A Rare Inflammatory Myofibroblastic Tumor of Pancreas in a Child With Cholestasis Presentation

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Abstract- Inflammatory myofibroblastic tumors (IMTs) are rare with unknown etiology. As pancreas involvement is rare in IMTs, here, we report a case of a girl with IMT, referred to our hospital. A 4-year-old girl presented with chief complaints of generalized itching and jaundice. Abdominopelvic computed tomography (CT) scans with contrast showed a homogeneous isodense mass lesion in the head of the pancreas with a compressive effect on the distal part of the common bile duct (CBD). Dilatation of intrahepatic bile ducts and CBD (8 mm) was observed. Magnetic resonance cholangiopancreatography (MRCP) examination showed a dilated gallbladder without stones and intrahepatic/extrahepatic bile ducts. The CBD was dilated, and a mass was found in the head of the pancreas. Immune-histochemical studies revealed spindle myofibroblastic tissues with lymphoplasmacytic and eosinophil infiltration. All of them were compatible with pancreatic IMTs. The surgery improved the symptoms. The IMTs of the pancreas can have symptoms like pancreatic cancer. The careful evaluation by imaging and pathology is recommended.

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Keywords: Inflammatory myofibroblastic tumors; Pancreas; Child

Introduction

Inflammatory myofibroblastic tumors (IMTs), which mostly occur among the youth and females, are a group of rare tumors that can develop in any organs (1). The IMTs are solid tumors consisting of myofibroblastic spindle cells and inflammatory cells (lymphocytes and plasma cells) (2). Although the exact cause is unknown, the human herpesviruses 3 and 8, Eikenella corrodens, and Epstein-Barr virus infections are considered as triggers of the IMT (3,4). Lungs, mesentery, and omentum are common sites of involvement, while the pancreas is a rare location of involvement (5).

In the case of pancreas involvement, abdominal pain, icterus, and weight loss are presenting symptoms, which can lead to misdiagnosis as pancreatic cancer.

In some cases, the mutations of genes will cause metastasis and/or recurrence (6-9).

For the pancreas, involvement is rare in IMTs; here, we report a case of a girl with IMT, referred to our hospital.

Case Report

A 4-year-old girl with generalized itching for three months, which worsened the last week before hospitalization, was referred to the Amirkola Children's Hospital in the north of Iran. Her chief complaints at admission were generalized itching and jaundice.

Her jaundice became worse three days before admission, and she had dark urine, steatorrhea, acholic stool as well as mild periumbilical abdominal pain with no fever or vomiting.

Her parents had no familial relationship, and she had no medical problem before her current complaint. Her growth was normal.

At the time of clinical examination, she was conscious, had icteric sclera and generalized excoriated cutaneous lesions as well as the liver was palpable 3cm under rib border with normal consistency, while other examinations were normal.

At admission, her vital signs were as follows: PR: 80/min, RR: 20/min, BP: 90/65 mmHg, T: 37° C.

The laboratory data revealed: CBC (complete blood count): WBC (white blood cell): 6.1×10^3/µL, Hb (hemoglobin): 11.3 g/dl, PLT (platelate): 341×10^3/µL,

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polymorphonuclear (poly): 62%, Lymphocyte (lymph): 35%, Monocyte: 1%, Eosinophil: 2%, ESR (erythrocyte sedimentation rate): 9 mm/h, AST (aspartate aminotransferase): 73IU/L, ALT (alanine aminotransferase): 71IU/L, ALP(alkaline phosphatase): 1891IU/L, GGT (gammaglutamyltransferase): 250U/L, T-Bil (total bilirubin): 4.9 mg/dL, D-Bil (Direct bilirubin): 3.5 mg/dl, BS (blood sugar), BUN (blood urea nitrogen), CREAT (creatinine), NA (natrium), K (kalium), Ca (calcium), P (phosphorus): Normal, CRP (C-reactive protein): 0/2 mg/dl, S/E (stool examination): Fat droplet positive, S/C (stool culture): Negative, U/A (urine analysis): Normal, U/C (urine culture): Negative, Amylase, Lipase, PT (prothrombin time), PTT (partial thromboplastin time), INR (international normalized ratio): Normal.

Abdominopelvic sonography revealed dilatation of the CBD in the proximal and distal part as well as intrahepatic bile ducts. Due to the incomplete bowel preparation, the pancreas evaluation was impossible. Kidneys and bladder were normal.

During admission, the patient became febrile, and then she underwent intravenous cefotaxime (50 mg/kg/q6h) with a possible differential diagnosis of acute cholangitis.

Her new laboratory data demonstrated

CBC: WBC: $11\times10^3/\mu L$, Poly: 70%, Lymph: 30%, ESR: 59 mm/h, CRP: 65mg/dl, AST: 345 IU/L, ALT: 215 IU/L, ALP: 1715 U/L, GGT: 680U/L, T-Bil: 6mg/dl, D-Bil: 4 mg/dl.

The PT, PTT, INR, Total Pr, Albumin, Amylase, Lipase, ANA, ASMA, Anti-LKM Ab, HIV Ab, Anti-HAV Ab, HBS Ag, HCV Ab, Wright and Widal tests, Alfa 1 Antitrypsin, ceruloplasmin, EBV and HSV Ab, CEA, CA19-9, αFP, βHCG were all normal.

The new ultrasound examination revealed dilatation of CBD (8 mm) as well as dilatation of intrahepatic portal and proximal pancreatic ducts.

Solid hypoechoic mass with a size of 19×21×22 mm was detected in the head of the pancreas. The liver, bladder, and kidneys were normal. Color Doppler evaluation of the portal venous system and hepatic artery were normal, too.

Abdominopelvic CT scan with contrast displayed a homogeneous and isodense mass lesion with a size of 28×25 mm in the head of the pancreas with a compressive effect on the distal part of the CBD. The pancreatic duct was visible and dilated (2.5 mm). Dilatation of intrahepatic bile ducts and CBD (8 mm) was found in her. The gallbladder was dilated. Spleen, kidneys, and bladder were all normal.

The magnetic resonance cholangiopancreatography (MRCP) examination illustrated the dilatation of gallbladder without stones as well as intrahepatic and extrahepatic bile ducts. The CBD was dilated (9 mm), and a mass was found in the head of the pancreas (25×29 mm) (Figure 1).

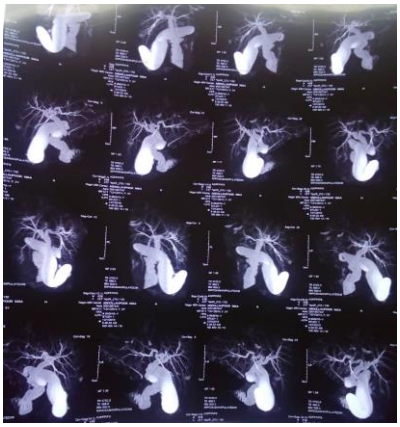

Figure 1. MRCP findings: Dilatation of intra/extrahepatic bile ducts and proximal pancreatic duct

The patient underwent Endoscopic Ultrasound (EUS) (Figure 2). Next, the fine needle aspiration (FNA) was taken from a mass under sonography guidance and finally was examined pathologically.

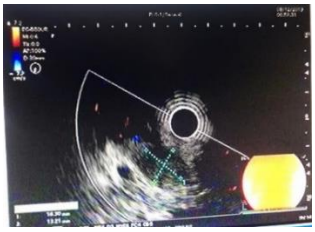

Figure 2. An endosonographic finding of pancreatic mass

The microscopic description was as follows

Moderately cellular lesion composed of bland-looking spindle cells with double-pointed dense, spindle-shaped nuclei and indistinct cytoplasmic borders arranged in a collagenous stroma. As shown in figure 3, the lesion entraps lobules of typical pancreatic acini and nerve fibers as well as contains small blood vessels in addition to a small number of lymphocytes and plasma cells.

Immune-histochemical studies through antibodies indicated

ALK1: Positive in spindle cells, CD138: Highlighting the small number of the plasma cell, IgG: Positive in a number of plasma cells, IgG4: No reaction, generally, all

of them were compatible with the pancreatic inflammatory myofibroblastic tumor.

The surgery was performed, and the mass (2.5-3 cm) was observed in the head of the pancreas. There was no ascites, peritoneal seeding, or metastasis. The size of the choledochal and pancreatic duct was 10 and 3 mm, respectively. The patient underwent Whipple (pancreaticoduodenectomy) surgery.

One week after surgery, she was discharged with a good general condition. Two weeks later, she was without icterus, and her liver enzymes were normal.

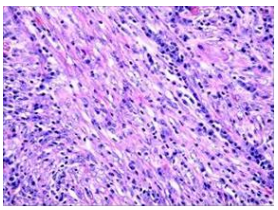

Figure 3. Pathology of pancreatic mass (IMT)

Discussion

The IMTs known as plasma cell granuloma, plasma cell pseudotumor, inflammatory pseudotumor, inflammatory fibroxanthoma, and histiocytoma are rare and tend to recurrence and metastasis (6,10). Though the exact cause is unclear, genetics and infections can play a role (3,11).

The IMTs can arise in different organs, but its occurrence in the pancreas is rare (12). The literature review has suggested that these tumors develop in a wide age range from 6 months to 82 years (1,13-15). Most cases (tumors) are asymptomatic and found incidentally. So, they could be misdiagnosed as pancreatic cancers. The symptoms could be abdominal pain, jaundice, weight loss, and palpable abdominal mass (14-18). A wide range of symptoms such as malaise, loss of appetite, and wasting may be appeared based on a compression effect on other organs (19).

Most previous pediatric cases, like the present case, were girls.

Our patient was a 4-year-old girl with generalized itching, abdominal pain, and generalized jaundice referred to the Amirkola Children's Hospital. She had elevated ALT and AST levels and normal levels of amylase and lipase.

Panda *et al*., reported a female patient with right upper quadrant pain as well as progressive jaundice, vomiting, and weight loss (16). The CT scan displayed a mass involving the head of the pancreas, dilated bile duct, and enlarged gallbladder.

In our case, the abdominopelvic CT scans with contrast indicated a homogeneous and isodense mass lesion in the head of the pancreas with a compressive effect on the distal part of the CBD. Dilatation of intrahepatic bile ducts and CBD (8 mm) was observed. The gallbladder was dilated.

Liu *et al*., represented a 15-year-old boy referred with abdominal pain (left upper quadrant) and fever. The ultrasound examination revealed a cystic lesion in the left upper quadrant. The tumor was in the pancreas tail, and the pathologic assessment illustrated the spindle cell tumor with myogenic differentiation. Immunohistochemical studies confirmed the existence of spindle cells stained positive for actin during a three-year follow-up, and he had no recurrence (20).

Battal *et al*., reported a 46-year-old man with acute abdomen symptoms, and the laboratory findings showed evidence of acute pancreatitis. Abdominal ultrasound demonstrated a mass in the head of the pancreas, and an abdominal MRI revealed a mass with solid and cystic components in the pancreas.

Our case underwent Whipple surgery, and her pathologic examination showed the lesion entrapped lobules of typical pancreatic acini and nerve fibers as well as contained small blood vessels in addition to a small number of lymphocytes and plasma cells. Moreover, the immune-histochemical studies using antibodies revealed a pancreatic inflammatory myofibroblastic tumor in her.

The IMT of the pancreas can be easily misdiagnosed as pancreas cancer, and the histological evaluation can be helpful for differential diagnosis. Sixty percent of pancreas IMTs are located in the head and remaining in the body and tail (16). More than two-thirds of patients have jaundice following pancreatic duct obstruction.

The primary treatment choice of IMTs is resection and careful follow-up of the patient. Chronic pancreatitis, fibrosarcoma, pancreatic carcinoma, and lymphoma are among the differential diagnoses (21).

In most cases, the prognosis is good, and the recurrence rate of IMTs is reported as 25% (19,22).

Our case, one week after surgery, had a good general condition. Besides, two weeks later, she was without icterus, and her liver enzymes were normal.

The IMTs of the pancreas may have symptoms such as pancreatic cancer. The careful evaluation by imaging and pathology is recommended.

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Ethics Approval

This study was approved by Health Research Institute of Babol University of Medical Sciences (ID: IR.MUBABOL.REC. 1399.094).

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