement or invasion, seen as irregularity and caliber changes, is rarely seen, though vessels may be stretched because of a mass effect. In our case, pancreatic head mass encased the common hepatic artery (and its branches), main portal vein, and left renal vein with mild attenuation, but no evidence of invasion. PPL can be distinguished from more common entities like pancreatoblastoma and solid-pseudopapillary tumor by the multiplicity of large nodal masses as the absence of internal heterogeneity, calcifications, necrosis, or vessel invasion.^{1,3} Double duct sign is highly suggestive, but not diagnostic, of pancreatic head malignancy or carcinoma of ampulla of Vater.¹⁴ Though ideal management algorithms are still to be developed, chemoradiotherapy is the modality of choice worldwide, while primary surgery is reserved for treatment failure cases or with diagnostic uncertainty.⁹

Disclosures

Author contributions: V. Sood is the article guarantor. V. Sood, N. Agrawal, D. Rawat, and R. Khanna contributed equally to assessing the clinical and laboratory information and to drafting the manuscript. V. Sood and N. Agrawal wrote the final manuscript. S. Alam and K. Bansal edited and revised the manuscript. C. Bihari assessed the pathology.

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References

- Shet NS, Cole BL, Iyer RS. Imaging of pediatric pancreatic neoplasms with radiologic-histopathologic correlation. *Am J Roentgenol*. 2014;202(6):1337–134.
- Rojas Y, Warneke CL, Dhamne CA, et al. Primary malignant pancreatic neoplasms in children and adolescents: A 20-year experience. *J Pediatr Surg*. 2012;47(12):2199–2204.
- Chung EM, Travis MD, Conran RM. Pancreatic tumors in children: Radiologic-pathologic correlation. *RadioGraphics*. 2006;26(4):1211–1238.
- Hamilton SR, Lauri LA, eds. Pathology and genetics of tumours of the digestive system. In: *World Health Organization Classification of Tumours*. Lyon, France: IARC Press; 2000.
- Saif MW. Primary pancreatic lymphomas. *JOP*. 2006;7(3):262–73.
- Arenas Garcia BR. Primary pancreatic lymphoma in pediatric patients [Article in Spanish]. *Radiologia*. 2007;49(2):125–127.
- Pietsch JB, Shankar S, Ford C, Johnson JE. Obstructive jaundice secondary to lymphoma in childhood. *J Pediatr Surg*. 2001;36(12):1792–1795.
- Eisenhuber E, Schoeffl R, Wiesbauer P, Bankier AA. Primary pancreatic lymphoma presenting as acute pancreatitis in a child. *Med Pediatr Oncol*. 2001;37(1):53–54.
- Yoon WJ, Yoon YB, Kim YJ, et al. Primary pancreatic lymphoma in Korea: A single center experience. *J Korean Med Sci*. 2010;25(4):536–540.
- Rosenberg SA. Validity of the Ann Arbor staging classification for the non-Hodgkin's lymphomas. *Cancer Treat Rep*. 1977;61:1023–1027.
- Luo G, Jin C, Fu D, et al. Primary pancreatic lymphoma. *Tumori*. 2009;95(2):156–159.
- Dawson IM, Cornes JS, Morson BC. Primary malignant lymphoid tumours of the intestinal tract. Report of 37 cases with a study of factors influencing prognosis. *Br J Surg*. 1961;49:80–9.
- Fujinaga Y, Lall C, Patel A, et al. MR features of primary and secondary malignant lymphoma of the pancreas: A pictorial review. *Insights Img*. 2013;4(3):321–329.
- Ahualli J. The double duct sign. *Radiology*. 2007;244(1):314–315.

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