

## Clinical Case

# Extrahepatic Biliary Myeloid Sarcoma: Surprise Diagnosis on Suspicious Obstructive Jaundice

## Sarcoma Mieloide Extra-Hepático das Vias Biliares: Um Diagnóstico Inesperado na Suspeita de Icterícia Obstrutiva

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### ABSTRACT

A man presents with appetite loss and jaundice with no further complaints. Computed tomography (CT) finds a stenosis of the common bile duct (CBD) compatible with a type II Bismuth tumor. After discussion in a multidisciplinary team meeting, the patient was referred to a Reference Center of Oncological Hepatobiliary Surgery, and left hepatectomy with biliary reconstruction was proposed.

Before the patient was submitted to surgery, a blood smear showed blastic cells and the patient was diagnosed with acute myeloid leukemia (AML). Intraoperatively, extemporaneous analysis of the biliary margin after left hepatectomy showed it was invaded by leukemic cells.

Final anatomopathological analysis revealed a myeloid sarcoma with medullary extent with *NPM1* mutation and the patient was referred to Hematology for further treatment.

**Keywords:** Bile Duct Neoplasms; Bile Ducts, Extrahepatic; Cholangiopancreatography, Endoscopic Retrograde; Cholestasis, Extrahepatic; Jaundice, Obstructive; Sarcoma, Myeloid

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## RESUMO

Um doente, sexo masculino, apresentou perda de apetite e icterícia, sem outras queixas associadas. Realizou tomografia computadorizada (TC) que revelou uma estenose do canal biliar comum (CBC), compatível com um tumor de Bismuth tipo II. Após discussão em reunião multidisciplinar, o doente foi referenciado para um Centro de Referência de Cirurgia Hepatobiliopancreática Oncológica, tendo sido proposta uma hepatectomia esquerda com reconstrução da via biliar. Antes da realização da cirurgia, o esfregaço de sangue periférico revelou a presença de células blásticas, tendo sido estabelecido o diagnóstico de leucemia mieloide aguda (LMA). Intraoperatoriamente, a análise extemporânea da margem biliar após a hepatectomia esquerda revelou invasão por células leucêmicas. O exame anatomopatológico definitivo revelou um sarcoma mieloide com extensão medular, associado a uma mutação no gene *NPM1*. O doente foi posteriormente referenciado para a Unidade de Hematologia para prosseguimento do tratamento.

**Palavras-chave:** Colangiopancreatografia Retrógrada Endoscópica; Colestase Extra-Hepática; Ductos Biliares Extra-Hepáticos; Icterícia Obstrutiva; Neoplasias dos Ductos Biliares; Sarcoma Mieloide

## INTRODUCTION

This report describes a case of obstructive jaundice with the initial suspicion of cholangiocarcinoma, with acute presentation. During staging, the patient was diagnosed with acute myeloid leukemia (AML) and a left hepatectomy with biliary reconstruction was performed. Final anatomopathological analysis revealed a myeloid sarcoma. This case demonstrates an extremely rare presentation form of AML.

## CASE REPORT

A man presents with appetite loss and jaundice for the previous three days (total bilirubin [TBrb] of 8.6 mg/dL and direct Bilirubin [DBrb] of 6.0 mg/dL), with no further complaints. This patient had no relevant pathological history, only previous inguinal hernia surgical repair.

Besides apparent mucocutaneous jaundice, there were no other relevant findings in the clinical evaluation.

Abdominal computed tomography (CT) (Fig. 1) and magnetic resonance imaging (MRI) (Figs. 2 and 3) show stenosis of CBD and hilum convergence, compatible with a type II Bismuth tumor. Thorax CT showed no signs of pulmonary metastasis. After discussion in a multidisciplinary team meeting, the patient was referred to a Reference Center of Oncological Hepatobiliary Surgery and left hepatectomy with biliary reconstruction was proposed.

Before the patient was submitted to surgery, about 1 month prior, he was submitted to biliary drainage since his TBrb levels were aggravated (11.4 mg/dL) and blood smear showed 18% of blastic cells. With these findings, Hematology diagnosed acute myeloid leukemia (AML).

After stabilization of bilirubin levels, the patient was submitted to left hepatectomy enlarged to segment I and biliary Y-Roux reconstruction with a jejunal loop. Intraoperatively there was no apparent metastasis, and extemporaneous analysis of the biliary margin after left hepatectomy revealed invasion with leukemic cells. Therefore enlargement of margins was necessary.

The patient recovered, although he has a post op cholangitis, with identification of *Candida albicans*, *Enterococcus faecalis* and *Serratia marcescens* (isolated from intraoperative biliary drainage), with treatment including concomitant



**Figure 1:** Abdominal CT compatible with stenosis of CBD and hilum convergence.

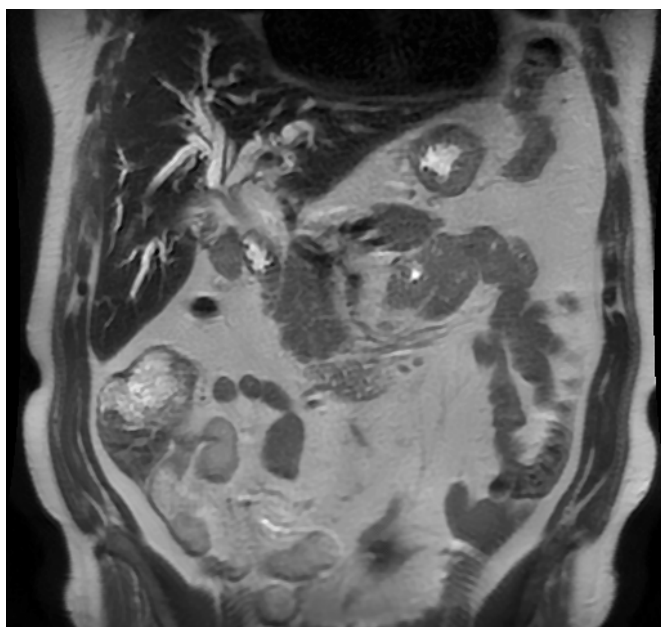


**Figure 2:** Abdominal MRI compatible with stenosis of CBD and hilum convergence.

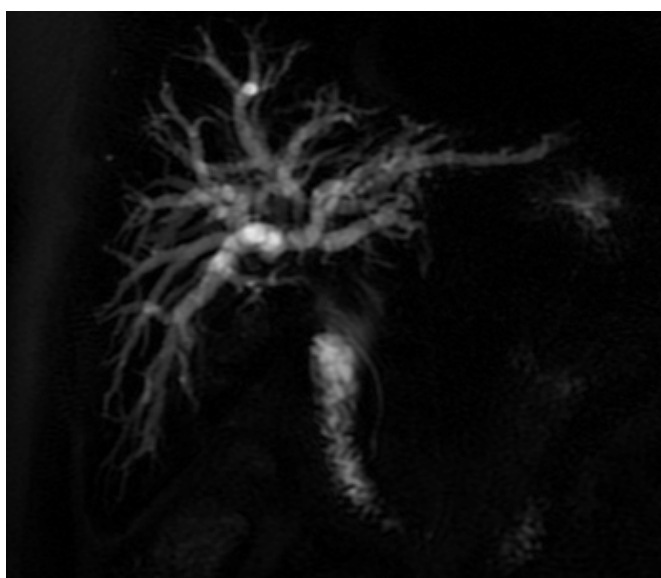


**Figure 3:** Abdominal MRI compatible with stenosis of CBD and hilum convergence.

antibiotherapy with fluconazole, trimethoprim/sulfamethoxazole and ampicillin.

Final anatomopathological analysis revealed a myeloid sarcoma with medullary extent with *NPM1* mutation and the patient was referred to Hematology for further treatment. Presently, he is 1 year post op, clinically well, still under consolidation therapy with idarubicin and cytarabine (IDAC) and waiting for allotransplant.

## DISCUSSION

### 1. DEFINITION AND EPIDEMIOLOGY

Myeloid sarcoma (MS) is a rare high-grade solid tumor consisting of myeloid blasts, at an extramedullary anatomical site. Classically, MS is described as a rare presentation of acute myeloid leukemia (AML). The mechanisms through which the leukemic cells migrate to form solid extramedullary tumors are still unclear. However, some authors suggest some mechanisms responsible for this migration, such as impaired adhesion to the Bone Marrow niche (due to altered expression of adhesion molecules, for example). According to the literature, MS typically presents between 40 and 70 years of age, mainly in male patients.<sup>1</sup> The incidence of MS with bone marrow involvement at AML diagnosis ranges from 2.8% to 0.8% for isolated MS.<sup>2</sup> The risk factors predisposing to the development of MS are similar to the factors predisposing AML, including environmental, lifestyle and genetic factors.

### 2. CLINICAL PRESENTATION AND DIAGNOSIS

MS frequently presents as tumors on the skin, lymph nodes, gastrointestinal tract and central nervous system. A hepatic myeloid sarcoma is one of the rarest forms, occurring in about 4% of MS cases. Due to this fact, MS usually mimics other primary or metastatic liver malignancies.

Extrahepatic biliary MS typically presents with obstructive jaundice and right upper quadrant abdominal pain. These symptoms, accompanied by fever, might resemble cholangitis and may precede or follow systemic AML.

Imaging through ultrasound or even CT scans may be used to detect the cause of the biliary obstruction and diagnose a focal mass, but cannot reliably distinguish MS from other causes, such as cholangiocarcinoma or inflammatory disease.<sup>3</sup>

Therefore, tissue biopsy is the only diagnostic instrument that can reliably establish a correct diagnosis. The biopsy might be obtained through endoscopic (ERCP) or percutaneous (TPC) whenever a biliary stricture is suspected to be malignant with atypical characteristics or is accompanied by suspected systemic hematologic disease. The most common histologic features are as follows: dense infiltrates of myeloblasts or myeloid precursors with medium-sized round nuclei, with diffuse infiltration of the bile duct.<sup>4</sup>

Immunohistochemistry is essential to confirm myeloid lineage and distinguish MS from carcinoma or lymphoma. The most common positive markers reported are myeloperoxidase, CD68, lysozyme, CD45, CD117 (c-kit), CD43 and CD33.<sup>4</sup>

### 3. TREATMENT

#### **Systemic chemotherapy**

Systemic chemotherapy is the cornerstone for the treatment of extrahepatic biliary MS and several published cases show its benefit in resolving biliary obstruction. These reports describe cytarabine-based induction (with anthracyclines or etoposide) producing mass regression and normalization of liver tests, with durable remission in both pediatric and adult patients.<sup>4</sup>

#### **Radiotherapy**

Radiotherapy is usually applied to patients for local control or palliation of symptomatic lesions.<sup>3</sup>

#### **Surgery and biliary drainage**

Surgery is frequently performed ad initium due to misdiagnosis as cholangiocarcinoma, but resection alone

does not reliably prevent systemic progression. These patients should be treated with adjuvant AML-directed chemotherapy (discussed above) to prevent progression.<sup>5</sup> Endoscopic or surgical biliary drainage can be useful to relieve obstructive jaundice and enable safe delivery of systemic therapy.

### LEARNING POINTS

Extrahepatic biliary MS is, with no doubt, a difficult diagnosis to make, especially due to the misdiagnosis related with biliary obstruction. Because of its rarity, there are no established guidelines for its treatment, but for now the literature bases its treatment on similar protocols as for AML. Surgical resection might be applied, however should not be the first choice nor an isolated one.

### ETHICAL DISCLOSURES

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### CONTRIBUTORSHIP STATEMENT

**IP:** Conceptualization, methodology, formal analysis, writing.

**AC:** Conceptualization, methodology.

**AR, RM, and JGT:** Formal analysis, writing.

All authors approved the final version of the manuscript for publication and assume responsibility for all aspects of the work, ensuring the accuracy and integrity of the data presented.

### DECLARAÇÃO DE CONTRIBUIÇÃO

**IP:** Conceptualização, metodologia, análise formal, redação.

**AC:** Conceptualização, metodologia.

**AR, RM e JGT:** Análise formal, redação.

Todos os autores aprovaram a versão final do manuscrito para publicação e assumem responsabilidade por todos os aspetos do trabalho, garantindo a exatidão e a integridade dos dados apresentados.

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