

## About A Rare Case of Primary Gallbladder Sarcoma

Gouton Ulrich Martial F.<sup>1\*</sup>, Ali Said Samah<sup>1</sup>, Adjadé Ganiou<sup>2</sup>, El Fadli Mohammed<sup>2</sup>, Adil Ait Errami<sup>1</sup>, Sofia Oubaha<sup>1</sup>, Zouhour Samlani<sup>1</sup>, Belbaraka Rhizlane<sup>2</sup>, Khadija Krati<sup>1</sup>

<sup>1</sup>Department of Hepato-Gastroenterology, Ar-razi Hospital, CHU Mohammed VI, University Cady Ayyad, Marrakech, Morocco

<sup>2</sup>Department of Medical Oncology, CHU Mohammed VI, University Cady Ayyad, Marrakech, Morocco

**\*Corresponding Author**  
Gouton Ulrich Martial F

### Article History

Received: 26.11.2020

Accepted: 16.02.2021

Published: 25.02.2021

**Abstract:** Primary gallbladder sarcomas are histological types rarely found in clinical practice. A few dozen cases have been identified in the literature since the first description made by Griffon Segall in 1897. In our 68-year-old patient, the diagnosis of the gallbladder sarcoma was possible by immunohistochemistry. It was a stage IV-B of TNM at imaging examinations (MRI and CT), which did not allow a curative approach. His evolution was quickly unfavorable with his passing within 6 weeks.

**Keywords:** Sarcoma, Gallbladder, Metastasis, Biliary Prosthesis, Marrakech, Morocco.

**Copyright © 2021 The Author(s):** This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

## INTRODUCTION

Cancer of the gallbladder is common and it's the most aggressive tumor damage of the bile tract [1]. The sarcomatoid histological type is a rare entity [2] and only a few dozen cases have been reported in the literature. It has only been described in subjects over the age of 60 [2], most often asymptomatic. When symptomatic, the typical clinical symptoms may include: pain in the right hypochondrium associated with jaundice and deterioration of the general condition [3]. Despite the improvement in diagnostic means, the majority of patients consult at a very advanced stage, making the therapeutic results unfavorable. We reported a rare case of locally advanced gallbladder sarcoma with liver metastasis in a 68-year-old woman.

## CLINICAL CASE

This is a 68-year-old woman, with no particular pathological history, admitted with a cholestatic jaundice preceded 2 months earlier by pain in the right hypochondrium with deterioration of the general condition. The physical examination objected a WHO performance index of 3, cutaneous-mucous jaundice, diffuse abdominal tenderness more marked in the right hypochondrium with painful irregular surface and hard consistency hepatomegaly.

An abdominal CT scan demonstrated suspicious tissue thickening of the gallbladder. The bili - MRI demonstrated a 40 mm tumor wall thickening of the vesicular wall, infiltrating the hepatic parenchyma, the pancreas, stomach and the right colic angle with significant dilation of the upstream intrahepatic bile tract. The liver presented nodular lesions at segments VI, V, and IV, with the largest one located at the level of segment V measuring 3.64 cm long axis. There was also a magma of hepatic, coeliomenteric, and lumbo-aortic hilar ADP encompassing the celiac trunk, its dividing branches, and micronodular infiltration of peritoneal fat (figures 1, 2).



**ig-1 : Heterogeneous Liver With Irregular Site Of Sparse Hypodense Lesions Consistent With Secondary Lesions**

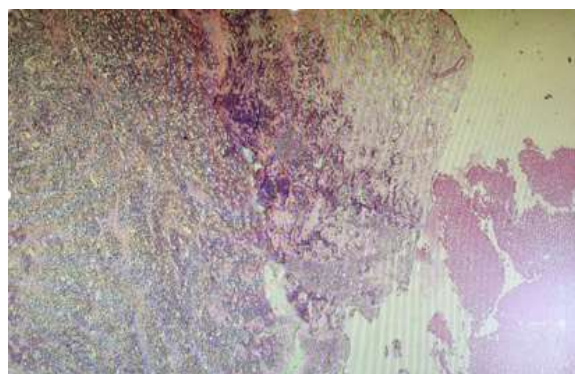


**Fig-2: Circumferential And Regular Wall Thickening With Partially Stenosing Right Colic Angle**

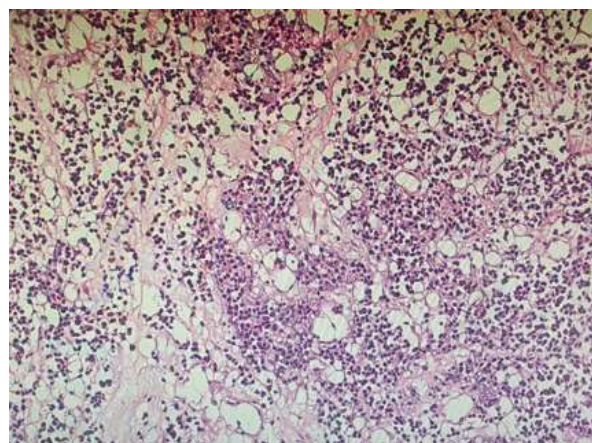
Biological assessment noted cholestasis with high alkaline phosphatase and gamma-glutamyl transpeptidase (ALP = 3046 IU / L, GGT = 1094 IU / L), predominantly conjugated hyperbilirubinemia at 368 mg / L, low prothrombin level at 36% (positive Koller test), associated with liver cytolysis with elevated liver enzymes (ALT = 168 IU / L and AST = 471 IU / L). The CA 19-9 tumor marker was elevated to 4907 IU / mL. Surgical exploration by right subcostal incision showed tumor proliferation of the gallbladder taking the main bile tract, invading the right colon, and the hepatic pedicle.

The histological study of the biopsies carried out a poorly differentiated tumor proliferation with round cells, organized in a rounded area without spans or acini, with a large nucleus without nucleolus, and scant eosinophilic cytoplasm.

The immunohistochemical study showed tumor cells expressing anti-CD4 and anti-vimentin in the vessels. The tumor did not express smooth muscle actin (SMA), anti-desmin, H-caldesmon, anti-myogenin, anti-hepatocyte, anti-pan cytokeratin, anti-PS100, anti CD30, anti PLAP, and anti LCA. (figures 3, 4 & 5). All these arguments concluded to a poorly differentiated sarcoma of the gallbladder stage IV-B.



**Fig-3: Hepatic parenchyma destroyed by layers of tumor cells (HEX10)**



**Fig-4: Medium-Sized round tumor cells sites of moderate cytonuclear atypia (HEX20).**



**Fig-5: Absence of expression of anti-vimentin antibody by tumor cells with positivity in blood vessels**

Given the patient's non-operability and non-resectability of the tumor, it was decided, through a multidisciplinary consultation meeting, to refer the patient to a palliative care with biliary drainage by fitting a metal endoscopic prosthesis. She passed within after 6 weeks of supportive care.

## DISCUSSION

Cancer of the gallbladder is the most common malignant neoplasm of the bile tract [1]. Adenocarcinoma is the most common histologic type at 98% [1]. Papillary, mucinous, and squamous cell carcinomas are other common histological types. Sarcomatoid carcinomas, small cell carcinomas and lymphomas are rare entities [1]. Sarcomas are extremely rare, for around 1.5% of all malignant tumors of the gallbladder [4]. It is an entity that is mostly diagnosed in women over the age of 60 [2].

It should be noted that it's asymptomatic in the majority of cases. The tumor was inadvertently discovered during the surgery of a benign pathology. Symptomatic patients generally have already locally advanced or unresectable metastatic tumor, as was the case of our patient. The clinical presentation is non specific, but most often includes epigastric or right hypochondrium pain, jaundice, digestive disorders and deterioration of general condition

[1].The preoperative diagnosis of gallbladder sarcoma is therefore difficult and tumor markers such as  $\alpha$ FP, CEA, CA19-9 are not specific [5].

Ultrasound is the first imaging test requested if hepatobiliary disease is suspected. The CT scan and MRI allow a better study of the gallbladder. They show hepatic and biliary infiltration, lymph node, vascular and metastatic extension [1]. But the diagnosis of sarcoma of the gallbladder is confirmed on histological examination. In the immunohistochemical study, mesenchymal markers such as desmin, actin and vimentin should be positive [5]. Monobloc R0 surgical resection is the standard of the therapy of localized tumors [4]. There is insufficient evidence in the literature regarding the efficacy of chemotherapy or adjuvant radiotherapy [2]. However, they are still indicated in high-grade aggressive sarcomas or in the event of R1 resection [4]. For tumors that are unresectable from the outset or have metastasis, radiotherapy and / or chemotherapy should be considered. The chemotherapy molecules that have been shown to be effective in this indication are dacarbazine, doxorubicin, epirubicin, gemcitabine, ifosfamide, liposomal doxorubicin and temozolomide. They are used as monotherapy or in combination as in the AD (doxorubicin, dacarbazine), AIM (doxorubicin, ifosfamide, mesna), or MAID protocols [4]. Our patient being inoperable with a locally advanced and metastatic tumor, supportive care remains the only therapeutic indicated. In addition, the prognosis of

sarcomas is very unfavorable with a 5-year survival rate estimated at 5%.

## CONCLUSION

Surgical resection is a standard in the localized sarcomas of the gallbladder. The prognosis of locally advanced and metastatic forms is unfavorable. Progress remains to be made for early detection in order to improve the prognosis of patients.

## REFERENCES

1. Kanthan, R., Senger, J. L., Ahmed, S., & Kanthan, S. C. (2015). Gallbladder cancer in the 21st century. *Journal of oncology*, 2015.
2. Paasch, C., Salak, M., Mairinger, T., & Theissig, F. (2020). Leiomyosarcoma of the gallbladder—A case report and a review of literature. *International journal of surgery case reports*, 66, 182-186.
3. Da Costa, A. C., Santa-Cruz, F., Sena, B. F., Lopes, A., Leite, N., Da Paz, A. R., ... & Figueiredo, J. L. (2018). Dedifferentiated liposarcoma of the gallbladder: first reported case. *World journal of surgical oncology*, 16(1), 1-5.
4. Park, E. Y., Seo, H. I., Yun, S. P., Kim, S., Kim, J. Y., & Han, K. T. (2012). Primary leiomyosarcoma of gallbladder. *Journal of the Korean Surgical Society*, 83(6), 403.
5. Park, S. B., Kim, Y. H., Rho, H. L., Chae, G. B., & Hong, S. K. (2012). Primary carcinosarcoma of the gallbladder. *Journal of the Korean Surgical Society*, 82(1), 54.