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Surgical management of gallbladder carcinosarcoma: A case report and review of the Literature

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ABSTRACT

INTRODUCTION: Gallbladder Carcinosarcoma is a rare and fatal cancer characterized by the presence of a combination of a cancerous epithelial and mesenchymal components.
PRESENTATION OF CASE: We present the case of a 66 years old patient with a carcinoma gallbladder carcinoma who received a surgical treatment. The histological assessment revealed two histological components consisting of an adenocarcinoma and a spindle cell sarcoma compatible with the carcinosarcoma.
DISCUSSION: This cancer is known to be extremely severe in that less than 100 cases have been documented in international medical review of literature. surgical treatment remains the only cure for gallbladder carcinosarcoma, The prognosis of this disease is extremely poor because it normally presents at advanced stages.
CONCLUSION: We present an extremely rare example of carcinosarcoma, and its attributes might be suggestive of a greater malignant potential. Consequently, the later the diagnosed worse is the prognosis.

1. Introduction

Gallbladder cancer, though generally considered rare, is the most common malignancy of the biliary tract. This cancer can be cured with radical surgery, and many efforts have been made in the attempt to improve resectability and the survival rate. Its carcinogenesis follows a progression through a metaplasia–dysplasia–carcinoma sequence. It is usually detected at an advanced stage mainly due to the ambiguity of its clinical presentation, its aggressive biology and its anatomic proximity to the liver [1]. In this article, we report the case of 61 years old male patient, who was brought to our hospital for a suspicious vesicular mass whose surgical exploration revealed a large locally advanced vesicular mass. The tumor was removed without any complications and Histological assessment revealed histological components consisting of an adenocarcinoma with a spindle cell sarcoma compatible with the carcinoma. This article presents a rare case and focuses on and explores the risks, management, and outcomes for primary gallbladder carcinoma. this work has been reported in line with the SCARE criteria [2].

2. Observation

Here we present the case of 66 years old male patient, from Moroccan northeast, with a history of bulbar ulcer perforation operated in 1985. He presented to our department with a history of six months history of upper abdominal pain spreading to the scapula and the right shoulder associated with vomiting. The patient had no history of weight loss, fever, malaise, bone pain or respiratory issues, nor any neurological implications suggestive of metastatic or paraneoplastic manifestations.

Physical examination shows an anticetric patient with the presence of a palpable and painful mass in the right hypochondrium.

Laboratory parameters were normal including hematological, biochemical, liver tests and tumor markers.

The abdomino-pelvic CT scan showed a distortion of the gallbladder fundus and an intraluminal tumor measuring 17 × 11 × 7 cm with no extension to lymph nodes or or distant metastases (Fig. 1).

Diagnosis and management were discussed with the patient. Surgical assessment demonstrated the presence of a huge gallbladder process invading the anterior abdominal side and the first duodenal portion with the presence of multiple nodes without any involvement of the liver or peritoneal lesions. The surgical intervention including an extended gallbladder cholecystectomy in Glenn's manner and duodenum, with gastroentero-anastomosis with lymphadenectomy.

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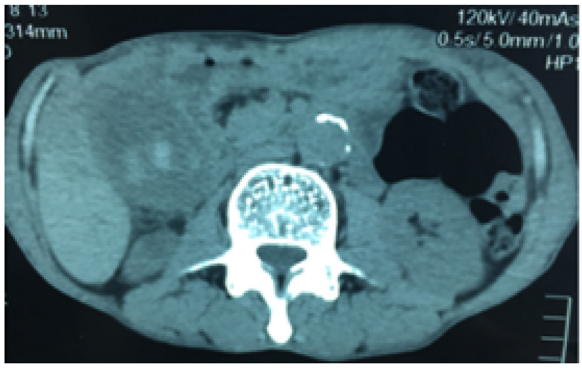

Fig. 1. Transverse iodinated contrast-enhanced MDCT phases showed a heterogeneous, expansile low soft tissue density mass abutting the inferior surface of the gallbladder.

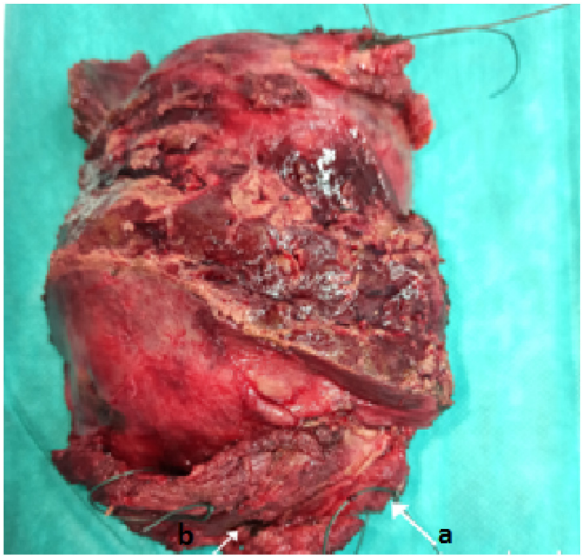

Fig. 2. Photograph of gallbladder. a: the duodenal section slice b: the parietal section slice.

The macroscopic examination of the specimen showed a 15 × 8 × 6 cm tumor which originated from the body of the gallbladder. It was a brown solid mass, with hemorrhagic and necrotic foci (Fig. 2).

Histopathological examination showed a neopalstic proliferation made of a mixture of carcinomatous component, made of glands of markedly atypical cells and a sarcomatous components made of a largely necrotic spindle cell pleomorphic proliferation, showing focal muscular and vascular differentiations.

The immunohistochemical study reported positive staining of tumor cells by CD31, CD34 in areas of vascular differentiation, desmin in areas of muscular differentiation. A focal positivity for Ck7 was also observed (Figs. 3–6).

Consequently, this represented a gallbladder carcinosarcoma stage IVA (pT4, PN0, M0), the case was staffed in multidisciplinary consertation, since there is no standard consensus in the adjuvant management of this type of tumor, we opted for clinical, biological and radiological monitoring by 6 months after a decline of 12 months, no recurrence was detected.

3. Discussion

Gallbladder cancer is the most common and aggressive biliary tract malignancy with the shortest median survival time. The first case of a carcinosarcoma of the gallbladder (CSGB) was reported by Landsteiner in 1907 [1]. Up to now less than 100 Cases have been reported in the literature. Carcinosarcomas of the gallbladder con-

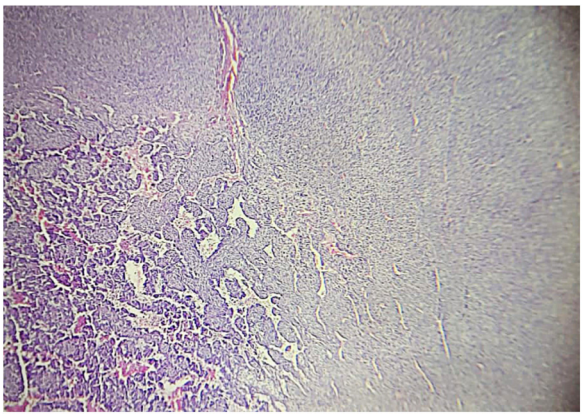

Fig. 3. Microphotography showing a neoplastic prolifération made of a glandular and a sarcomatous component. HE, 100×.

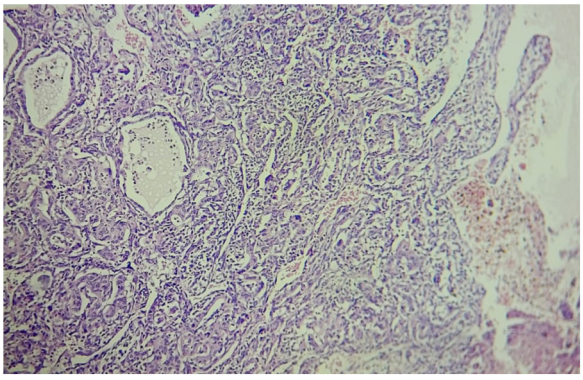

Fig. 4. Microphotography showing a vascular focal differentiation. HE, 100×.

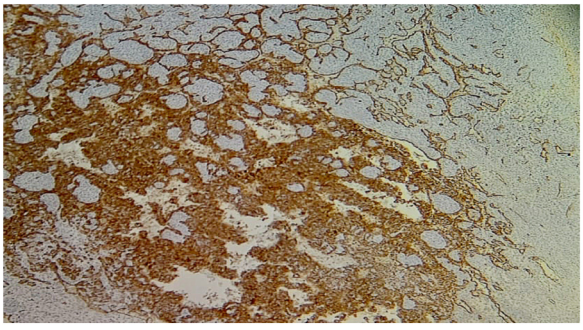

Fig. 5. Microphotography showing posiitve staining by CD34 of cells undergoing a vascular differentiation.

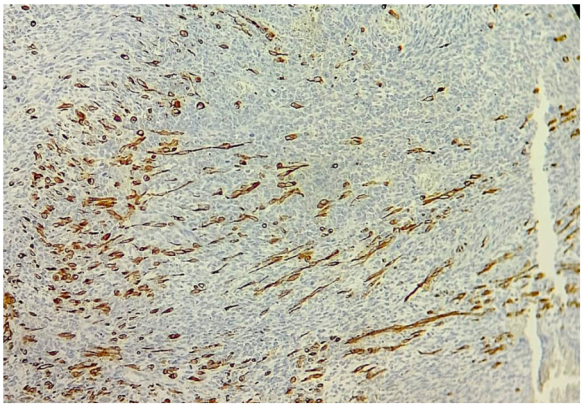

Fig. 6. The sarcomatous component also showed a focal muscular differentiation as shown with desmin stain.

sist of both epithelial and mesenchymal components. The diagnosis requires the presence the two histological components. The epithelial component usually consists of adenocarcinoma, although an element of squamous cell carcinoma is also occasionally observed. Carcinosarcomas are rare and represent less than 1% of gallbladder cancers [3].

Gallbladder Carcinoma is a mixed combination of epithelial and mesenchymal components. According to Zhang's et al. meta-analysis of 68 cases, the age of the patients ranged from 45 to 91 years, with a mean of 68. As for female prevalence, it was presented with a sex ratio of 2,7 [1].

The symptoms and signs of gallbladder carcinosarcoma are non-specific. Representative clinical symptoms include abdominal pain, vomiting, anorexia, weight loss and jaundice. There are four main clinical scenarios a physician may come across when diagnosing a patient with gallbladder cancer. A malignancy may be suspected preoperatively due to clinical presentation. The cancer may be incidentally found on radiologic imaging. Suspicion may arise intraoperatively in a patient undergoing cholecystectomy for presumed benign gallbladder disease, lastly and most commonly, that of incidentally diagnosed malignancy on pathologic examination following a simple cholecystectomy [4].

As far as the biological results are concerned, including inflammatory sign, hepatic result as well as cancerous signs, they were not specific and most often normal. Also,the pre-surgical diagnosis of CSVB is very difficult. With regard to radiology, the radiological literature describing CSVB is rare and there are no specific radiological signs for CSVB.

According to a Portuguese study conducted by JOÃO CRUZ et al., CSVB can appear in an infiltrating or in fibrous forms. Infiltration may happen in a focal asymmetrical way or in a diffusing manner ranging from 20% to 30%, most often invading the subserous level in the liver. On the other hand, fibrous tumours generally present a polypoid 'cauliflower-like' appearance which tends to fill or replace the gallbladder in 40–65% of cases [5].

It appears as a polypoid intraluminal lesion with or without a dense implantation base for 15–25% cases. In MRI, CSGB seems to be hypoid on T1-weighted images and heterogeneous hyperintense moderate on T2-weighted ones. The signal that is moderately high on T2-weighted images, with a heterogeneous partitioned appearance, may be the strongest determinant because of its resemblance to the appearance of visceral sarcomas [5].

The final diagnosis is anatomopathological of the specimen, Pathologic tissue diagnosis is unnecessary in patients where the tumor is considered resectable. In unresectable patients planned for neoadjuvant or definitive chemotherapy, percutaneous biopsy is a reliable method of diagnosis with a sensitivity of approximately 88% [6]. The most common epithelial constituent is adenocarcinoma that is less frequent than squamous carcinoma. As for the mesenchymal component, it is mainly represented by spindle cells, and less frequently by heterogeneous elements that are chondroid, rhabdomyoid, osteoid or mixed cells. On the one hand, for immunohistochemistry, the epithelial component is positive for cytokeratin. On the other hand, the mesenchymal component is positive for vimentin [7].

With respect to therapeutic measures, based on the literature review, only 54 cases have undergone surgical resection as a treatment. There are mainly two researches conducted on this regards; one done by Okabayashi et al. who analyzed the medical records of 36 patients who underwent surgical resection of the CSGB between 1971 and 2009, and the other one belongs to Kunio Mochizuki et al's study carried out between 2009 and 2019. According to them, surgical treatment remains the only cure for gallbladder carcinosarcoma. A simple or an extensive cholecystectomy; including an extended one to the adjacent liver site, and to the main biliary tract with partial resection of the small intestine and/or colon, as well as with duodeno-pancreatectomy, were carried in (36.0%) and (64.0%) of cases respectively, as most of these patients had a locally extensive mass involving adjacent organs [3,4].

Systemic therapy has been used in curative and palliative settings in the management of carcinosarcoma of the gallbladder as an adjuvant treatment alone or in combination with radiotherapy after surgical resection, in a disease not locally advanced unresectable metastatic alone or in combination with radiotherapy, and in advanced metastatic disease.

There are a few reported cases using a single chemotherapy regimen as adjuvant therapy. However, these adjuvant chemotherapies have not seen any added benefit in remission or survival. the diagrams used were essentially based on: UFT: tegafur / uracil, gemcitabine, S-1: tegafur / gimeracil / oteracil [8]

5-Fu is a main chemotherapy regimen in the treatment of gallbladder carcinoma and oxaliplatin is used in the treatment of sarcomas such as soft tissue sarcoma and gynecologic sarcoma. The combination of 5-Fu and oxaliplatin has shown a significant survival benefit in the treatment of aggressive colon cancers [9]. The use of these two chemotherapy regimens as adjuvant chemotherapy has been reported in a recent case for the treatment of carcinosarcoma of the gallbladder [10] otherwise no case of neoadjuvant treatment has been reported. elsewhere Currently, outside of a clinical trial, neoadjuvant therapy is not recommended for surgically resectable gallbladder cancer [11].

In spite of the complementary or the palliative Chemotherapy and/or radiotherapy's roles, they remain unclear because of the limited number of cases described in the literature, as chemotherapy has shown no a significant benefit in terms of morbidity or mortality [12].

However, among resected cases, 31% represents the overall 5-year survival rate while 8.3% stands for the percentage of people who died in hospitals. As it is apparent, it is likely that the overall survival rate is low because two thirds of the patients had severe invasion and/or damage to other organs [1].

These findings imply that carcinoma is riskier than gallbladder adenocarcinoma in that it has malignant capacities, which explains the low number of cases that can be cured. In addition, the 5-year survival rate after a curative resection for gallbladder carcinoma was 88.9% when tumour penetration was limited to muscularispropria [13].

Zhang's s et al. metanalysis suggests that tumour size is a good prognostic factor that marks the endurance of CSGB (tumours smaller than 5 cm had longer prolongation) as well as AUCC stage I and II and curative resection. It also demonstrated that there were no significant differences between survivors in terms of age groups, gender, gallbladder stones 'presence, epithelial and mesenchymal components [3].

The prognosis of this disease is normally poor. Most cases present with locally advanced disease. Liver metastasis and peritoneal dissemination are widespread in these cases. Adrenal glands, pancreas, diaphragm, and lower thoracic vertebrae are the other metastatic sites reported in the literature. Regional, retroperitoneal, and para-aortic lymph nodes may be involved. The mean survival time after diagnosis is usually only a few months [4].

4. Conclusion

Among the many histopathogical subtypes of gallbladder neoplasm, carcinosarcoma is an exceedingly rare type. Vague symptoms often delay the diagnosis of gallbladder cancer, contributing to its overall progression and poor outcome. Surgery represents the only potential for cure. Clinical decision making should be personalized to improve patient outcomes and survival. Hence, There are no randomized, prospective studies assessing the surgical treatments for GB cancer. Moreover, there are only a few

systematic reviews of the GB cancer surgical literature. Management of these cases should benefit from an electronic health record system and a digitized cancer registry. This will allow for data capture and analysis as well as the seamless integration of a patient's pathology, clinical algorithm, response, and survival into a national precision medicine framework.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Not required for this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Author contribution

Dr. Madani Ayoub, Dr Rachid Jabi, Pr Bouziane Mohamed: Data collection, Data analysis and interpretation, article writing.

Dr Achraf Mery, Pr Benani Amal: Interpretation of histological data.

Dr Soumia El Arabi, Pr Imane Kamaoui: Interpretation of Radiological data.

Registration of research studies

N/A.

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