



Case Report: Primary Liver Mature Teratoma in Childhood

Reporte De Caso: Teratoma Maduro Hepático Primario en la Infancia

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Resumen

Presentamos el caso de un paciente de 2 años de sexo masculino, que presentó una masa abdominal asociado a distensión y dolor abdominal de leve intensidad. Cursó con valores normales de perfil hepático, así como de Alfafetoproteína, Gonadotrofina coriónica y antígeno carcino embrionario. La tomografía computarizada de abdomen y la resonancia magnética nuclear de abdomen reportaron una lesión hepática expansiva de predominio quístico con algunos componentes sólidos internos. Se valoró el caso y se decidió su intervención quirúrgica, realizándose hepatectomía derecha y derivación biliodigestiva por elongación y compromiso del colédoco, con una evolución favorable. El teratoma hepático maduro primario es un tumor raro en la población con mayor incidencia en niños, por lo que debe sospecharse en todos los pacientes con tumores hepáticos sólidos que no presenten valores elevados de AFP.

Palabras clave: Teratoma hepático; niños; hepatectomía; derivación biliodigestiva; alfafetoproteína.

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Abstract

We present the case of a male 2-year-old patient who underwent surgery for a 13-cm-long liver teratoma that caused elongation of the common hepatic duct and the portal vein. The boy had an abdominal mass and abdominal distension with pain of mild intensity. Abdominal computed tomography and abdominal nuclear magnetic resonance reported a predominantly cystic expansive liver lesion with some internal solid components. The case was evaluated and surgical intervention was decided, performing a right hepatectomy and biliodigestive derivation due to elongation and compromise of the common bile duct. Primary mature liver teratoma is a rare tumor in population with a higher incidence in children, so it should be suspected in all patients with solid liver tumors that do not present with high AFP values.

Keywords: Liver teratoma; children; hepatectomy.

Introducción

The origin of the word teratoma comes from the Greek “teratos” which means monster. A teratoma derives from 2 or more germ cells: ectoderm, mesoderm or endoderm.¹ Teratomas are classified as mature and immature, with a slow growing rate (<2 mm/year).² Primary liver teratoma (PLT) represents less than 1% of documented tumors located in the liver, with less than 50 cases reported worldwide, mostly in children under 3 years old.³

The present study features the case of a 2-year-old male patient presenting an abdominal mass in right hypochondrium and flank. The mass was measured finding a 10 x 7 cm tumor associated with mild abdominal pain, said tumor underwent right hepatectomy. This is a rare case, with risk of malignancy. However, there is still no literature that reports a similar case of PLT in pediatric patients. This case report was approved by the ethics committee of the sponsoring institution for publication.

Clinical case

This study presents the case of a 2-year-old boy who came into the emergency room,

he presented an abdominal mass in right hypochondrium and right flank that grew towards the right iliac pit, associated with distension and mild abdominal pain. He had normal liver profile values, as well as alpha-fetoprotein.

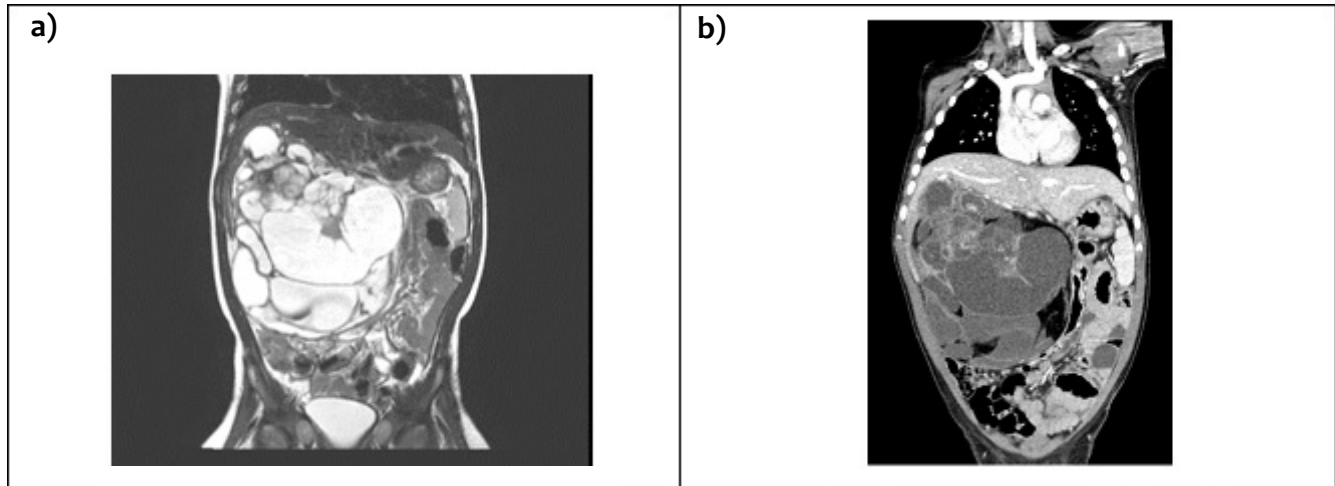
Physical examination revealed a solid mass of 10 x 7 cm that was not mobile, with regular edges and located in the right upper quadrant of the body/abdomen? Laboratory tests showed a normal blood count, with negative AFP, BHCG and CEA tumor markers, liver enzymes within normal values and slightly elevated C-Reactive Protein.

Abdominal computerized tomography scan and abdominal nuclear magnetic resonance test shown a predominantly cystic expansive liver lesion with some internal solid components in segments V, VI, VII and VIII. The tumor had a multilocular appearance with calcium and fat content inside and with the presence of bile ducts that entered the tumor with small vascular branches from the hepatic artery and portal vein that irrigate the capsule of the tumor in its superior aspect. The main portal vein was collapsed and displaced posteriorly by the tumor, parallel to and anterior to the inferior vena cava. The drainage of the suprahepatic

veins was normal. The gall bladder was previously displaced by the tumor. Mass effect and compression of the head of the pancreas was observed, which was in close contact with

the tumor in its lower left lateral aspect. The first and second portions of the duodenum were in contact with the tumor and the transverse colon with inferior displacement. (Figure 1).

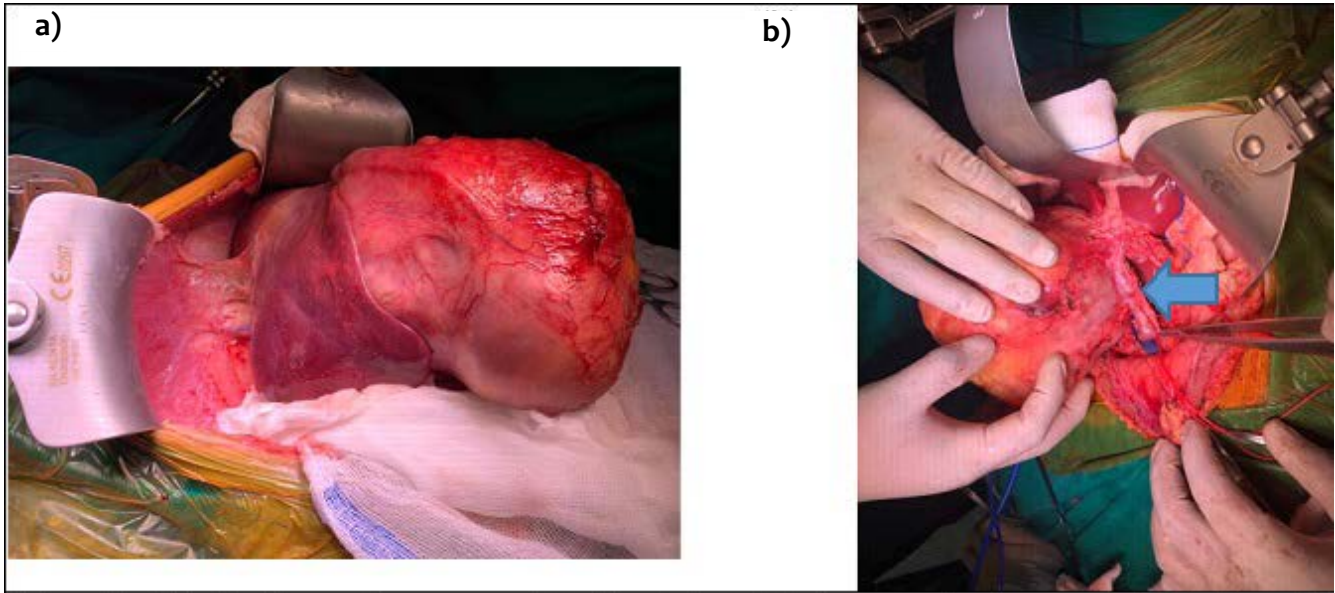
Figure 1. a) Abdominal magnetic resonance: cystic tumor compatible with liver teratoma measuring 11.9 cm x 13.8 cm x 9.3 cm (SD x DL x DBH). It is centered in segment V and partially compromises segment VIII. **b)** Abdominal computerized axial tomography: cystic type tumor with thin walls, heterogeneous with solid and cystic areas.



The case was reviewed and surgical intervention was decided, we found a reddish-brown tumor of +- 13 cm in length that compromised segments V, VI, VII and VIII. The common hepatic duct was 12 cm long and 3 mm in diameter and ran along the anteromedial aspect of the tumor, while the portal vein was 10 cm long and 8 mm in diameter posterior to the tumor, that is why a right hepatectomy was conducted, additionally a biliodigestive derivation was also performed due to elongation and compromise of the common bile duct. (Figure 2 a and b)

The adhesions of the tumor were released towards the duodenum, dissection of the triangular and round ligaments, ligation of the right hepatic artery and right portal vein. Cantlie's line was identified and hepatic transection was performed with monopolar and bipolar. The right suprahepatic vein was sutured with PDS 4/0 continuous stitches. The common bile duct, which ran over the anteromedial aspect of the tumor, was sectioned distally and remodeled for a subsequent end-to-side hepatoduodenal anastomosis with PDS 4/0 separate stitches.

Figure 2: a) Reddish-brown tumor measuring 13 cm in length that compromised segments V, VI, VII, and VIII. b) Elongated portal vein 10 cm long and 8 mm in diameter medial to the tumor.



The macroscopic study of the anatomopathological specimen revealed a dark brown tumor measuring 13 x 12.5 x 10 cm, with

multiple cystic cavities containing friable yellow mucoid and sebaceous material and solid areas of bone and cartilaginous tissue. (Figure 3.)

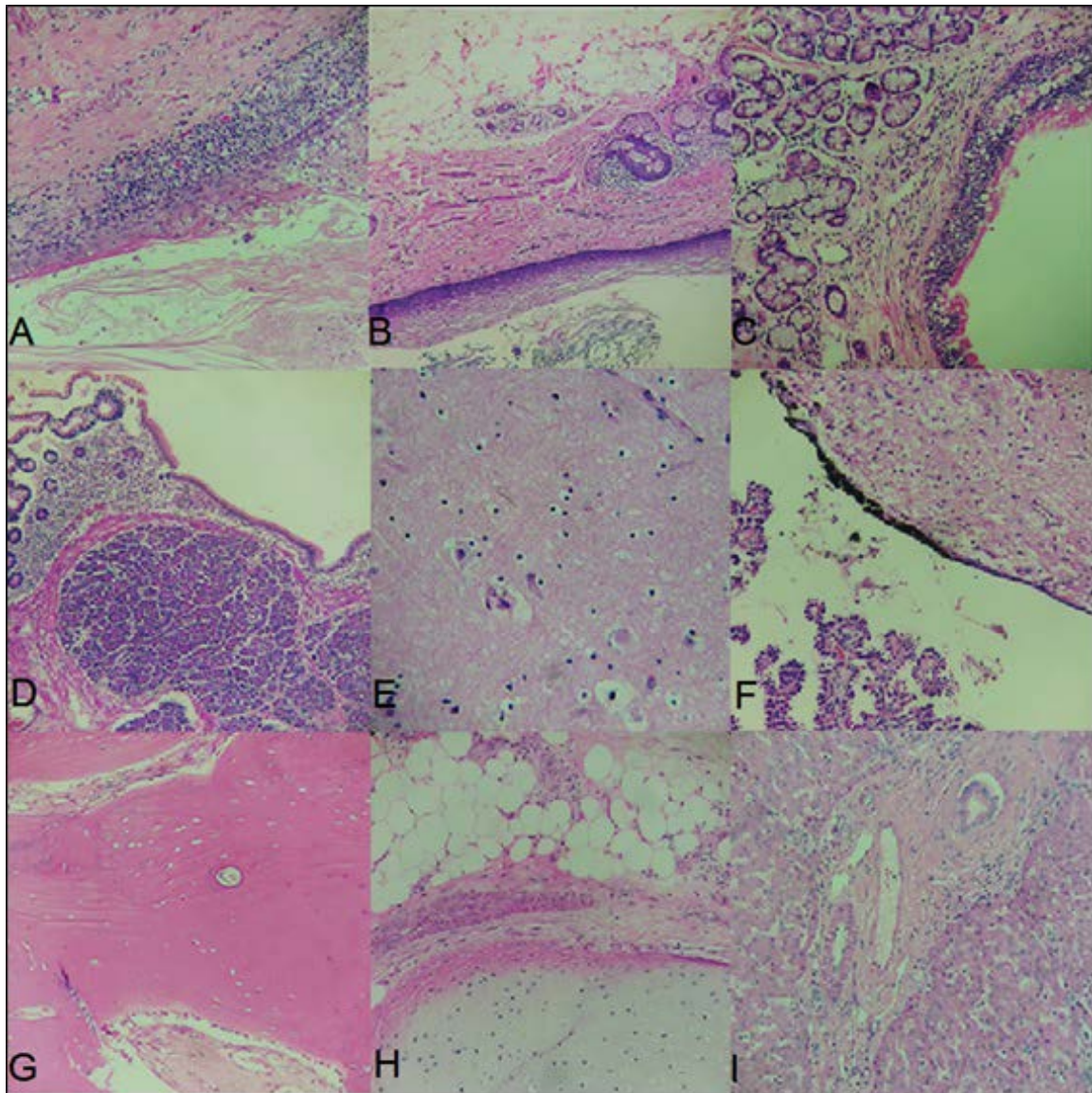
Figure 3: (a) Liver Tumor: Liver tissue (left) and tumor with multiple cystic cavities with mucoid and sebaceous material, yellowish in color and friable, with solid areas of bone and cartilaginous tissue are observed.



Light microscopy examination revealed structures of ectodermal (stratified keratinizing epithelium, mature glial tissue and retina), mesodermal (lymphoid tissue, lymph node, muscle, adipose tissue, cartilaginous tissue, and mature bone) and endodermal (pancreatic tissue, intestinal and respiratory epithelium)

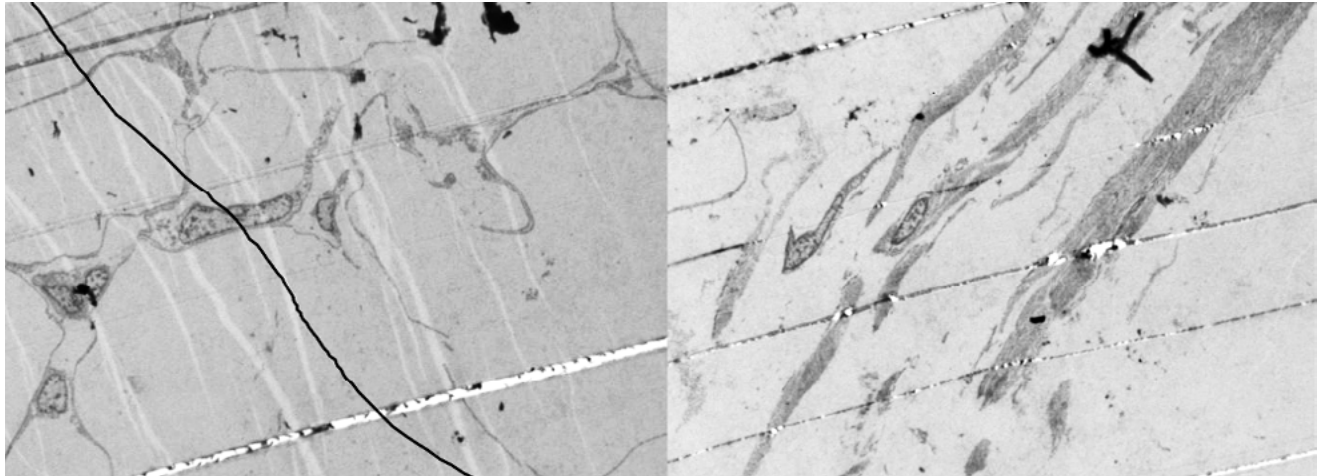
origins. With these findings, the diagnosis of Liver Mature Teratoma was confirmed. Non-tumor liver tissue showed short fibrous expansions of some portal vein spaces and mild inflammatory infiltrate. The Gall bladder did not show significant alterations. (Figure 4.)

Figure 4: Hepatic Teratoma: A,B.- Keratinizing squamous epithelium (H&E, original magnification x200). C,D.- Intestinal and pancreatic tissue (H&E, original magnification x100 E.- Glial tissue (H&E, original magnification x400). F.- Glial tissue, retina and choroid plexus (H&E, original magnification x200). G.- Tissue Bone (H&E, original magnification x200) H.- Adipose and cartilaginous tissue (H&E, original magnification x200).I.- Non-tumor liver parenchyma.(H&E, original magnification x200).



Transmission Electron Microscopy revealed neural and adipose tissue, fibroblasts and collagen fibers. The cells and tissues proved to be mature. (Fig.5.)

Figure 5. Hepatic Teratoma with Transmission Electron Microscopy: adipose tissue (left), fibroblasts and collagen fibers (right). The cells and tissues proved to be mature. (Electromicrophotography, original magnification x4000).



Histochemical staining such as Masson's Fontana, Pas, Reticulin and Masson's Trichrome and CD68, CK7 and S-100 Immunohistochemical staining confirmed the cell lineage. Transmission Electron Microscopy revealed neural and adipose tissue, fibroblasts and collagen fibers. The cells and tissues proved to be mature. (Fig.5.)

In the postoperative period, the patient had a residual abdominal collection despite peritoneal drainage, so percutaneous drainage was performed and antibiotic coverage was improved leading to remission in the following days. Currently the patient is not showing any signs or symptoms of illness/disease and did not require chemotherapy. Until now, the patient has survived for more than 3 years.

Discussion

The teratoma is a germ cell tumor that occurs most frequently in the ovaries and

testicles, followed by the anterior mediastinum, retroperitoneum and sacrocoxygeal region.⁴ The pathological anatomy study confirmed the diagnosis of teratoma and should establish which type it belongs to: mature or immature. Immature teratomas are histologically graded (based on the Norris System) according to the proportion of immature elements (mainly neuroectodermal) found on low-field microscopy. Grade I correspond to immature tissue limited to no more of 1 field at low magnification per slide, Grade II consists of finding immature tissue that does not exceed 3 low-power fields per slide and Grade III is defined as finding immature tissue in more than 3 low-power fields per slide, the latter considered malignant.⁵⁻⁶

Malignant transformation must also be ruled out of some differentiated element of a mature teratoma, called Teratoma with malignant transformation.⁶

The location in the gastrointestinal tract and liver comprises 1% of all extragonadal organs.⁵

Among the differential diagnoses of liver tumors are hepatoblastoma, hepatocarcinoma, myelolipoma, lipoma and angiomyolipoma.²

This patient's case was a primary mature liver teratoma, without gonadal organ compromise, and without elevation of AFP, indicating a favorable prognosis. This findings are similar to those found by O'Really with a 100% survival in mature liver teratoma (LT).⁷ However, mortality might be higher in all patients older than 15 years with immature primary liver teratomas that produce AFP as documented by Cöl.⁸

The symptoms of mature and immature PLT are nonspecific and are conditioned by mechanical pressure from the growing tumor, including abdominal distention and pain, constipation, fever, loss of appetite, a feeling of fullness in the right upper quadrant, vomiting, as well as acute abdominal pain.^{8,9} According to O'Reilly, abdominal pain was the most frequent symptom reported by patients with LT⁷, similar to the experience of this patient, whom presented with distension, and intermittent abdominal pain due to the large size of the hepatic tumor found.

According to García-Ríos, this condition is more frequent in females and the size of the tumor can range from 4 to 18 cm, being located more frequently in the right hepatic lobe.¹³ Unlike Gupta et al.'s case of a 4-year-old patient with a 4cm LT located in the caudate lobe.⁴ However, in this case it was a large tumor in a young boy that caused elongation of the common hepatic duct and portal vein, being the first case know to have this complication to the best of our knowledge.

Initial contrast enhanced CT imaging and MRI scan demonstrated a large mass and allowed an adequate diagnosis and surgical planning. Surgery with complete resection of the tumor is the appropriate treatment, as well as a correct histological evaluation of the

tumor.¹ Laparoscopic surgery has a diagnostic advantage in which peritoneal liquid sampling and exploration of the abdominopelvic cavity for malignancy can be conducted in order to assess the involvement of adjacent structures. However, laparotomy is recommended in complex cases like cyst ruptures with an acute emergency, massive complex masses or advanced stage malignancy detected.¹¹ Malignant LT has a poor prognosis; however, complete resection of a benign LT is curative.¹² In the present case a hepatectomy was needed because of the extension and complex location of the tumor.

A hepatectomy was performed considering the size of the tumor, despite the possible complications like bleeding or liver failure. This patient also needed remodeling of the bile duct and choledochoduodenal derivation due to its great elongation and high risk of kinking without leaks or stenosis. There is currently no published literature regarding this kind of pediatric or adult patients at the national level which justifies its publication.

Conclusions

Primary mature liver teratoma is a rare tumor more frequent in children, so it should be suspected in pediatric patients with solid liver tumors that do not present with high AFP values. Likewise, due to the large tumor size with elongation of the bile duct and main portal vein, a right hepatectomy with biliodigestive derivation was performed. Imaging studies and surgical planning were crucial for a successful surgery, especially for this patient whom is the first national reported case for these findings.

Ethical Considerations

This manuscript complies with all ethical aspects according to COPE guidelines.

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Conflict of Interest

The authors declare that they have no conflicts of interest.

Contributions

Carolina Paz Soldán Mesta: Original idea, data acquisition, analysis and interpretation, manuscript writing and review.

José Luis Apaza León: Original idea and data acquisition.

Mariela Tello: Data acquisition

Roxana Lipa Chancolla: Data analysis and manuscript writing

Carlos Enrique Paz Soldán Oblitas: Data análisis and interpretation, manuscript review.

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