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Case report: Sarcomatoid carcinoma, a rare gallbladder carcinoma.

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ABSTRACT

Background: Sarcomatoid carcinoma, a rare malignancy, is difficult to diagnose preoperatively, and often presents at a late stage with poor clinical outcome. The most common histology subtype of gallbladder (GB) carcinoma is adenocarcinoma which accounts for more than 70% of all GB tumors histologically. Sarcomatoid carcinoma of the gallbladder accounts for less than 1% of all subtypes, and there are only 109 cases reported worldwide. Case presentation: A 64 year old gentleman presented with right hypochondriac pain for 2 weeks, with a palpable mass clinically over the right hypochondriac region. Initial Imaging suggested gallbladder empyema, and was treated with antibiotics for a week, but pain was unresolved, and subsequently underwent laparoscopic cholecystectomy, which turned out to be sarcomatoid carcinoma under histopathological examination. Conclusion: Sarcomatoid carcinoma can only be diagnosed via histopathology as there are no substantiate screening tools available for now.

KEYWORDS

Sarcomatoid carcinoma, spindle cell carcinoma, cholelithiasis, laparoscopic cholecystectomy

INTRODUCTION

Gallbladder carcinoma, a rare malignancy accounts for 0.8% of all new cancers diagnosed in Malaysia, surmounting to 1.1% mortality in all diagnosed cases in 2018 [1]. According to ASCO, it is projected that 11,980 adults will be diagnosed with gallbladder and other biliary cancers in 2020 in the USA alone, in which 4 out of 10 adults are diagnosed with GB cancer. An estimation of about 4000 deaths will be from this group in 2020 [2]. The risk factors for GB carcinoma include gallstones, gallbladder polyps, age exceeding 70, H.pylori infection or carcinogen exposure [3]. The most common presentations include pain, jaundice and fever, which are often misdiagnosed as acute cholecystitis, cholangitis or gallbladder empyema. The majority of patients are offered elective operations on a later date once their acute issues have subsided, in which the gallbladder specimen obtained was reported as malignancy via histopathological examination. Sarcomatoid carcinoma is a rare variant of undifferentiated carcinoma composed of both malignant epithelial and mesenchymal components [4]. This report aims to improve the

diagnosis method and management of this rare type of carcinoma. This work has been reported in line with SCARE criteria and PROCESS guidelines [5]. We report a case of a gentleman presented with right hypochondriac pain which clinically appears as gallbladder empyema and discuss our management strategies.

CASE

The patient presents with on and off right hypochondriac pain for 2 weeks which is colicky and bloating in nature. There is no yellowish discolouration of the face, no pale coloured stools or tea colored urine, no loss of weight or loss of appetite, no history of malignancy in family, no history of alcohol consumption and he denied traditional medication intake or any high risk behaviours. On examination, the patient is alert with good hydration and is not septic looking. Examination revealed a soft abdomen with tenderness over the right hypochondriac region (Murphy's sign positive) and epigastrium, associated with guarding. There is a mass palpable over the right hypochondriac and epigastric region. He has no stigmata of chronic liver disease and bowel sounds are normal. Preliminary blood parameters including liver enzymes are in normal range.

We proceed with a CT scan of the abdomen (Figure 1) to investigate the abdominal mass. It shows that the gallbladder is enlarged and appears folded. The GB wall is enhancing the inferior portion of the GB, and is disproportionately ballooned. There is pericholecystic fluid with septation, and a layer of subtly hyperdense material in the dependent portion. A suspicious defect is seen in the folded portion of the wall at the mid section of the GB. The wall in this region measures 4.5mm. There is no hyperdense gallbladder of biliary tree calculus. The proximal common bile duct (CBD) is thickened and enhanced. The distal CBD at the region of the pancreatic head is prominent measuring 7mm. The cystic duct unable to be visualized. The enlarged GB causes mass effect onto the D1/D2 of duodenum medially. Portal vein was opacified with no filling defect. Presence of periportal edema/fluid. Based on the CT scan findings, the patient is treated in the ward as gallbladder empyema, and started on IV Cefobid 1g BD and IV metronidazole 500mg TDS for one week. His symptoms subsequently improved within a week and he was discharged well.

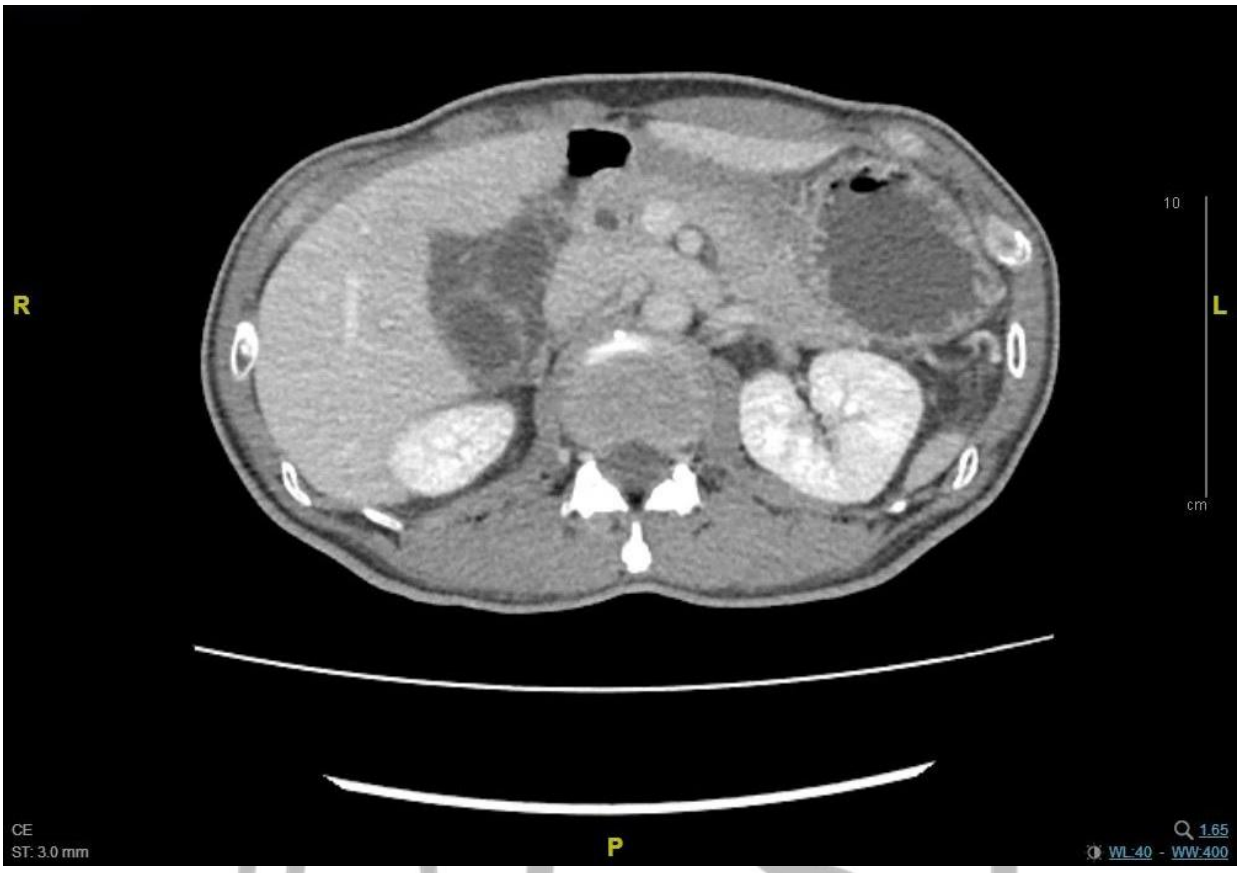

Figure 1: CT scan showed that the gallbladder is enlarged and appears folded. The GB wall is enhancing the inferior portion of the GB, and is disproportionately ballooned. A suspicious defect is seen in the folded portion of the wall at the mid section of the GB.

A week after his discharge, the patient developed abdominal pain and vomiting for 1 day. We proceeded with an ultrasound(USG) scan of the abdomen (Figure 2) which showed the gallbladder is enlarged with a thickened wall. There are multiple calculi and sludge within with minimal pericholecystic fluid. CBD is dilated and measured 0.9cm in maximum diameter. Soft tissue lesions were seen within the distal CBD.

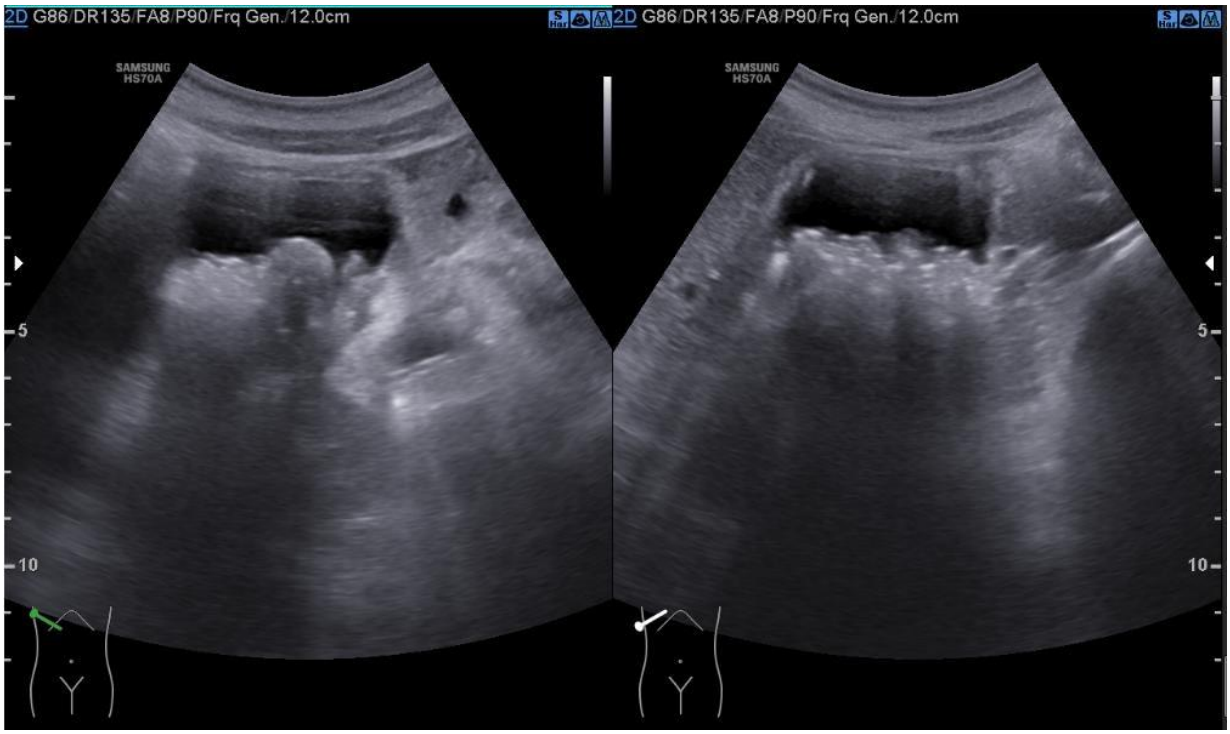

Figure 2: USG abdomen showed thickened GB wall with multiple calculi and sludge.

The patient proceeded with Endoscopic retrograde cholangio-pancreatography (ERCP) and Endoscopic Ultrasonography (EUS), which showed stone in dilated CBD, and thickened Gallbladder's wall. Pancreas was normal. CBD stones were removed via sphincterotomy, and sphincteroplasty was done. Post procedure cholangiogram showed no residual stone in CBD.

Patient was scheduled for an elective Laparoscopic Cholecystectomy, which was performed 6 weeks later. The patient was admitted one day prior to the operation date. Pre-operative assessments and blood parameters were normal. Intraoperatively,GB was distended, and its fundus had flimsy adhesion to transverse colon and stomach. The surgeon experienced difficulty in grasping the GB fundus, and suspected some stones were impacting over the fundus. Hartmann's pouch was soft, and the cystic duct was dilated. However, upon milking the cystic duct, no stone was present. The Lymph node of Lund was enlarged, measuring about 1.5cm in size. GB was successfully removed, and was dissected. Dissected GB showed (Figure 3) multiple cholesterol stones in varying sizes, the largest measured about 2cm. There was organised slough within the gallbladder, likely due to chronic inflammation and cholecystitis.

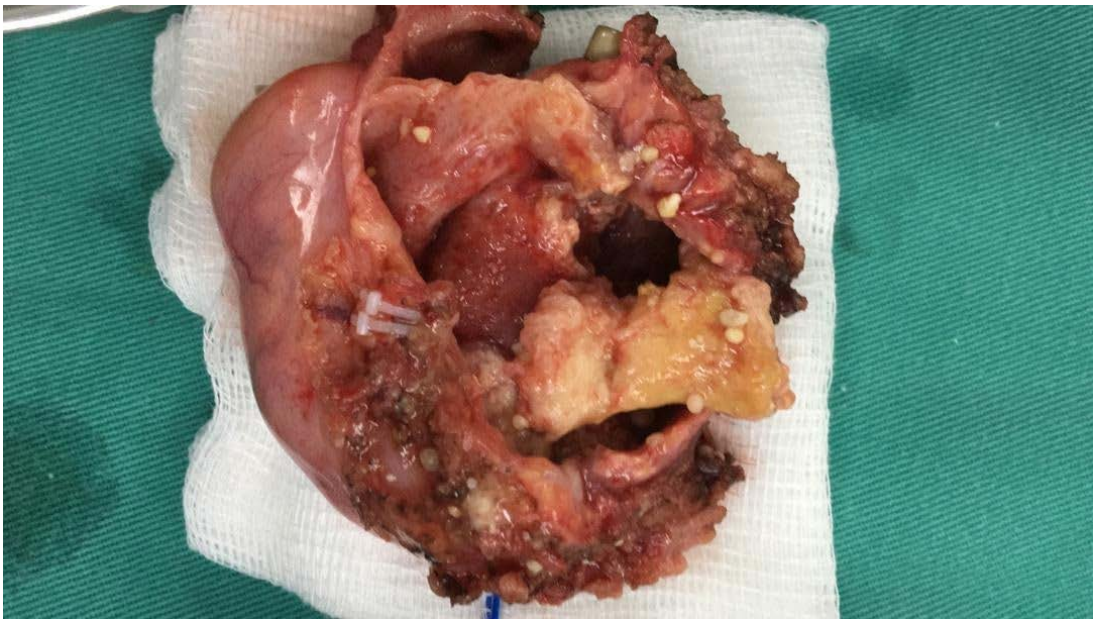

Figure 3: Dissected GB showed organised slough within GB, and irregular yellowish tan lesion near the hepatic bed. Mucosa is ulcerated with a small multinodular surface with presence of necrotic tissue.

The gallbladder and lymph node specimen were sent for histological examination. Post-operatively, the patient had an uneventful recovery and was discharged home the next day.

Gross examination for the GB specimen (Figure 3), measured 95mm in length and 30mm in greatest diameter. The GB wall was thickened, measured 3mm to 10mm. There was a firm, irregular yellowish tan lesion near to the hepatic bed measuring 20x15x10mm, which is 20mm from the cystic duct margin. The mucosa is ulcerated with some small multinodular surface which is 10mm from the cystic duct, necrotic tissue noted as well. Histology showed an infiltrative ulcerated tumour mass consist of diffuse sheets and trabeculated pattern of poorly differentiated tumour cells displaying marked nuclear pleomorphism with enlarged vesicular nuclei, prominent eosinophilic nucleoli and indistinct pale cytoplasm. These cells showed ovoid to spindly nuclei. Mitotic figures of aberrant forms were frequently seen. Marked geographical tumour necrosis and lymphoplasmacytic cells infiltrate were seen among the tumour cells.The tumour cells were strong positivity to panCK, CK7 and Vimentin. Focal positive to Actin, CD68 and EMA is suggestive of sarcomatoid carcinoma. For the lymph node specimen, there was no tumour infiltration seen.

Upon follow up for the patient a month later, a CTTAP was arranged, and showed distant metastasis to the liver and lungs. Patient opted for palliative chemotherapy, and passed away 3 months later.

DISCUSSION

According to meta analysis by *Zhang LJ*, the average median age for patients who had SC was 68.8, with female to male ratio of 3:2. The median size of the tumour is about 5cm. Two important good prognostic factors include Japanese ethnicity and tumour size of <5cm [7]. Diagnosing gallbladder carcinoma during the early stage is a challenge as only 50% of GB carcinomas are diagnosed preoperatively including imaging investigations such as CT

scans/MRI/MRCP. Routine bloods and tumour markers are not specific and sensitive to GB ca, with certain cases showing an obstructive picture if the tumour invades the biliary tract [8]. Majority of cases were diagnosed at advanced stages, and most are found incidentally during elective surgery for cholelithiasis [9]. Survival outcome of this disease depends on the tumour stage, which will affect the prognosis of the patient and the subsequent mode of management including palliative surgery in advanced cases.

During elective laparoscopic cholecystectomy, patients appear to have a survival advantage if resected with curative intent. Other methods such as radical surgical approach with the intent of achieving R0, reported to have a 5 year survival rate of average 37% in Japan and USA [10][11].For sarcomatoid carcinoma, a study conducted by *Liu KH* showed no significant benefit with radical resection of tumour, limited role of chemotherapy and radiotherapy. Mean survival rate is about 2 months in their study [12]. According to *Wada*, one patient was started with adjuvant chemotherapy with gemcitabine chloride for 3 years, after curative surgery and was followed up for 5 years without any signs of recurrence [13]. Overall prognosis of this subtype is poor, following curative resection due to recurrence, systemic metastasis or peritoneal dissemination [14].

CONCLUSION

Sarcomatoid carcinoma is a rare subtype of gallbladder carcinoma which has high mortality and morbidity. It can only be diagnosed via histopathology as there are no substantiate screening tools available for now. High clinical suspicion of malignancy should be suspected in patients presented clinically as gallbladder empyema, as malignancy can be masked by it.

ABBREVIATION

SC:Sarcomatoid carcinoma

CBD: Common Bile duct

GB: Gallbladder

CONFLICT OF INTEREST STATEMENT

There are no conflicts of interest to declare.

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This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

ETHICS APPROVAL

Ethics approval was not required from the institution for the publication of this case report.

CONSENT

Informed consent was obtained from the patient for the publication of this case report and accompanying images

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