Caroli's disease as an hepatic exophytic mass: a case report

Doença de Caroli sob a forma de massa hepática exofítica: relato de caso

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Abstract

Purpose: to report on a rare case of Caroli's disease presented as an hepatic exophytic mass.
Methods: the authors reviewed the literature about Caroli's disease and its forms of presentation, and report an atypical case of the disease.
Results: Caroli's disease is characterized by a segmental and sacular dilatation of intrahepatic biliary ducts. We report an atypical case of the disease, presented as an hepatic exophytic mass on a 3-year-old female child. We have resected the mass and the patient is asymptomatic after a 33-month follow-up.
Conclusions: Caroli's disease is a differential diagnosis to hepatic exophytic masses.
Key words: Caroli's disease; liver neoplasms; diagnostic; child.

Resumo

Os autores relatam um caso da doença de Caroli apresentado como uma massa exofítica do lobo direito do fígado, revisaram a literatura, as suas formas de apresentação e reportam uma apresentação atípica da doença e o seu tratamento cirúrgico em uma paciente de 3 anos de idade.
A doença de Caroli se carateriza pela dilatação segmentar e sacular dos ductos biliares intra-hepáticos.
***Palavras-chave:** doença de Caroli; neoplasias hepáticas; diagnóstico; criança.*

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Recebido em maio de 2001.

INTRODUCTION

Caroli's Disease is characterized by a segmental and sacular dilatation of intrahepatic biliary ducts. We report on an atypical case of that disease, that presented as an hepatic exophytic mass.

CASE REPORT

We present the case of a 3-year-old female child presenting hepatomegaly for 7 months.

Serum dosage of α-phetoprotein was normal (1,38 ng/dl). Other serum dosages were AST=38 UI/ml, ALT=15 UI/ml, alkaline phosphatase of 552 UI/ml, total bilirubin of 0,36 mg/dl and indirect bilirubin of 0,24 mg/dl. Abdominal ultrasound showed a cystic image on the right lobe of the liver. Abdominal computarized tomography showed an expansive mass on the right hypocondrium, which contained multiple cysts of various sizes and specific density, contiguous to the inferior limb of the liver. Marked hematia scintigraphy of the liver showed hypocaptation of the radioisotope on the lower 2/3 of the liver.

We found an hepatic exophytic tumor, which was cystic at palpation, at surgery. We performed an hepatic segmentectomy (V and VI segments) and cholecystectomy.

Histopathologic examination revealed congenital dilatation of intra-hepatic biliary ducts (Caroli's disease).

Follow-up of 33 months without abnormalities

Caroli's disease is a congenital disease that may be asymptomatic during infancy, or presented with hepatomegaly and jaundice.[1] It may be associated with other hepatobiliary diseases such as biliary stones (up to 95% of the cases[2]), congenital hepatic fibrosis, cystic degeneration of extra-hepatic biliary ducts, and extra-hepatic biliary ducts atresia.[3-5]

It may coexist with renal malformation, which is presented on up to 80% of the cases on Brazil.[1]

Reviewing the papers on the subject, we were able to find only another case of Caroli's Disease presented as an exophytic mass of the right lobe.[6]

Caroli´s disease is often diagnosed at an adult age.[2,3] In Brazil, however, the median

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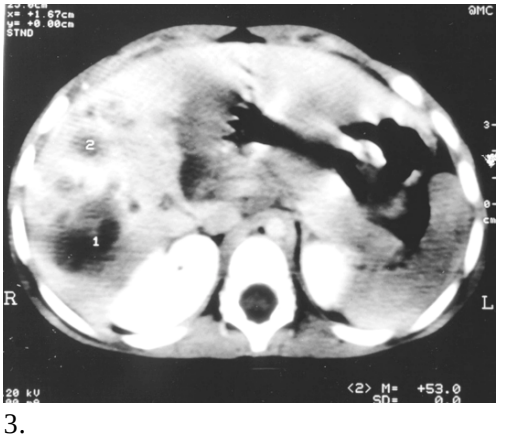

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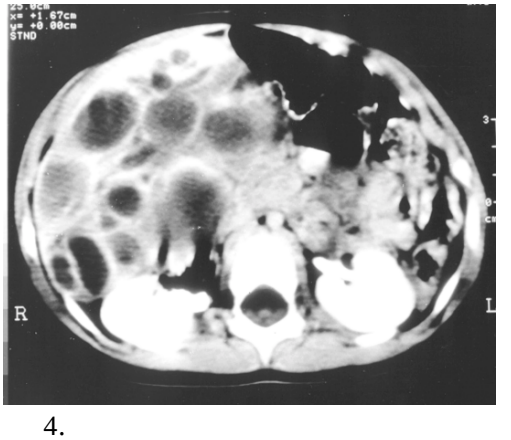

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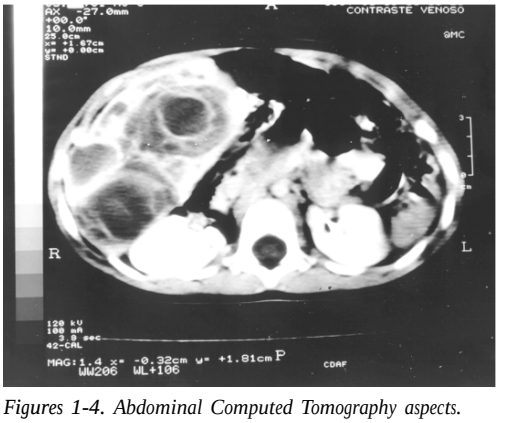

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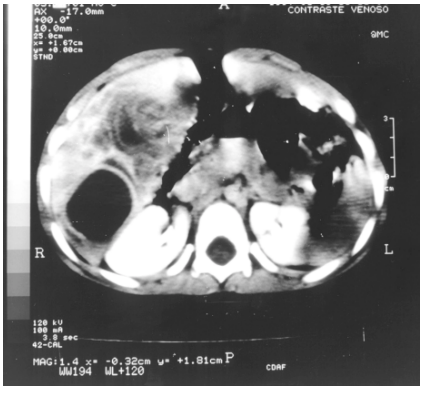

Figures 1-4. Abdominal Computed Tomography aspects.

age for symptoms to appear is 5,5 months old, and diagnosis is made at 12 months old.[1]

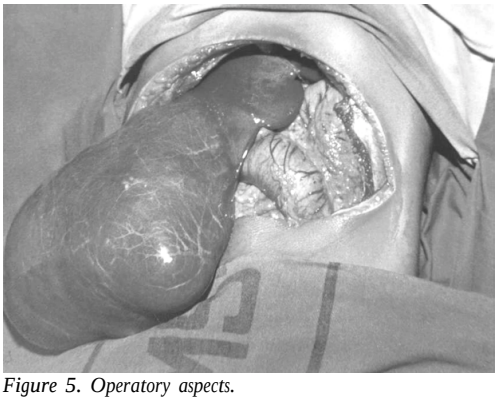

Figure 5. Operatory aspects.

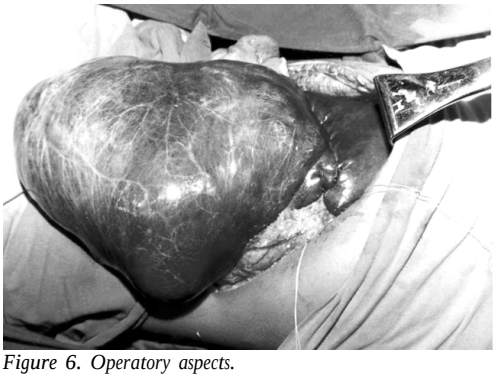

Figure 6. Operatory aspects.

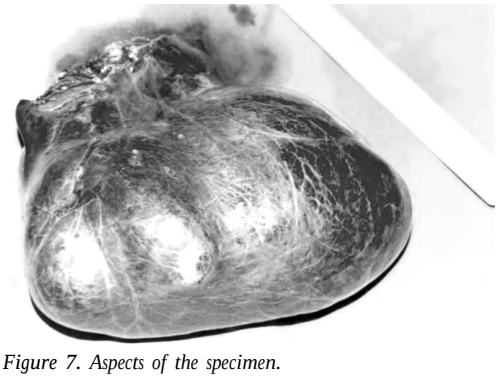

Figure 7. Aspects of the specimen.

There are no relevant differences about sex distribution

Diagnosis is made through abdominal CT scan and US. On CT scan the disease is defined by dilated intra-hepatic biliary ducts, portal bridges, intraluminal protrusions and indirect signs of cholangitis.[7] Other tests may be valuable to establish diagnosis of the disease: dietil-iminodiacetic acid scintigraphy, helicoidal cholangiotomography[8] and magnetic cholangioressonancy.[9]

The primary treatment is surgery, for biliary drainage and symptoms relief.[2] Exophytic masses must be totally resected,[6] as we did.

We reported the case because the disease is rare and the form of presentation even more rare in children.

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