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CLINICOPATHOLOGICALCASE

Hepatic mesenchymal hamartoma

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OVERVIEW OF THE CLINICAL HISTORY (A-10-28)

We report on a 2-year-old male patient who presented due to an increase in abdominal girth.

Family Hereditary History. Mother is 36 years old, married, with secondary education. She is a housewife. She is Catholic, without any addictions and apparently healthy. Father is 35 years of age, married, with a secondary education. He is a merchant, Catholic, without addictions and apparently healthy. There are two brothers, ages 6 and 12, both apparently healthy. A sister died at birth likely due to neonatal hypoxia. Type 2 diabetes mellitus was reported for both maternal and paternal branches. One maternal aunt died due to complications of puerperium. Another maternal aunt died of leukemia at age 53. A maternal uncle died at 9 months of age due to an unknown cause but had lower extremity paralysis. Another maternal uncle died at 3 months due to pneumonia.

Non-medical Personal History. The family is originally from and resides in the State of Mexico. They live in a home with all basic services. They have two dogs and two cats as pets. Normal daily hygiene is reported.

Nutrition. The patient was breastfed until 9 months of age and was weaned at 5 months of age with mashed fruits and vegetables. At the present time he was integrated to the family diet. There is no past medical history.

Psychomotor Development. The patient demonstrated head support at 2 months of age, crawled at 7 months, walked at 1 year 3 months, monosyllabic at 9 months, and without bowel or bladder control.

Immunizations. Immunizations received are as follows: BCG (one dose), hepatitis B (three doses), acellular pentavalent (four doses), rotavirus (two doses), pneumococcus (three doses), influenza (three doses), and triple viral (one dose).

Perinatal and Medical History. The patient was the product of a planned pregnancy (G IV, P IV). Parents are nonconsanguineous and the mother received adequate prenatal care. Mother reported urinary tract infection during the first trimester. Three ultrasounds during the second and third trimesters were reported as normal. Folic acid and ferrous sulfate intake were reported from the first month. Vaginal delivery was accomplished. The patient cried and breathed at birth. Birth weight was 3,200 g, length 52 cm, and Apgar 8/9.

Perinatal and Pathological Background. Right hand polydactyly was detected. Neonatal icterus was treated with phototherapy. The patient was discharged at 4 days. Amputation of the first accessory digit of the right hand and circumcision were done. At 2 years of age, the mother noted an increase in abdominal girth. An abdominal ultrasound was done, which showed a large intrahepatic cyst, probably a liver abscess.

Abdominal CAT. Heterogeneous mass dependent on the right hepatic lobe (Figure 1) was demonstrated on abdominal CAT. The patient was referred for outpatient consultation with Oncology Surgery at the Hospital Infantil de México Federico Gómez (HIMFG). He was admitted to the Surgical Service for resection of the liver tumor.

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Table 1. Anthropometric characteristics

Weight	Length	HR	RF	BP	Temp.
13.5 kg	92 cm	100/min	22/min	90/50 mmHg	36.7ºC

FHR, heart rate; RF, respiratory frequency; BP, blood pressure.

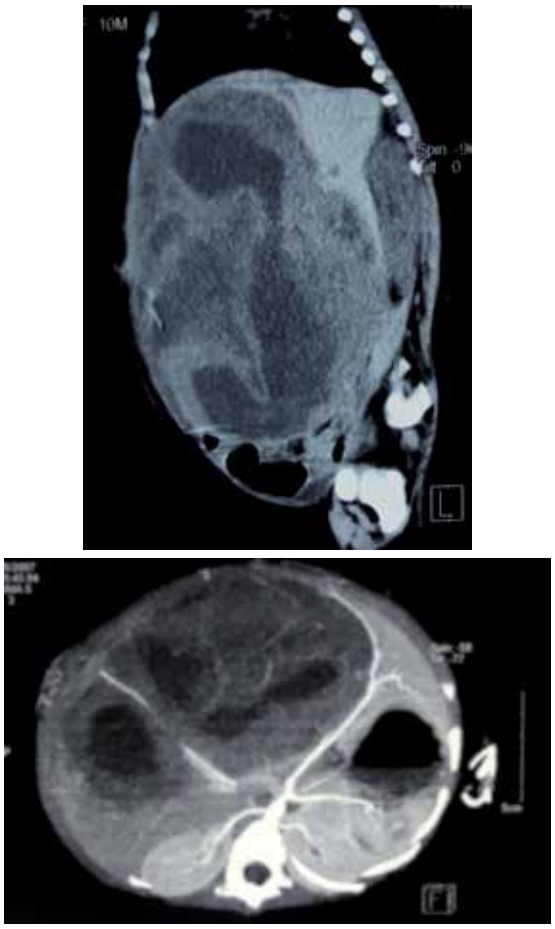

Figure 1. Heterogeneous lesion occupying segments of the right lobe of the liver and displacing the vascular structures.

Physical Examination. The patient was conscious, active and reactive (Table 1). He was normocephalic with round and reactive equal pupils, depressed nasal bridge, broad forehead, and cylindrical neck without masses or adenopathies. Cardiopulmonary system was without compromise and there were no murmurs, rales or wheezing. Abdomen was globular, soft, depressible, normal peristalsis, hepatomegaly 4-4-3 cm below the right costal margin, and without hepatodynia. Extremities were intact without pulses present and immediate capillary refill.

Surgery. Exploratory laparotomy and extended right liver lobectomy were carried out. A cystic liver tumor occupied the right lobe with ~200 ml of rust-colored fluid. A 5.5 Fr central venous catheter was placed 13 cm left subclavian at the first attempt (Table 2).

Intensive Therapy. Due to hypoperfusion, the patient required boluses of crystalloid and norepinephrine (0.1 µg/kg/min) with mechanical ventilation, base solutions of 1,800 ml/m^2 SC/day, clindamycin 40 mg/kg/day and amikacin 15 mg/kg/day; fentanyl 2 µg/kg/h and midazolam 4 µg/kg/min. Plasma was indicated for prolonged coagulation times with 100% of the Penrose drainage being replaced by 5% albumin. Penrose drainage was 1,059 ml/24 h.

April 13, 2010 (Intensive Therapy). The patient was extubated. Due to seizures he was administered diphenylhydantoin (DFH) at 20 mg/kg/dose and was reintubated. Penrose drainage was 74 ml/24 h.

Neurology. Change in the state of alertness, aphasia, no interaction with the environment, bilateral mydriasis, decreased strength, hyperreflexia, spasticity, bilateral prominent Babinski. Pyramidal syndrome and hypoxic ischemic brain disease.

Surgery. Central venous catheter exchange carried out.

April 15, 2010 (Infectious Disease). On the seventh postoperative day the patient had fever up to 38.4 ºC during 5 h without infection, and abdominal CAT was suggested (Figure 2).

Surgery. Enteral feeding was initiated. The patient presented abdominal distention and vomiting (Table 2).

April 16, 2010 (Intensive Therapy). The patient continued with neurological deterioration and fever without evidence of clear focus of infection (Table 2). Parenteral nutrition started. Probable pancreatitis.

Infectious Diseases. Fever probably not due to an infectious cause. Continued with the same management.

April 18, 2010 (Infectious Diseases). Nosocomial pneumonia associated with ventilator of late start (Table

Table 2. Laboratory results

March 24, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Plat	PT	PTT	INR			
12.5 g/dl	36%	5700	39%	48%	6%	6%	222,000	11.9”	26.7”	0.96			
April 8, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Plat	PT	PTT	INR	BUN	Cr	UA
18 g/dl	50.5%	13,500	70%	10%	10%	0	222,000	30.6”	44.8”	2.63	8 mg/dl	0.4 mg/dl	4 mg/dl
Na	K	Cl	Ca	P	Mg								
146 mEq/l	3.6 mEq/l	116 mEq/l	7.3 mg/dl	3.8 mg/dl	1 mg/dl								
BD	IB	TB	AP	TGP	TGO	GGT	LDH	Alb					
0.38 mg/dl	1.55 mg/dl	1.93 mg/dl	153 U	497 U	685 U	59 U	552 U	1.2 g/dl					
April 15, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Plat	BUN	Cr	UA	Na	K	
10.1 g/dl	28.6%	22,600	74%	22%	4%	0	250,000	9 mg/dl	0.4 mg/dl	0.7 mg/dl	138 mEq/l	4.4 mEq/l	
Cl	Ca	Mg	DB	IB	TB	TGP	TGO	Alb	Lipase	Amylase			
105 mEq/l	9.4 mg/dl	2 mg/dl	2.74 mg/dl	2.8 mg/dl	5.54 mg/dl	135 U	42 U	4.8 g/dl	878 U	96 U			
UA	pH	UD	Alb	Hb	Erithr	Scarse	Epithelial cells						
	7	25 mg/dl	50 mg/dl	0-1/c	3-4/c	bacteria	3-4/c						
pH	PaO_2	$PaCO_2$	HCO_3	SaO_2	BE	Lact							
7.42	128	24.9	15.2	99.9	-7.5	1.3							
April 18, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Plat	BUN	Cr	UA	Na	K	
9 g/dl	26%	19,100	82%	13%	2%	1%	280,000	5 mg/dl	0.3 mg/dl	1 mg/dl	137 mEq/l	4.1 mEq/l	
Cl	Ca	Mg	Procalcitonin	pH	PaO_2	$PaCO_2$	HCO_3	SaO_2	BE	Lact			
102 mEq/l	8.6 mg/dl	1.8 mg/dl	6.64 ng/ml	7.40	78.1	33.1	20.2	96.8	-3.8	1.2			
May 2, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Plat	PT	PTT	INR			
11.1 g/dl	32.2%	11,700	70%	23%	4%	2%	250,000	10.8”	18.5”	0.87			
BUN	Cr	UA	DB	IB	TB	GPT	GOT						
5 mg/dl	0.3 mg/dl	1 mg/dl	5.03 mg/dl	1.65 mg/dl	6.68 mg/dl	77 U	48 U						
May 3, 2010													
Hb	Hct	Leuk	Neu	Lymph	Mono	Eos	Platelets	PT	PTT	INR	Gluc	BUN	Cr
8.8 g/dl	26.9%	18,400	49%	32%	4%	0	74,000	20”	>120”	1.68	350 mg/dl	8 mg/dl	0.4 mg/dl
Na	K	Cl	Ca	Mg	DB	IB	TB	GPT	GOT	Alb			
145 mEq/l	4.9 mEq/l	111 mEq/l	9.5 mg/dl	1.4 mg/dl	2.15 mg/dl	1.61 mg/dl	3.76 mg/dl	716 U	1329 U	3.1 g/dl			
pH	PaO_2	$PaCO_2$	HCO_3	SaO_2	BE	Lact							
7.06	45	68.7	18.6	73	-10.2	8.5							

Hb, hemoglobin; Hct, hematocrit; Leu, leukocytes; Neu, neutrophils; Lymph, lymphocytes; Mono, monocytes; Eos, eosinophils; Plat, platelets; PT, prothrombin time; PTT, partial thromboplastin time; INR, international normalized ratio; BUN, blood urea nitrogen; Cr, creatinine; UA, uric acid; DB, direct bilirubin; IB, indirect bilirubin; TB, total bilirubin; AP, alkaline phosphotase; GPT, glutamate pyruvate transaminase; GOT, glutamate oxalate transaminase; GGT, gammaglutamyl transferase; LDH, lactate dehydrogenase; Alb, albumin; UA, urinalysis; UD, urinary density.

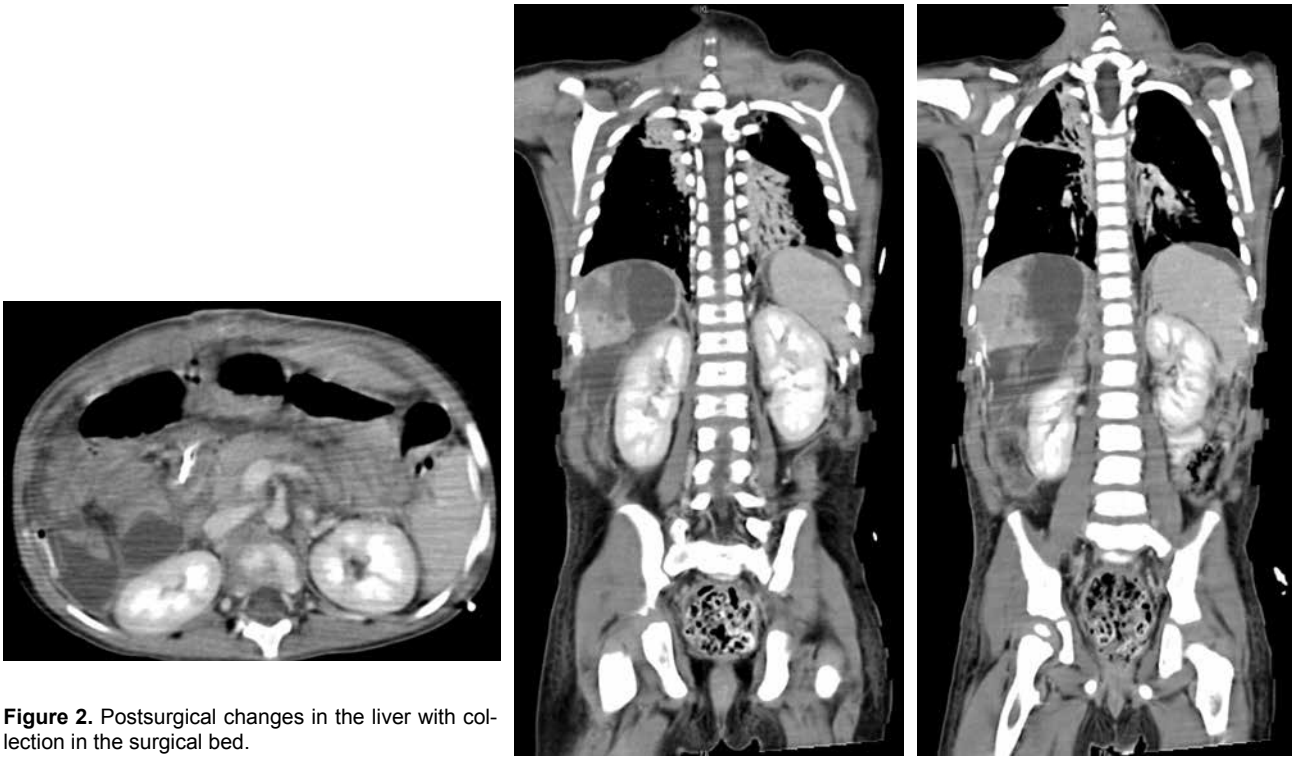

Figure 2. Postsurgical changes in the liver with collection in the surgical bed.

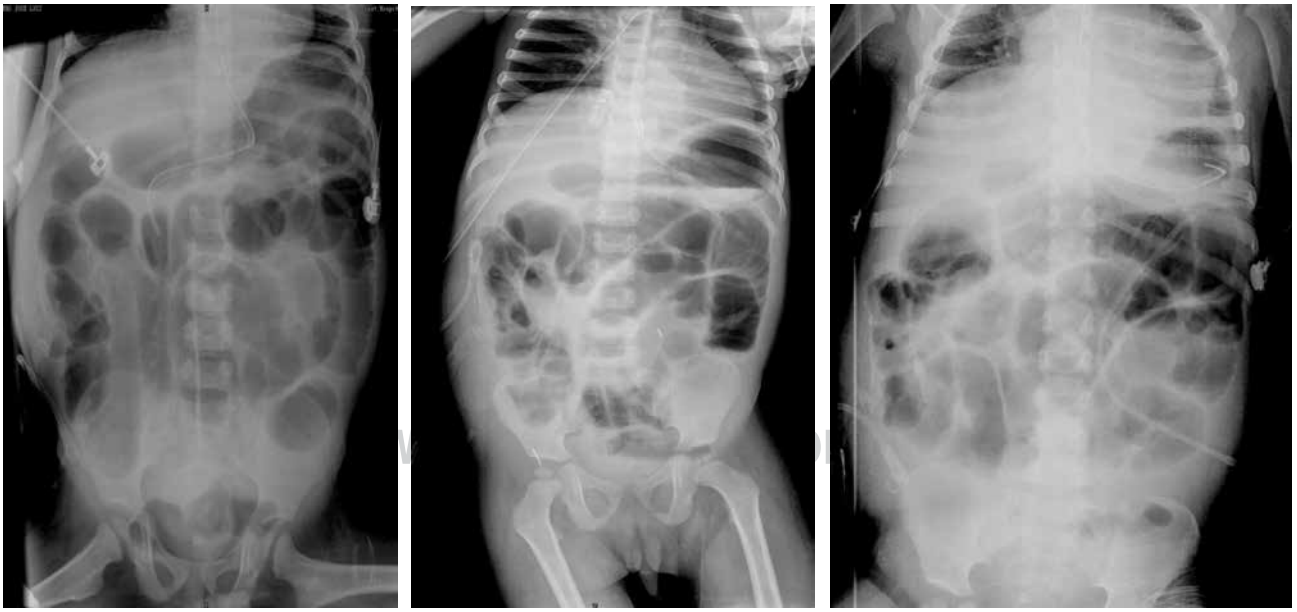

Figure 3. Plain films that demonstrate loop dilation, which suggests intestinal ileus.

2). Cefepime was started (150 mg/kg/day) and amikacin (15 mg/kg/day). X-rays were taken (Figure 3).

April 19-20, 2010 (Intensive Therapy). Output from the Penrose had biliary characteristics.

Surgery. Penrose output of 860 ml with biliary characteristics.

Infectious Diseases. Probable bilioenteric fistula. Coverage was widened for *Enterococcos* sp. with ampicillin at 100 mg/kg/day.

April 21-22, 2010 (Intensive Therapy). Removed from mechanical ventilation.

April 23, 2010 (Surgery). With adequate tolerance to liquid diet, diet was progressed and amikacin was stopped.

April 24, 2010 (Surgery). Central blood culture. *S. hominis*. CVC was removed, and the patient continued with fever.

April 26, 2010 (Surgery). Biliary secretion culture: *S. aureus*.

April 28, 2010 (Surgery). Left basal hypoventilation, mild expiratory wheezing, micronebulizations with salbutamol. Penrose drainage 56 ml, biliary. Completed 17 days with cefepime and was stopped; continued with ampicillin.

May 3, 2010 (Surgery) 15:30 h. Biliodigestive bypass in "cobra head," end to end anastomosis, and invaginating appendectomy (Table 2). Multiple loop to loop, loop to wall friable flanges were found that easily. A bile leak was not located. Penrose at the site of biliodigestive bypass. Bleeding 500 ml. Patient was transfused with 400 ml red blood concentrate. Admitted to surgical therapy. Bleeding via the Penrose 400 ml with hypotension, delayed capillary refill and peripheral pulses not palpable.

Surgery 19:26 h. Exploratory laparotomy, packing and Bogota pouch placement. Layered and profuse bleeding was found in the surgical bed, abdominal wall and raw liver area with hemoperitoneum of ~250 ml.

Anesthesia. The patient was admitted with adrenalin (0.1 µg/kg/min) and vasopressin (0.2 µg/kg/min). He had a cardiorespiratory arrest, electrical activity without pulse. Various advanced resuscitation maneuvers were carried out, adrenalin and calcium gluconate 1 g were administered. There was a new cardiac arrest and asystole which was managed with adrenaline and sodium bicarbonate. He presented ventricular fibrillation and was defibrillated with return to sinus rhythm.

May 4, 2010 (Intensive Therapy) 03:20 h. Oliguria, weak pulses, generalized pallor, capillary refill time of 3 sec. Management with adrenalin and milrinone was added with slight improvement. Patient finally presented cardiorespiratory arrest without response to resuscitation maneuvers.

CASE PRESENTATION

(Dr. Jorge Cortés Sauza)

The clinical family history in relation to the death of family members is considered of importance, not necessarily directly related to the primary condition. The syndromatic diagnoses were established as mentioned below:

1. Abdominal mass dependent on the liver. It is unknown if it was solid, mobile or with or without limiting margins. During its evolution the patient was evaluated outside this institution. An ultrasound was performed and reported a giant cystic mass. It was complemented by computerized tomography that reported a heterogeneous tumor in the right lobe of the liver (Figure 1).
2. Convulsive syndrome due to crisis after the first surgical event. The patient was managed with DFH at conventional doses that warranted evaluation and follow-up by the Neurology Department.
3. Infectious syndrome characterized by sustained temperature elevations up to 38 ºC. At times, fever was accompanied by tachycardia and tachypnea as well as having data of hypoperfusion with alterations in blood count with leukocytosis and bandemia and prevalence of neutrophils.
4. Cholestasis shown by elevation of total bilirubin with predominance of direct bilirubin and with a progressive postsurgical behavior towards increasing.

The principal diagnoses are as follows:

1. Liver tumor:

Liver tumors are classified into two large groups-the malignant and benign. Malignant tumors are the third most frequent abdominal tumors in the pediatric age after neuroblastoma and Wilms tumor. The most frequent liver tumors are hepatoblastoma, liver sarcoma and hepatocarcinoma. Benign tumors represent up to 30% of liver masses and are divided into hemangiomas, hamartomas, mesenchymal and focal hepatic nodular hyperplasia. The

study of the patient with a liver mass should be initiated with a plain film of the abdomen to see calcifications, abdominal air distribution, data on compression of structures or data of secondary intestinal occlusion. Subsequently, Doppler abdominal ultrasound should be carried out to see sonographic characteristics, i.e., cystic, solid, vasculature, presence of thrombus and compromise of the intra- or extrahepatic bile duct.[1] There are then two alternatives: CAT and MRI. In this case, the former was utilized. CAT assists with the anatomic description: the extent, volume, presence of thrombus in the portal vein or vena and presence of distance metastasis (Figure 2). It is believed that in this patient there are two diagnostic possibilities. First there is the hepatic mesenchymal hamartoma that represents ~6% of liver tumors. It is rare and is mainly present in the right lobe of the liver. The most frequent age at presentation and diagnosis is up to 2 years of age. For those cases there may be three types of surgical approaches. One may be nonanatomic enucleation, another is marsupialization with unroofing of the cyst and placement of sclerosing agents in the area and, finally, wide resection of the lesion. According to the literature, it has been noted that this lesion can have a sarcomatous component. For this reason, the technique of choice should be complete wide resection and, if possible, with negative margins. Another probable tumor is liver sarcoma, which can be seen in 6% of malignant liver tumors. The average age at diagnosis is usually school-age children. It presents rapid and progressive growth; therefore, it can begin as a mass with liver rupture. In accordance with the approach per the image, the hamartoma can be seen as a poorly vascularized cystic lesion such as liver sarcoma, although it mainly presents solid tissue.[1,2]

It must be kept in mind that surgical management has its complications. The most frequent complication is bleeding, which could be active but not severe. The severe form may present itself in <2% of the processes. Bile leakage from the surgical bed, which could cause a biliary fistula, is observed in <5% of the cases. Another reported complication could be intestinal occlusion or sepsis. Hepatic insufficiency, which may not be recoverable, may require a liver transplant in <5% of cases.[3,4]

2. Hypovolemic shock:

After the first surgery, during postsurgical care the patient presented hypotension and hypoperfusion and required management is crystalloids and vasopressors. Hospital stay in intensive therapy was 14 days with Penrose output up to 800-1,200 ml and up to 20 days with bile characteristics between 500 and 1,000 ml in 24 h accompanied by progressive total cholestasis up to 6.6 mg/dl with elevation of transaminases after the surgical procedure with an intermittent behavior. Twenty-four days after the first surgical procedure, due to the presence of a bile leak, a biliodigestive bypass was performed in "cobra head," which consists of lifting a bowel loop to the surgical bed where a cut in the liver is done. Unfortunately, the patient presented postoperative bleeding. He also presented data of hemodynamic compromise with hypoperfusion, hypotension, prolonged capillary refill, and non-palpable pulses. Four hours later, urgent exploratory laparotomy was performed due to Penrose drainage of 400 ml. A bleeding surgical bed was found. The patient was most probably in a stage of consumption; therefore, it was decided to pack bleeding areas and placement of a Bogota bag with the consequent risk of progressing to compartment syndrome. During the third surgery the patient had two cardiorespiratory arrests. He recovered from these events; however, 8 h later in the surgical recovery unit he had mixed acidosis, hyperlactatemia and hypoxemia, anemia, thrombocytopenia and prolonged coagulation times.

3. Disseminated intravascular coagulopathy:

Clinically, due to bleeding in the surgical bed and abdomen, the patient required the indicated therapy with prolonged coagulation times, decrease in fibrinogen and thrombocytopenia.

4. Convulsive crisis:

He was evaluated by neurology with magnetic resonance and electroencephalogram, taking into consideration a pyramidal syndrome due to hypoxia.

5. Sepsis:

From the patient's management, he presented sustained temperature elevations. Antimicrobial therapy was changed from cefepime to amikacin. Bacteria were isolated from the catheter and in the Penrose fluid. No lumbar puncture was done, which would have been useful for the risk factors of hospitalization and invasion and neurological deterioration. With this in mind the following final diagnoses were made:

1. Benign liver tumor: hepatic mesenchymal hamartoma
2. Hypovolemic shock
3. Disseminated intravascular coagulation

PATHOLOGY DEPARTMENT (Dr. Stanislaw Sadowinski Pine)

Histopathological Findings

The right lobe of the liver was received for histopathological studies and weighed 100 g and measured 16 x 15 x 7 cm. The external surface was covered by a smooth, shiny Glisson capsule. The gallbladder was identified on its posterior face without alterations; macroscopically the surgical margins were without tumor (Figure 4). On serial cuts, a well-defined nonencapsulated, solid, heterogeneous tumor was observed, demonstrating a yellowish-white cyst with pearly areas. The cysts were variable in size. The largest was 9.5 cm in diameter and contained whitish material and others showed rust-colored fluid. Histological cuts demonstrated cysts covered by biliary epithelium surrounded by mesenchymal tissue separated by hepatocyte trabecula. On Masson trichromic stain there was proliferation of cholangioles and expansion of dense connective tissue in the portal spaces of the residual liver tissue (Figure 5).

The left lobe of the liver weighed 400 g and demonstrated a smooth surface. Accentuation of lobular pattern and greenish color was observed on the cut after fixation. Microscopic examination showed ductal and cellular cholestasis, proliferation of cholangioles and fibrous septa that united various portal spaces, secondary to a partial obstruction of bile flow, cholangitis, and data of shock characterized by perivenular hemorrhagic necrosis and sinusoidal congestion. There was no residual tumor (Figure 6). Portal fibrosis could be seen in the ductal plate abnormalities, although in this case they are believed to be secondary to obstruction of bile flow due to the tumor and the damage to the bile duct.

At autopsy, the abdomen was observed with a Bogota bag containing 75 ml of free blood as well as packing material, and 185 ml of blood in the cavity. The biliodigestive bypass in "cobra head" had intact sutures, and a bile deposit was observed on the Glisson's capsule and soft tissues adjacent to the anastomosis along with inflammatory infiltrate to foreign body and fibrosis (Figure 7). Intestinal serosa was thickened by fibrosis and mixed inflammatory infiltrate, and mucosa showed extensive areas of superficial hemorrhage. Lungs showed vascular congestion, focal collapse and focal pneumonitis. The arterioles showed fibrin and septic thrombus. Also observed were changes secondary to shock in the heart, intestine, kidney and pancreas. The brain showed a decrease in weight, data of edema with widening of the convolutions, dilation of the lateral ventricles due to atrophy and isolated necrosis of the neurons. The thymus showed acute involutional changes and cystic Hassall corpuscles with abundant cellular debris.

Liver tumors in children are uncommon. The records of liver tumors from the Pathology Department were reviewed. During a 13-year period, there were 179 cases recorded (corresponding to 0.6% of the total tumors). The most common tumors, after hepatoblastoma, were mesenchymal hamartoma and hepatocellular carcinoma (Table 3). Frequency of these tumors in the HIMFG agrees with

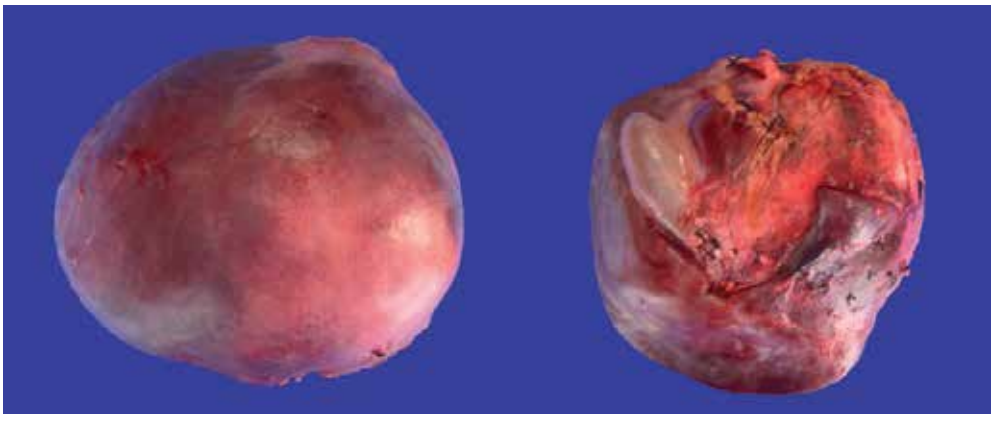

Figure 4.

Result of the extended right hepatectomy with intact surface, bile duct and surgical margin without tumor.

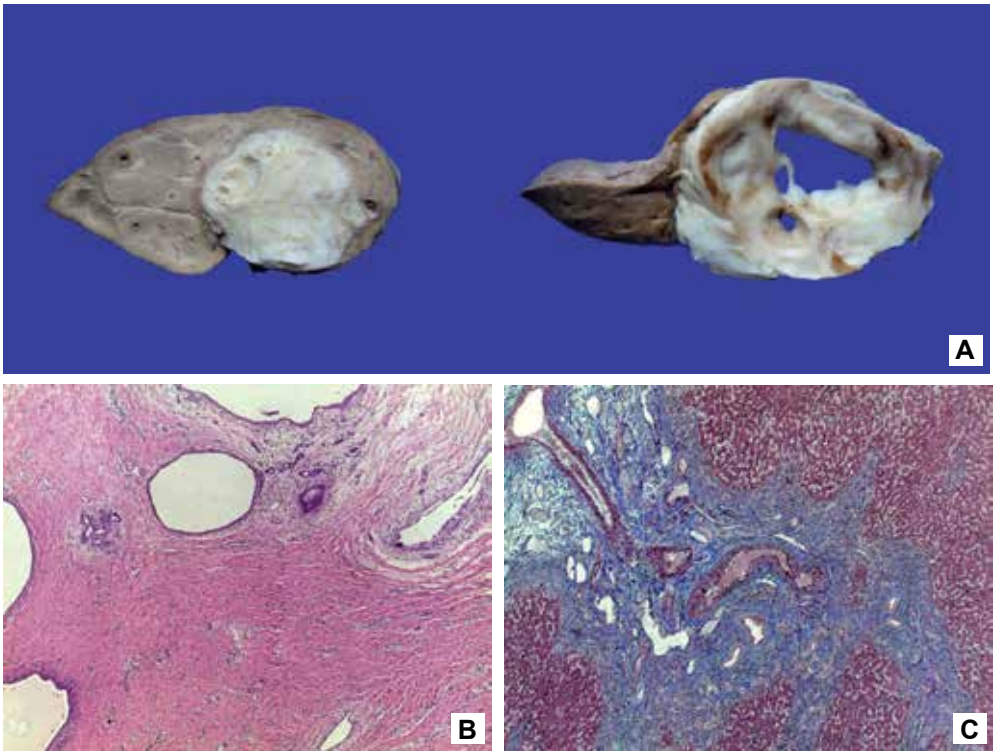

Figure 5.

(A) Solid well-defined and cystic tumor (11.5 x 9 cm) surrounded by congestive liver parenchyma. **(B)** The tumor has cysts covered by biliary epithelium, separated by dense connective tissue (HE x20, adjustable objective [AO]). **(C)** Portal space with fibrous expansion and proliferation of cholangioles (Masson x20 AO).

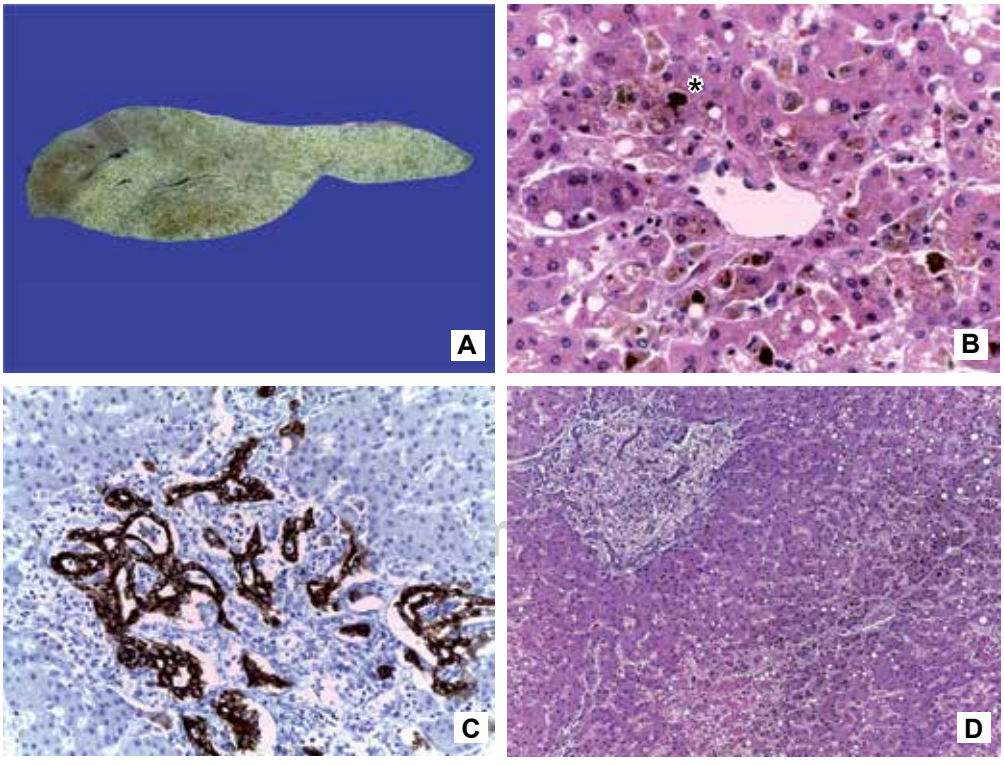

Figure 6.

(A) Left lobe of the liver without residual tumor and greenish color. **(B)** Hepatocytes with bile in the cytoplasm and bile plugs in canaliculi (*). **(C)** Proliferation of cholangioles in the portal space expressing CK7 (immunohistochemistry x40 AO). **(D)** Liver parenchyma with centrolobular necrosis and macrovesicular steatosis (HE, x20 AO).

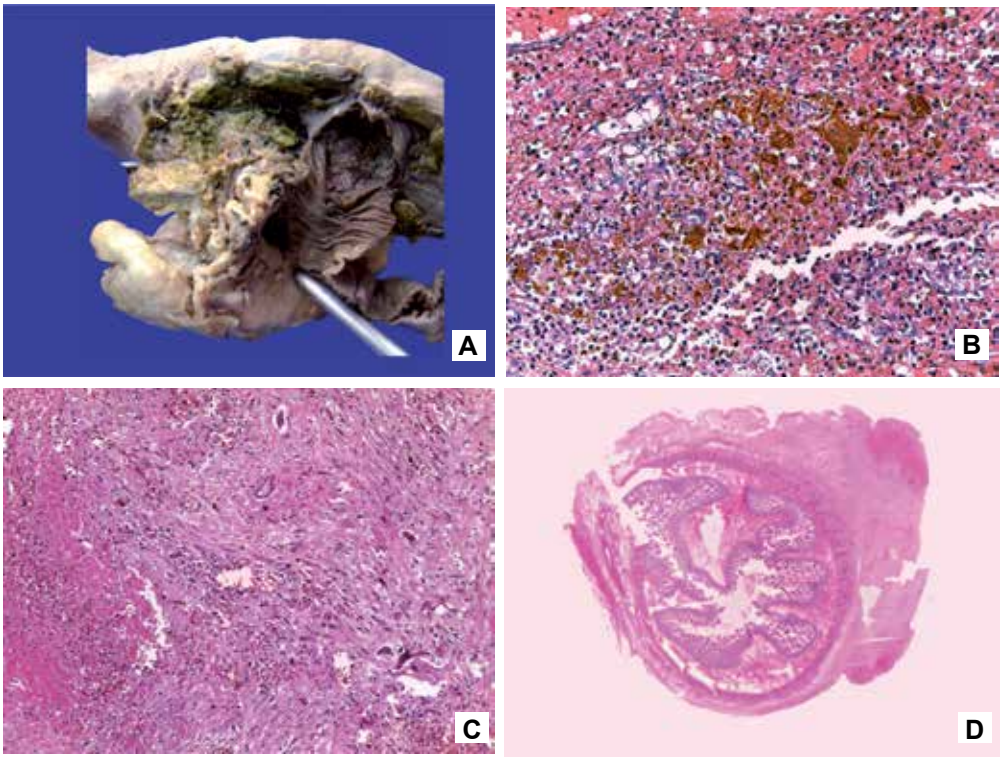

Figure 7.

(A) Anastomosis in "cobra head," adjacent fibroadipose tissue with bile deposits. **(B)** Biliary pigmentation surrounded by inflammatory infiltrate and fibrin (HE x40 AO). **(C)** Fibrosis and hemorrhage in the fibroadipose tissue (HE x20 AO). **(D)** Transverse cut of the small intestine with acute and chronic peritonitis.

Table 3. Liver tumors registered in the Hospital Infantil Federico Gómez, 2000-2012 (Department of Clinical and Experimental Pathology)

	No. of tumors	%
Liver tumors	179	0.60*
Hepatoblastoma	108	60.34**
Mesenchymal hamartoma	14	7.82**
Hepatocellular carcinoma	14	7.82**

*With respect to total tumors.
**With respect to liver tumors.

what has been reported in other series.[5] Hepatic mesenchymal hamartoma is a rare, benign tumor that presents itself in neonates and infants. Complete surgical resection is curative. It has recently been reported that patients with hepatic mesenchymal hamartoma present recurrent cytogenetic changes with translocation of 19q13.4 and deletion of this locus[5,6] as well as monosomies 2, 3, 4, 9, 10 and 22.[7] Morphological changes are similar to mesenchymal dysplasia of the placenta, so these alterations may be associated. There have also been cases reported of patients with Beckwith-Widemann syndrome and hepatic mesenchymal hamartoma.[8] It is believed that this group of alterations corresponds to a possible imprinting defect.[9] This is the first case study with autopsy in the Clinical and Experimental Pathology Department of the HIMFG. The death of the patient was related to surgical complications, which led to a consumption coagulopathy and intraabdominal bleeding.

Final Diagnoses

Principal disease

1. Completely resected right hepatic mesenchymal hamartoma

Concomitant disorders

1. Localized biliary peritonitis secondary to biliary fistula
2. Status postbiliodigestive bypass in "cobra head," end to end anastomosis and invaginating appendectomy
3. Status postexploratory laparotomy and abdominal packing
4. Hemoperitoneum 260 ml (75 ml in Bogota bag)
5. Left lobe of the liver with changes secondary to partial obstruction of bile flow and moderate fibrosis
6. Acute cholangitis
7. Sepsis due to *Staphylococcus aureus*

8. Bilateral pleural effusion (right side 140 ml; left side 40 ml)
9. Anatomic data of shock and disseminated intravascular coagulopathy. Hepatic periventricular necrosis, acute tubular necrosis, ischemic hypoxic encephalopathy, ischemic hypoxic visceral myopathy, pulmonary fibrin thrombus

Independent Alterations

1. Right-hand polydactyly
2. Clinical history of amputation of first finger of the right hand
3. Accessory spleen

COMMENTARY

(Dr. Pablo Lezama del Valle)

It must be clear that faced with any tumor or palpable mass in the abdomen in a pediatric patient, the patient should be hospitalized for studies and treatment to rule out malignant tumor as soon as possible. The absence of signs such as lack of weight gain, fever, and lymphadenopathy does not rule out the possibility of cancer because in various malignancies a palpable mass may be the only clinical data. The most useful initial screening study is abdominal ultrasound. In a patient of this age with a solid liver tumor, the first possibility would be a hepatoblastoma. A heterogeneous tumor could be a hamartoma or focal hepatic hemangiomal (hemangioendothelioma), although for the latter a Doppler would demonstrate data of characteristic vascular flow. In this patient, the first suspicion was a hepatic hamartoma corroborated with simple and contrast CT study. It was a complex, predominantly cystic tumor that affected the right lobe of the liver and mid-portion of the left lobe, i.e., corresponding to Couinaud segments I, IV, V, VI, VII and VIII-leaving only the left lateral segment free or segments II and II. Treatment of this benign tumor of the liver ideally consists of complete excision by means of anatomic resection. It was a very large tumor. In order to perform an extended or wide right lobectomy, also known as trisegmentectomy (although this terminology causes some confusion with the already mentioned Couinaud segmentation), the cystic portion was partially decompressed and was very close to the left hepatic vein in its most proximal portion. During hepatic resection, the work of the anesthesiologist is fundamental. The anesthesiologist should foresee the possibility of massive bleeding on an overloaded patient and replace the losses with blood products as required, using crystalloids with moderation. It is fundamental to prevent acidosis and hypothermia. Ligation of the right branch of the portal vein, right hepatic artery and right hepatic vein was carried out, as well as of the veins that drain directly into the inferior vena cava. To resect the parenchyma the afferent flow was interrupted with a Pringle maneuver. The surgery was carried out successfully with interruption of the hepatic vascular flow and bleeding control. To corroborate that there was no active bleeding before closure, an invaginating appendectomy was carried out after waiting a few minutes. The incision was closed once the patient was replaced with the volume lost and it was confirmed that there was no bleeding. The patient was taken to intensive therapy with assisted ventilation. After a few hours there was clinical evidence of bleeding at the surgical site, based on hemodynamic changes as well as decrease in the hematocrit count and blood loss via the abdominal drain. The patient was transferred to the operating room for a new exploration with the findings of bleeding in layers, for which liver packing was carried out and a laparostomy bag placed (Bogota bag). For the coagulopathy, which was not reversed with blood transfusions, and even with administration of recombinant factor VII, the patient continued to bleed in an uncontrolled manner and then expired.

I would like to mention that in the HIMFG a large number of liver resections are carried out primarily for malignant tumors, and intraoperative mortality has been significantly decreased. During the immediate postoperative period, an outcome such as this is extremely uncommon.

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REFERENCES

1. Bisogno G, Pilz T, Perilongo G, Ferrari A, Harms D, Ninfo V, et al. Undifferentiated sarcoma of the liver in childhood: a curable disease. Cancer 2002;94:252-257.
2. Wei G, Tang LF, Chen ZM, Tang HF, Li MJ. Childhood undifferentiated embryonal liver sarcoma: clinical features and immunohistochemistry analysis. J Pediatr Surg 2008;43:1 912-1919.
3. Pérez-Gómez RM, Herrera-Medina H, de León-Bojorge B, Ortiz-Hidalgo C. Sarcoma indiferenciado embrionario de

hígado. Estudio clínico-patológico e inmunohistoquímico de ocho casos con énfasis en el diagnóstico diferencial con tumores intraabdominales en niños y adultos jóvenes. Patología 2011;49:25-32.
4. Gupta DK, Carachi R. Pediatric Surgical Oncology. McGraw-Hill Education; 2008.
5. Rakheja D, Margraf LR, Tomlinson GE, Schneider NR. Hepatic mesenchymal hamartoma with translocation involving chromosome band 19q13.4: a recurrent abnormality. Cancer Genet Cytogenet 2004;153:60-63.
6. Talmon GA, Cohen SM. Mesenchymal hamartoma of the liver with an interstitial deletion involving chromosome band 19q13.4: a theory as to patoghenesis? Arch Pathol Lab Med 2006;130:1216-1218.
7. Baboiu OE, Saal H, Collins M. Hepatic mesenchymal hamartoma: cytogenetic analysis of a case and review of literature. Pediatr Dev Pathol 2008;11:295-299.
8. Cajaiba MM, Sarita-Reyes C, Zambrano E, Reyes-Múgica M. Mesenchymal hamartoma of the liver associated with features of Beckwith-Weidemann syndrome and high serum alpha-fetoprotein levels. Pediatr Dev Pathol 2007;10:233-238.
9. Reed RC, Kapur RP. Hepatic mesenchymal hamartoma: a disorder of imprinting. Pediatr Dev Pathol 2008;11:264-265.