CLINICOPATHOLOGICAL CASE

Congenital hepatic fibrosis: Complications and management

Fibrosis hepática congénita: complicaciones y manejo

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1. Summary of the clinical history (A-11-23)

We present the case of a 5-year-old male patient with fever, abdominal pain and icterus.

1.1. Family medical history

The patient’s mother is an apparently healthy 27-year-old female with a primary school education. She is Catholic, denies substance abuse, and lives with a partner in a free union. The patient’s father is an apparently healthy 35-year-old male who is a mason. He is also Catholic. There is a family history of diabetes mellitus and liver disease.

1.2. Social history

The family is originally from and resides in the DF (Federal District). They own a home with basic services. The family lives with a pet dog.

1.3. Perinatal and pathological history

The patient was the product of a second pregnancy with adequate prenatal care, normal ultrasonography, and intake of multivitamins. Pregnancy was of normal evolution and the patient was born at term, weighing 2700 g. Height and Apgar score are unknown. There were no perinatal complications. Patient was under treatment of the Hospital Infantil de México Federico Gómez (HIMFG) from January 2007 due to chronic liver disease with history of icterus since the first week of life.

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septic shock, and probable cholangitis.

normocytic anemia, hyperuricemia, thrombocytopenia, cirrhosis, grade III hepatic encephalopathy, normochromic

as follows: severe acute malnutrition, cryptogenic liver

treatment. Steroids were slowly decreased and stopped. IgG

He later presented with elevated transaminases despite

mottling, antiribosome 9.4.

were pH 7.31, pCO$_2$ 49,000, FA 558, BUN 36, calcium 7.7. Arterial blood gases

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lack of sleep during the night.

sleep-wake cycle of 24-h evolution. The mother reported

blood. He had sudden, progressive onset of cephalocaudal

movements of 24 h evolution on six occasions that were

sudden progressive onset of decreased consistency bowel

peritoneal irritation and globulous due to ascites. Peristalsis

pupils. Abdomen was soft, distended, depressible without

yellowish tinge with dry mucosa and hyporeflexic isocoric

liver and kidney) 2

was elevated, LKM (antimicrosomal antibodies type I of the

for 2 months. In August the dose of steroids was decreased

applied patients who have undergone liver transplantation, in
cases of malignancy are transplanted. The patient was also

It is possible to obtain a good quality of life and healthy

infections. In addition, the patient must undergo a careful

pathology. A diagnosis of partial bile flow

Evolution.

The patient's condition was treated

Diagnoses upon admission were

Laboratory findings were as follows:

The patient was also evaluated by the Transplant

Treatment was initiated with prednisone

+ ANA (antinuclear antibodies) 1:80 fine

A liver biopsy was performed, which

The patient was also evaluated by the Transplant

The patient presented with fever of 72 h evolution of sudden

The patient had progressive, diffuse, moderately intense abdominal

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7. Acute respiratory insufficiency syndrome—based on
6. Brain syndrome—based on alteration of the sleep/
5. GI hemorrhagic syndrome—manifested by repeated
4. Acute diarrheal syndrome with dehydration—according
3. Systemic inflammatory response syndrome—according to
2. Portal hypertension syndrome—manifested by ascites,
1. Chronic liver disease due to persistent elevation of

Based on the previously mentioned syndromatic diagnoses

The case presented is that of a 5-year-old male with multiple

Discharge diagnoses

The parents decided that no advanced resuscitation maneuvers

In the summary of the clinical history no evidence was

Liver cirrhosis can be a result of multiple causes. In the

This is a preschool-age child with baseline chronic liver

Clinically important toxins are another entity to rule

It is important to note that the patient should have been

Having documented neonatal cholestasis and

At 1 year 6 months of age. It is unknown if there was

This is a preschool-age child with baseline chronic liver

As the cirrhosis advanced, involvement of the liver

The fact that the patient had biliary cirrhosis from the

This is a preschool-age child with baseline chronic liver

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The parents as well as the poor short-term prognosis. The

Upon continuing with the diagnostic considerations, it was

Given the complexity

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Vitamin C deficiency is a cause of neonatal jaundice.

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The main complication presented by the patient was upper gastrointestinal (GI) bleeding secondary to intrahepatic portal hypertension. In the presence of a patient with the ascites, it is necessary to perform a liver biopsy. However, there is a disorder that combines the ascites with the acute liver failure that was not present in the patient. Another important complication that can affect the liver to such a degree as to cause fulminant hepatic failure, which is the liver disease. This is mandatory because of the characteristics already described as well as the presence of anti-microsomal, renal-hepatic type one and antinuclear antibodies which, at a point in time, required steroid treatment. Administration of platelet concentrates is recommended only in the case of bleeding. It is also not indicated to be used prophylactically in patients with portal hypertension and biliary cirrhosis, considered to be curative but palliative.

Based on what has been described, it is important to take into account the following diagnostic possibilities: hepatic veno-occlusive disease; GI bleeding with the purpose of correcting a coagulopathy; and when there is a platelet count less than 30,000 and a recommended amount of esophageal varices tends to aggravate the previously presented by the patient was ascites, which increased due to the decrease in osmotic pressure and increase of the renin-angiotensin-aldosterone system, which conditions a leak to the third space, principally at the peritoneal level. The main complication presented by the patient was upper gastrointestinal (GI) bleeding secondary to intrahepatic portal hypertension. In the presence of a patient with the ascites, it is necessary to perform a liver biopsy. However, there is a disorder that combines the ascites with the acute liver failure that was not present in the patient. Another important complication that can affect the liver to such a degree as to cause fulminant hepatic failure, which is the liver disease. This is mandatory because of the characteristics already described as well as the presence of anti-microsomal, renal-hepatic type one and antinuclear antibodies which, at a point in time, required steroid treatment. Administration of platelet concentrates is recommended only in the case of bleeding. It is also not indicated to be used prophylactically in patients with portal hypertension and biliary cirrhosis, considered to be curative but palliative.

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failure (Table 1).

As the multiple bleeding episodes, progressing to multi-organ deterioration, both due to the infectious processes as well as the decrease in leukocyte count and function, leading to a secondary state of immunocompromise compounded by the systemic factors, thus affecting the innate and adaptive system.

A decrease in immunoglobulins and complement should not be surprising given that liver failure conditions are known to be an essential part of the treatment of hepatic encephalopathy, with prolonged times of coagulation and massive pulmonary bleeding.

Anaphylaxis and hypoglycemia may occur due to the use of antibiotics such as rifaximine that have little enteral absorption with similar effectiveness as lactulose and fewer side effects.

It is recommended to begin with a visually high protein diet that can use antibiotics such as rifaximine that have little enteral absorption with similar effectiveness as lactulose and fewer side effects.

The use of lactulose was correct; however, it was initiated in the second week of hospitalization. In place of lactulose one can use antibiotics such as rifaximine that have little enteral absorption with similar effectiveness as lactulose and fewer side effects.

This is defined as acute liver failure in the context of a patient with chronic liver disease. Therefore, the matter would be to know if there was sufficient evidence to suspect autoimmune hepatitis. Currently, if the diagnosis of autoimmune hepatitis is suspected, the question would be the validity of treatment. As explained above, given the complexity of this case, it was not easy to reach an etiological diagnosis of liver disease. Therefore, the matter would be to know if there was sufficient evidence to suspect autoimmune hepatitis.

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3.1. Gastroenterology (Dr. Rodrigo Vázquez)

Within the management of hepatic encephalopathy, the option of hemodynamic stability has been assured.

It is important that once hemodynamic stability has been obtained, an adequate caloric intake is ensured in order to avoid catabolism. If the patient was not able to be completed during the 35 months after it was known by the majority of the services involved, it was not easy to reach an etiological diagnosis of liver disease. Therefore, the matter would be to know if there was sufficient evidence to suspect autoimmune hepatitis.

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risk of bleeding and the esophageal varices are treated, esophageal varices. If there are large gastric varices at the presence of gastric varices prior to treatment of the sclerosis should be continued. It is important to rule out bled. Pharmacological treatment and treatment based on is done in a patient who has varices that have already been concluded that variceal sclerosis in a patient who has been confirmed. The use of nonselective beta blockers in pediatric patients is still under discussion as opposed to adult patients in whom it has already been proven. As is often common, in the pediatric population as is often common, in the pediatric population.
Congenital hepatic fibrosis: Complications and management

The cytogenic alteration is a mutation in the short arm of chromosome 6 where the PKHD1 gene (of polycystic kidney and liver disease) is found. Dysfunction of this protein affects the bile ducts of the liver, the collecting tubules of the kidney and the pancreatic ducts (Fig. 3). Histologically, there is fibrosis in the portal spaces and formation of irregular, cystic ducts due to fusion of the ductal plate. The ductal plate is two layers of cells located around a portal vein during its fusion and remodelation. When there is lack of this protein, there is an increase in the number of bile ducts formed. Intrahepatic bile ducts, which are dilated, are tortuous and the epithelium is flattened. The portal veins also demonstrate changes; they are increased in number and are small.

In the pancreas, the ducts are dilated and have dense eosinophilic material in its lumen. Necrosis of the epithelial component and adipose tissue is also seen. The other organ affected is the kidney (Fig. 4). Both kidneys are increased in size and weight as has been described in autosomal recessive polycystic kidney disease. Histologically, small cysts originating in the collecting tubules covered with flattened cubic epithelium are observed and correspond to the terminal branches of the collecting tubules. In this disease, when the renal changes are more apparent, liver damage is less and vice-versa.

Due to liver damage, the kidney experiences other changes such as focal glomerulonephritis, mesangial proliferation due to IgA deposits, liver cirrhosis and liver failure, and damage due to bile material in the kidney tubules. Also, because of the liver damage, in the esophagus and stomach there are dilated and congested varices associated with ulcerated areas in the esophagus. In these areas there is fibrin, cellular detritus and yeast, which could correspond to Candida sp. Thirty five mL of blood was found in the stomach.

The lungs were increased in size and weight with extensive areas of reddish-brown color. Histological cuts demonstrated extensive areas of pulmonary hemorrhage. There were no data of infection found. Focally there were...
Hyaline membranes associated with alveolar damage. There was edema and dilated lymph vessels seen in the pleura. The spleen was increased in size and weight with chronic passive congestion.

In conclusion, this was a patient with congenital liver fibrosis in whom the renal and pancreatic damage was discretely present.

5. Final comments

5.1. Nephrology (Dr. Luis Velásquez Jones)

The majority of the studies in patients with chronic liver disease who developed a renal disorder have been done in adults. Very few studies have been carried out in the pediatric population. In adults, 50% of patients with cirrhosis have an IgA elevation. An IgA elevation was noted in this patient. It is known that adult patients with cirrhosis develop mesangial proliferation of the kidney. According to the immunofluorescence study, there are abundant IgA deposits found that correlate with the elevated serum levels of IgA observed in 90% of these patients. In this case the patient presented with hematuria and elevated IgA levels. It can be said with great certainty that he had an IgA nephropathy with creatinine elevation. This disease is uncommon although secondary IgA nephropathy has been reported in patients with biliary duct atresia. After renal transplant, in these patients the IgA nephropathy disappears.

Conflict of interest

The authors declare no conflict of interest of any nature.

References


Figure 4

Renal damage due to cirrhosis.