Available online at www.sciencedirect.com
SCIENCE DIRECT
Hepatology Research 35 (2006) 147–150
HEPATOLOGY RESEARCH
http://www.elsevier.com/locate/hepres

Case report

# Ductal plate malformation and congenital hepatic fibrosis Clinical and histological findings in four patients

Olga Giouleme [a], Nikolaos Nikolaidis [a], Konstantinos Tziomalos [a,*], Kalliopi Patsiaoura [b], Themistoklis Vassiliadis [a], Nikolaos Grammatikos [a], Vassilios Papanikolaou [c], Nikolaos Eugenidis [a]

[a] *Gastroenterology Section of 2nd Propaedeutic Department of Internal Medicine, Aristotle University of Thessaloniki, Hippokration General Hospital, 63 Solonos Street, Thessaloniki 54248, Greece*
[b] *Department of Pathology, Aristotle University of Thessaloniki, Hippokration General Hospital, Thessaloniki, Greece*
[c] *Organ Transplant Unit, Aristotle University of Thessaloniki, Hippokration General Hospital, Thessaloniki, Greece*

Received 7 September 2005; received in revised form 20 November 2005; accepted 8 February 2006
Available online 10 March 2006

**Abstract**

Congenital hepatic fibrosis belongs to the fibrocystic diseases of the liver and represents ductal plate malformation of interlobular bile ducts, along with a destructive cholangiopathy associated with fibrosis. Four patients with congenital hepatic fibrosis are described. Their median age at presentation was 25 years; none of them had a family history of liver or renal disease. Variceal bleeding was the initial manifestation in three patients. All of them required frequent endoscopic variceal ligation sessions and distal splenorenal shunting was also performed in two, almost obviating the need for further variceal ligation. Variceal bleeding did not recur during follow-up. One of these three patients rarely exhibited acute cholangitis; administration of ursodeoxycholic acid resulted in complete remission. In contrast, the fourth patient showed frequent severe episodes of acute cholangitis but normal cholangiographic findings. He underwent liver transplantation but died 2 months later. Laboratory findings disclosed pancytopenia in all patients whereas hepatic synthetic capacity was well preserved. Renal function was unaffected despite the presence of polycystic kidneys in two patients. In summary, congenital hepatic fibrosis can also be diagnosed in older ages, might have strikingly different manifestations and is associated with prominent portal hypertension necessitating aggressive management in order to prevent variceal bleeding.
© 2006 Elsevier Ireland Ltd. All rights reserved.

*Keywords:* Congenital hepatic fibrosis; Fibrocystic diseases of the liver; Ductal plate malformation; Distal splenorenal shunting; Endoscopic variceal ligation; Portal hypertension

## 1. Introduction

Fibrocystic diseases of the liver are a group of conditions originating from abnormal persistence or defects in the progressive remodeling of the ductal plate. Ductal plate malformation (DPM) is now considered to be the basic component of these diseases and DPM at different levels of the biliary tree give rise to the wide spectrum of these disorders [1,2]. Congenital hepatic fibrosis (CHF) belongs to this group, is an autosomal recessive inherited condition and represents DPM of interlobular bile ducts, along with a superimposed destructive type of cholangiopathy (of variable speed and duration) associated with scarring fibrosis [2,3]. We report the clinical and histological findings of four patients who were diagnosed with CHF in our department.

## 2. Case series

Table 1 summarizes the main characteristics of the patients. Their median age at presentation was 25 years; none

* Corresponding author. Tel.: +30 2310823487; fax: +30 2310848354.
*E-mail address:* ktziomalos@yahoo.com (K. Tziomalos).

1386-6346/$ – see front matter © 2006 Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.hepres.2006.02.004

Table 1
Characteristics of the patients

| No. | Gender | Age at presentation (years) | Family history of liver or renal disease | Initial manifestation | Magnetic resonance cholangiography | Renal involvement | Follow-up (months) | Remarks |
|---|---|---|---|---|---|---|---|---|
| 1 | Male | 25 | Negative | Variceal bleeding | Normal findings | Polycystic kidneys | 26 | Splenorenal shunting |
| 2 | Male | 25 | Negative | Variceal bleeding | Normal findings | Polycystic kidneys | 70 | Splenorenal shunting |
| 3 | Female | 25 | Negative | Variceal bleeding | Normal findings | No | 72 | Episodes of acute cholangitis |
| 4 | Male | 44 | Negative | Acute cholangitis | Normal findings | No | 86 | Liver transplantation |

of them had a family history of liver or renal disease. Variceal bleeding was the initial manifestation in three patients. All of them required frequent endoscopic variceal ligation (EVL) sessions, approximately every 3 months, in order to obliterate their gastroesophageal varices. Distal splenorenal shunting was performed uneventfully in patients 1 and 2 approximately 8 and 30 months after the diagnosis; the frequency of EVL sessions has fallen significantly after the operation, and was only required once every year. During a follow-up time of 26, 70 and 72 months, respectively, none of these patients experienced variceal bleeding. Patient 3 also exhibited rare episodes of acute cholangitis but hospitalization was never required; ursodeoxycholic acid was administered at a dosage of 15 mg/kg per day and resulted in complete clinical and biochemical remission. Magnetic resonance cholangiography (MRC) was performed in all patients and showed normal findings, thus excluding the coexistence of Caroli's disease. Patient 4 showed a markedly different clinical course, characterized by frequent severe episodes of acute cholangitis that required prolonged hospitalizations. Both MRC and endoscopic retrograde cholangiography (ERC) were performed and showed normal findings, thus excluding the coexistence of Caroli's disease. He underwent orthotopic liver transplantation 7 years after the initial presentation and died 2 months later due to hepatic artery thrombosis. He never exhibited variceal bleeding and prophylactic EVL was performed only once.

Laboratory findings were within normal limits in all patients, except for thrombocytopenia and mild leukopenia and anaemia, consistent with the presence of portal hypertension-associated hypersplenism; hepatic synthetic capacity was well preserved (Table 2). It is noteworthy that renal function was also unaffected in all patients, despite the presence of polycystic kidneys in two of them (Table 2).

Liver biopsy was performed in all patients and similar histological findings were observed in all of them (Figs. 1 and 2). The portal tracts were enlarged due to severe fibrosis which was developed in them; the fibrous tissue was mature and collagenized. The other characteristic feature was the numerous uniform and generally small bile ducts, scattered in the fibrous tissue. Bile duct morphology and arrangement (anastomosing biliary channels) was consistent with the diagnosis of DPM. The ducts were lined by cuboidal to low columnar epithelium and some of them contained bile. Broad fibrous bands encircled single hepatic acini or groups of them. In

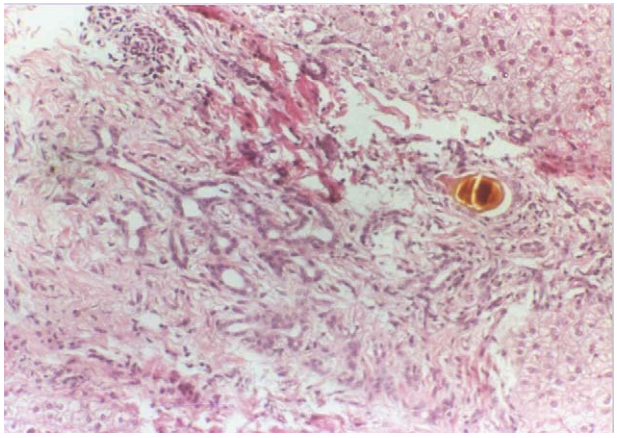

Fig. 1. Enlarged portal tract with small bile ducts, some of which are dilated and contain bile. Haematoxylin and eosin, ×200.

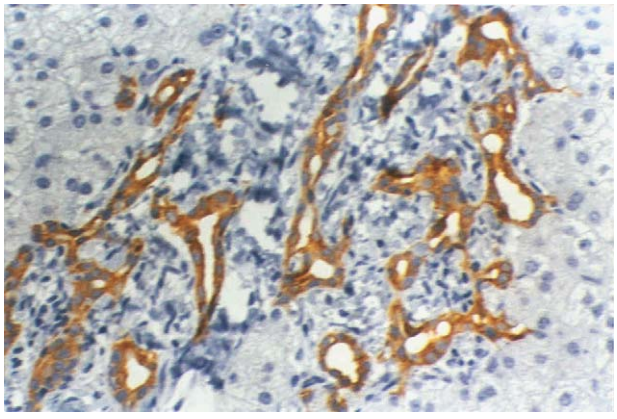

Fig. 2. Appearance of ducts which suggests DPM. PAP (Keratin 7), ×200.

immunohistochemical staining, both peripheral and central bile ducts showed strong immunoreactivity for cytokeratin (CK) 7 and CK19 whereas positive staining for CK8 and CK18 was observed in the hepatocytes; we did not use antibodies against CD34 in our immunohistochemical analysis.

## 3. Discussion

Our case series highlights several interesting aspects of CHF; this inherited fibrocystic disease of the liver might be diagnosed in older ages, family history of liver or renal

Download English Version:

https://daneshyari.com/en/article/3311555

Download Persian Version:

https://daneshyari.com/article/3311555

Daneshyari.com