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A Case of Idiopathic Fibrosing Pancreatitis

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We experienced a case of chronic fibrosing pancreatitis in an 18/12-year-old girl, which was idiopathic because there were no familial back ground, no cystic fibrosis of pancrease, no ductal anomalies and obstruction. The patient presented intermittent colicky abdominal pain and progressive obstructive jaundice, but T-tube drainage and removal of the lymph nodes around the common bile duct relieved her symptoms and disease process. This seems to be the first case reported in a Korean child. Idiopathic fibrosing pancreatitis should be considered in the differential diagnosis of abdominal pain with obstructive jaundice in children.

Key Words: *Idiopathic fibrosing pancreatitis, chronic fibrosing pancreatitis, obstructive jaundice.*

INTRODUCTION

Idiopathic fibrosing pancreatitis is a chronic disease of unknown etiology characterized by pancreatic fibrosis, which is both intra-and extralobular (Williams et al., 1967). Chronic intermittent colicky abdominal pain is the main manifestation. Obstructive jaundice is so rare that it is rarely considered in the differential diagnosis of chronic abdominal pain and obstructive jaundice in a child. Only 16 pediatric cases have reported in the English literature. We experienced a case of idiopathic fibrosing pancreatitis in an 18/12-year-old girl with intermittent colicky abdominal pain and progressive obstructive jaundice as the first case in a Koera child.

CASE REPORT

An 18/12-year old girl was admitted to the Pediatric Department of Ewha University Hospital on September 8, 1988 due to intermittent colicky abdominal pain of five weeks' duration and progressive obstructive jaundice.

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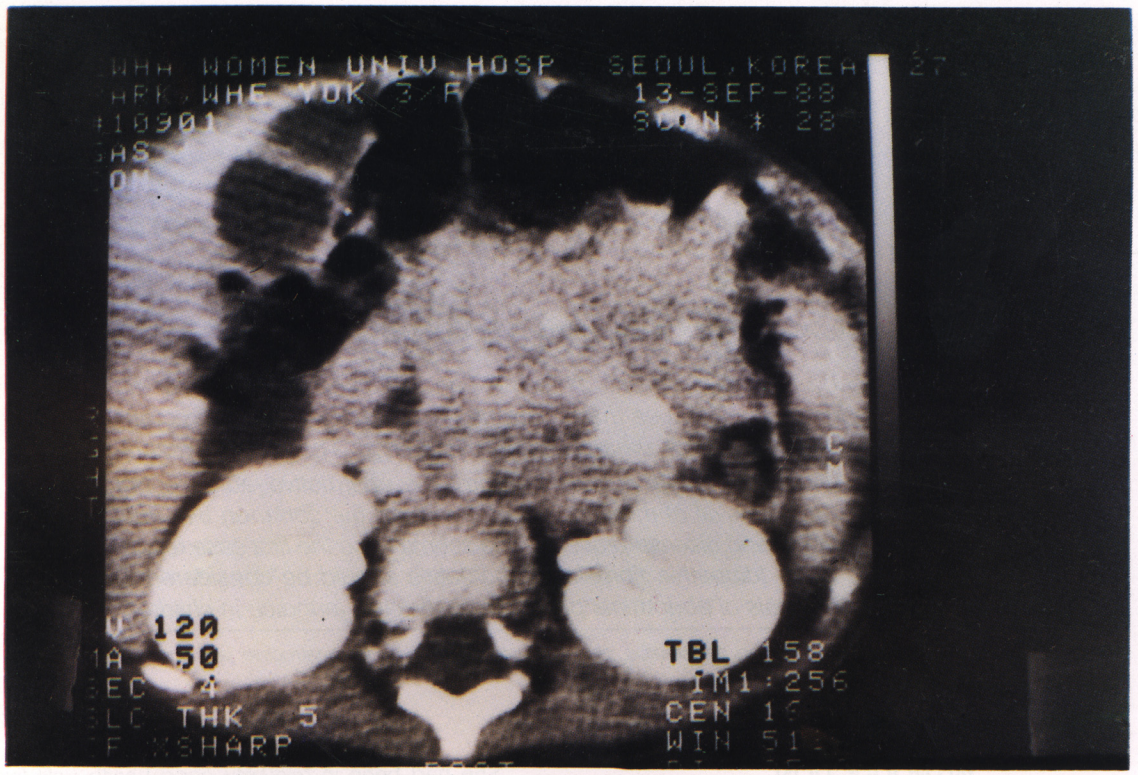
She had been in relatively good health until five weeks prior to admission, when she began suffering from colicky abdominal pain. The abdominal pain occurred spontaneously without special medical intervention. She had been brought to a local clinic, where jaundice was first noticed and liver function tests were found to be abnormal. She was admitted to another hospital on August 9, 1988 and treated for infectious hepatitis. The abdominal pain became less colicky, and jaundice seemed to reveal only mild hepatomegaly. But hepatitis A and B markers were all negative. On the 17th hospital day, her general condition became worse again. Intermittent mild fever, anorexia, nausea and colicky abdominal pain became more severe. The jaundice worsend again in a progressive obstructive pattern. Her weight loss was 3kg. She was transferred to the Pediatric Department of Ewha University Hospital.

On admission, she was acutely ill looking with deeply icteric skin and sclera. The liver was palpable 3cm below the right costal margin with a firm consistency. The spleen was not palpated. Liver function tests were abnormal with increased direct bilirubin (Table 1). Abdominal ultrasound revealed hepatomegaly, a moderately dilated biliary tree and an ill-defined retroperitoneal mass. In addition to hepatomegaly, an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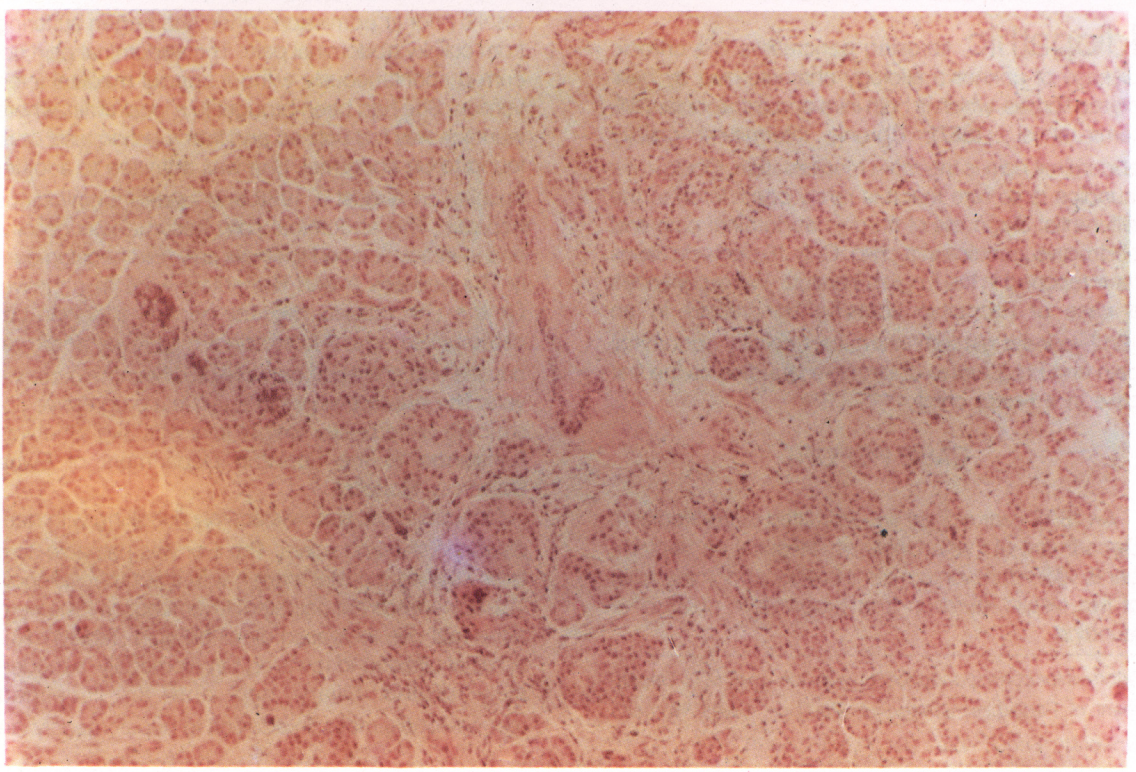

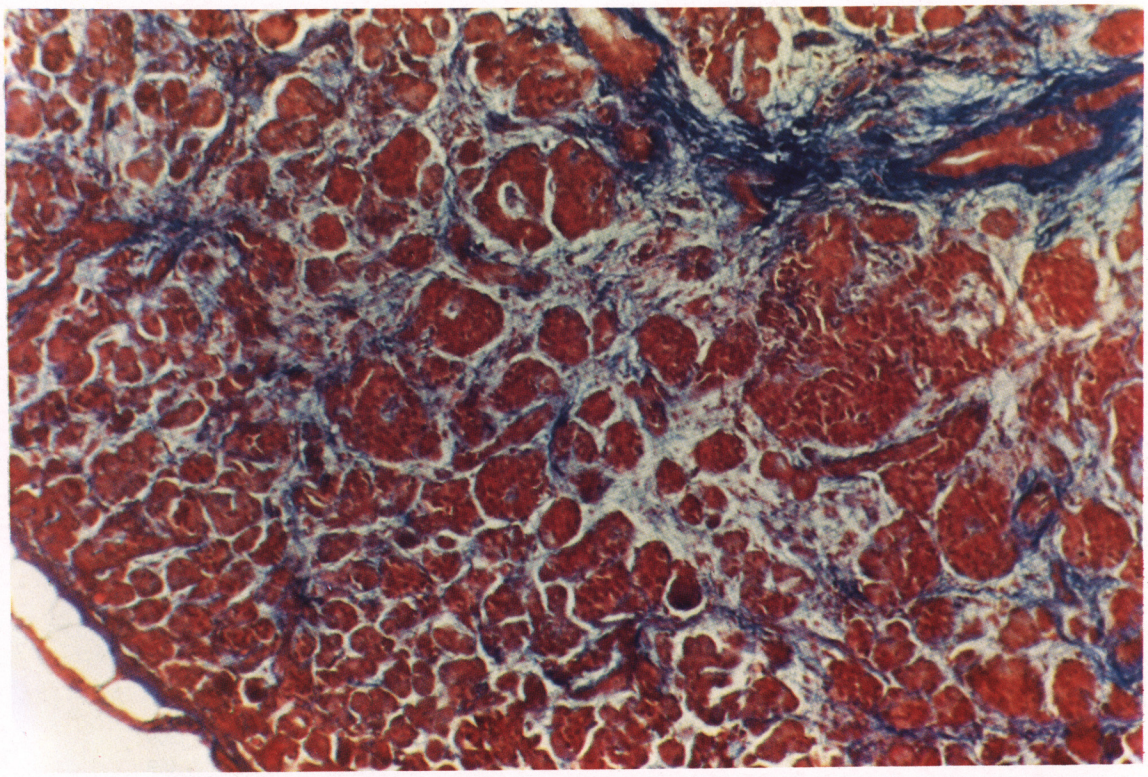

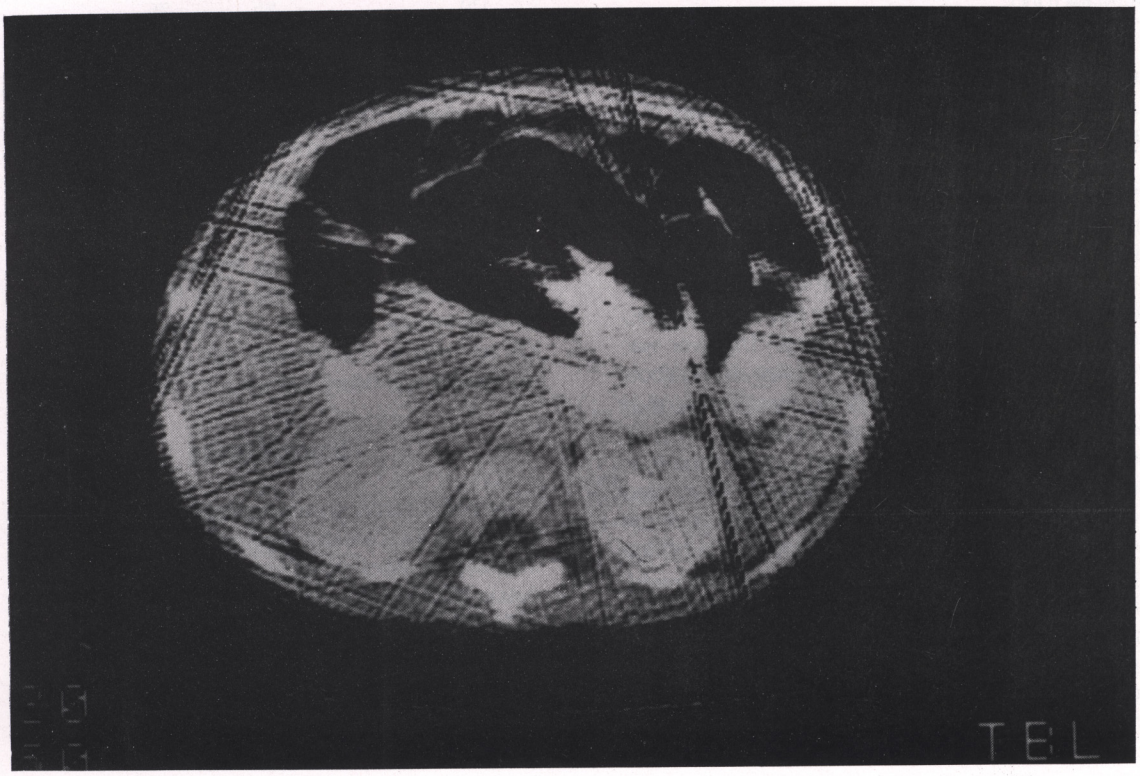
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Fig. 1. Preoperative abdominal CT reveals a huge retroperitoneal mass shadow.
Fig. 2. Microscopically the pancreas shows mild intra-and interlobular fibrosis with intact acini and islets (H & E, ×100).
Fig. 3. Microscopic finding of the pancreas (Masson's Trichrome, ×100)
Fig. 4. Postoperative abdominal CT reveals no retroperitoneal mass.

Microscopically, the pancreas showed mild intra- and interlobular fibrosis with intact exocrine acini and islets. Inflammatory infiltration was not significant. The pancreatic duct appeared patent (Fig. 2, 3). The liver revealed portal expansion with fibrosis, ductular proliferation and bile plugging. The gallbladder and cystic duct showed mild degree of fibrosis and chronic inflammation.

Table 1. Laboratory data

	Before operation						After operation		
	'88 8/2	8/9	8/19	9/6	9/8	9/15	9/17	9/30	'89 4/11
Total bilirubin (mg/dL)	8.5	5.5	1.4	6.3	8.4	6.8	3.4	1.3	0.5
Direct bilirubin (mg/dL)	0.7	0.9	1.0	5.9	5.8	5.0	2.1	0.7	0.1
Alanine aminotransferase (U/L)	305	80	67	126	162	157	146	39	
Aspartate aminotransferase (U/L)	257	136	91	101	135	162	138	57	
Alkaline phosphatase (U/L)	866	143.5	93.1	270.5		1678	449	369	
Cholesterol (mg/dL)					270	300	183	159	
Amylase (Unit)					170		95	75	

abdominal CT scan demonstrated 9×9cm-sized huge undefined retroperitoneal mass with markedly dilated biliary tree and gallbladder (Fig. 1).

An operation was done under the impression of retroperitoneal malignancy. On exploratory laparotomy, the pancreas was diffusely enlarged and "rubbery hard."

The pancreatic head had adhered to the duodenum and the distal common duct but was easily dissected from the surrounding tissue. The liver was enlarged and the common bile duct was dilated about 1.4cm in diameter. The walls of the common bile duct and gallbladder were extremely thickened. An operative cholangiogram demonstrated a dilated common bile duct and intrahepatic and extrahepatic biliary trees and no drainage to duodenum due to distal common bile duct obstruction. Two lymph nodes around the distal common bile duct near the pancreatic head, each measuring 1.2×1.2cm, were removed. After easy insertion of Baker's dilator #4 into the ampulla of Vater, drainage to the duodenum was excellent. Wedge biopsy from the enlarged pancreatic head and cholecystctomy were performed. A T-tube was inserted into the common bile duct.

Postoperatively, the jaundice was progressively resolved and the patient remained asymptomatic. A T-tube was removed on the 11th postoperative day. She was discharged in good general condition with almost normal liver function tests on the 12th postoperative day.

About seven months later, the patient's general appearance and activity were good. She gained 1.5kg of body weight. On repeat abdominal ultrasound and CT, no more mass shadow was detected, and the pancrease and biliary tree were normal in size (Fig. 4).

DISCUSSION

Idiopathic fibrosing pancreatitis is a very rare chronic disease in the pediatric age group characterized by interstitial fibrosis of the pancrease of undetermined cause (Williams et al., 1967).

The first reported case in the pediatric age group was an 11-year-old boy with chronic abdominal pain for two years (Comfort et al., 1946). Since then 16 cases of idiopathic fibrosing pancreatitis in the pediatric age group were reported. Most cases were as-

sociated with chronic abdominal pain, but Meneely et al. (1981) reported a case without abdominal pain. Alvear and Petro (1976) reported a first case of idiopathic fibrosing pancreatitis with obstructive jaundice.

Clinically, this disease is characterized by exacerbation and remission of abdominal pain and discomfort. The abdominal pain is the most constant symptom, transitory and mild at first. It locates anywhere in the upper abdomen, especially in the epigastrium (Williams et al., 1967). The progressive obstructive pattern of jaundice can be associated with chronic abdominal pain, as in our case. But painless obstructive jaundice was also reported as a primary manifestation of idiopathic fibrosing pancreatitis (Meneely et al., 1981)

The gross pathology of idiopathic pancreatitis is characterized by a diffusely or focally enlarged pancrease or a fibrotic, atrophic nodular pancreas (Meneely et al., 1981). Microscopically, diffuse collagenous tissue bands enclose normal-appearing acini. Pancreatic ducts are normal in caliber and free of metaplastic change. Islets of Langerhans are spared. Varying amounts of inflammatory reaction (polymorphonuclear leukocytes, lymphocytes and plasma cells) may be seen within the connective tissue and acini but are not prominent feature. There is no evidence of autodigestion such as fat necrosis, hemorrhage and pseudocyst formation (Williams et al., 1967). Our patient showed microscopic pancreatic features of mild intra-and interlobular fibrosis with intact exocrine acini and islets.

Abdominal ultrasound and CT demonstrate a diffusely or focally enlarged pancreas or a fibrotic, atrophic nodular pancreas and sometimes the presence of dilated bile ducts. Pancreas biopsy under the guidance of CT may confirm the diagnosis preoperatively (Atkinson et al., 1988)

Differential diagnosis in children with abdominal pain, obstructive jaundice and pancreatic enlargement includes acute and chronic pancreatitis and pancreatic carcinoma (Atkinson et al., 1988). Acute pancreatitis could be ruled out by normal serum amylase, no ultrasonic evidence of pancreatic swelling and inflammation such as pancreatic edema, inflammatory cells and fat necrosis (Weizman and Durie, 1988). The most common causes of chronic pancreatitis in the age group are hereditary pancreatitis, usually presenting in the second decade as an autosomal dominant with incomplete penetrance (Meneely et al., 1981). Forty families were reported in the literature, but no case had common bile duct obstruction (Forbes et al., 1984; Stafford and Grand, 1982). Three cases of common bile duct obstruction were reported in chronic pancreatitis in cystic fibrosis (Patrick et al., 1986). Primary carcinoma of the pancreas, although rare, has been reported in children demonstrating localized or diffuse enlargement of the pancreas and obstructive jaundice (Moyan et al., 1964). Our patient was not compatible with acute pancreatitis, had no family history of hereditary pancreatitis and demonstrated no malignancy on gross and microscopic examination.

Treatment of idiopathic fibrosing pancreatitis varies (Synn et al., 1987). The indications for surgery are an obstructive jaundice and recurrent severe abdominal pain (Meneely et al., 1981). Early surgery appears to offer two advantages: possible prevention of exocrine pancreatic nsufficiency and diabetes mellitus and normal life without frequent episodes of severe pain and hospitalization (Ghishan et al., 1983; Arvidsson et al., 1985). The choice of surgery depends on the anatomy of the pancreas and biliary trees (Ghishan et al., 1983). In case of complete bile duct obstruction, Roux-en-Y drainage and sphincterotomy were successfully done (Ghishan et al., 1983). In those cases of severe fibrosing process in the pancreas with normal biliary tree, direct pancreatic drainage is a rule: distal pancreatectomy with drainage by Duval (1954) or split pancreaticojejunostomy by Puestow (1958). Our case showed dramatic relief of abdominal pain and obstructive jaundice after the removal of the lymph nodes around the common bile duct and T-tube drainage. Our experience indicates that the choice of surgical procedure needs to be considered, case by case.

In summary, idiopathic fibrosing pancreatitis should be considered as a cause of obstructive jaundice and chronic abdominal pain in children. Abdominal ultrasound or CT is crucially important in demonstration of the diffusely or focally enlarged pancreas and dilated biliary tree. Surgical relief of the obstruction is mandatory.

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