Case Report

Sarcomatoid carcinoma of the gallbladder: A case report and review of the literature

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ABSTRACT

Sarcomatoid carcinoma of the gallbladder is a rare variant of undifferentiated carcinoma composed of both malignant epithelial and mesenchymal components. Preoperative differentiation of this tumor from adenocarcinoma is difficult because of its overlapping clinical and radiological features. However, the prognosis of this tumor is poor with a median survival of only 5.5 months. Surgical resection is the treatment choice for these tumors, but chances of recurrence are high. To our knowledge, 80 cases of sarcomatoid carcinoma of the gallbladder have been reported in the world literature. We report a case of sarcomatoid carcinoma of the gallbladder in a 65-year-old male who was treated with a radical cholecystectomy with extrahepatic bile duct resection. A hepaticojejunostomy with Roux en Y jejunojejunostomy and regional lymphadenectomy was done. A Ro resection was achieved in this case. We report this case because of its rarity, difficulty in its specific preoperative diagnosis and to emphasize the need of immunohistochemistry to confirm the diagnosis.

Key words: Gallbladder, immunohistochemistry, sarcomatoid carcinoma

INTRODUCTION

Primary gallbladder carcinoma is a malignant neoplasm with an incidence of 1.2 cases per 100,000 people each year.[1] Adenocarcinoma is the most common type of gallbladder neoplasm whereas sarcomatoid carcinoma comprises <1% of all gallbladder neoplasms.[2] It is characterized by the presence both carcinomatous and sarcomatous components, sometimes with heterologous sarcomatous elements.[3] The occurrence of this tumor in many sites such as the uterus, lung, esophagus, pancreas, and kidney is well-known.[4] Like other biliary tract disease, it is frequently associated with cholelithiasis.[5] It presents as a large polypoidal mass usually at an advanced stage and thus has a poor prognosis. Both histopathological examination and immunohistochemistry (IHC) are required for its diagnosis.[3] Patients with this tumor are generally treated with radical or palliative surgery.[2]

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CASE REPORT

A 65-year-old male presented with a 3-month history of intermittent pain in the right hypochondrium, anorexia, and weight loss. The liver was nonpalpable, and there was no free fluid in the abdomen. His laboratory examination showed hemoglobin of 12.6 g/dl, leukocyte count of 13600/cumm and normal liver function tests. The ultrasonographic (USG) examination showed a distended gallbladder with a hypoechogenic mass and multiple gall stones. Computed tomography and magnetic resonance cholangiopancreatography scan showed a polypoidal mass with a heterogeneously enhancing soft tissue density occupying the fundus and body of the gallbladder with infiltration into the adjacent liver [Figure 1a and b]. Cancer antigen (Ca) 19-9 levels were within normal limits. A cholecystectomy with segment IVb and V lobe hepatic resection along with regional lymph node dissection and extrahepatic bile duct resection was done. A hepaticojejunostomy at the confluence with Roux en Y jejujnojejunostomy was done. The surgical specimen showed a dilated gallbladder measuring 13 cm × 9.5 cm with the liver tissue attached at one side measuring 7 cm × 4 cm × 3 cm. Cut section showed a polypoidal grey white, soft, fleshy tumor measuring 8 cm × 7 cm in size and occupying the fundus and body of the gallbladder. Tumor was extending into the liver contiguously for a depth of

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1 cm and the cystic duct. Liver cut margin was 2 cm away; proximal bile duct cut margin was 0.5 cm away and distal bile duct cut margin was 1.5 cm away. Multiple gall stones were also seen [Figure 2]. Microscopic examination revealed a biphasic tumor composed of predominantly sarcomatoid areas with focal carcinomatous component in the form of few scattered glands [Figure 3]. The sarcomatous areas showed sheets of spindle cells with nuclear pleomorphism. Large areas of necrosis and frequent mitosis were seen. Tumor was infiltrating into the liver. A total of 10 lymph nodes were dissected, and all were negative for metastasis. Thus, the tumor was classified as Stage IIIA (T3N0M0). On IHC, both carcinomatous and sarcomatous components showed positivity for cytokeratin (CK) and sarcomatous component showed a strong vimentin positivity [Figure 4a and b]. Based on the histopathology and IHC findings, a diagnosis of sarcomatoid carcinoma was given. Patient had an uneventful postoperative course and was doing well 2 months after surgery.

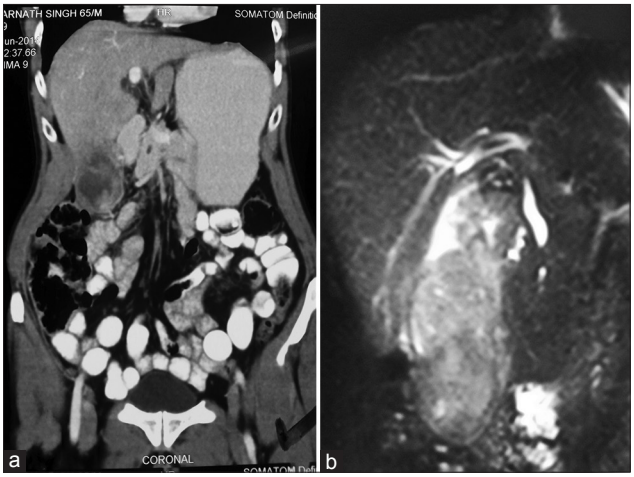

Figure 1: (a) Computed tomography scan showing wall thickening with enhancing mass in the gallbladder. (b) Magnetic resonance cholangiopancreatography scan showing a gallbladder mass causing compression and infiltration of the common hepatic duct

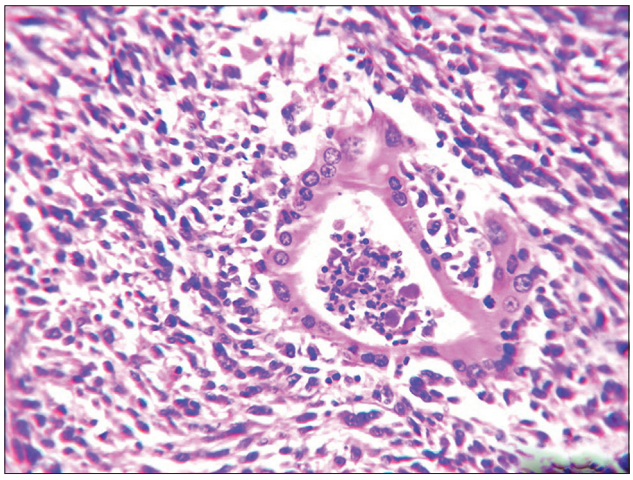

Figure 3: Microscopy of the tumor showing dual differentiation with malignant glandular cells and sarcomatous spindle cells (H and E, ×400)

DISCUSSION

Sarcomatoid carcinoma also known as carcinosarcoma or spindle cell carcinoma is a type of highly aggressive gallbladder cancer which is classified as a type of undifferentiated carcinoma according to World Health Organization classification of gallbladder tumors.[6]

Since Landsteiner *et al*. published the first case of carcinosarcoma of gallbladder in 1907, there have been <80 cases of this lesion reported in world literature.[5] It is more common in females with a female to male ratio of 2:1–5:1 with an average age range of 60–70 years.[7] Similar to adenocarcinoma these patients usually present with abdominal pain, jaundice, nausea, anorexia, and loss of weight. A palpable abdominal mass is felt in some cases. It is not associated with specific radiological findings or tumor markers like carcinoembryonic antigen or Ca 19-9.[3] USG shows a polypoidal mass as

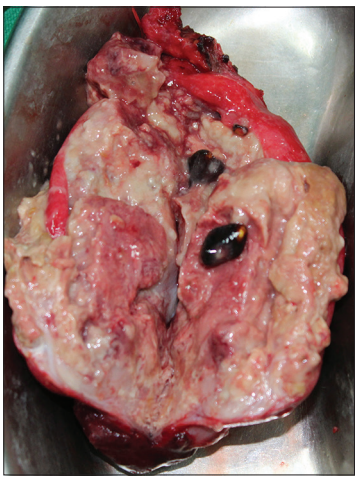

Figure 2: Gross specimen showing a soft and fleshy tumor occupying the entire gallbladder lumen along with gall stones

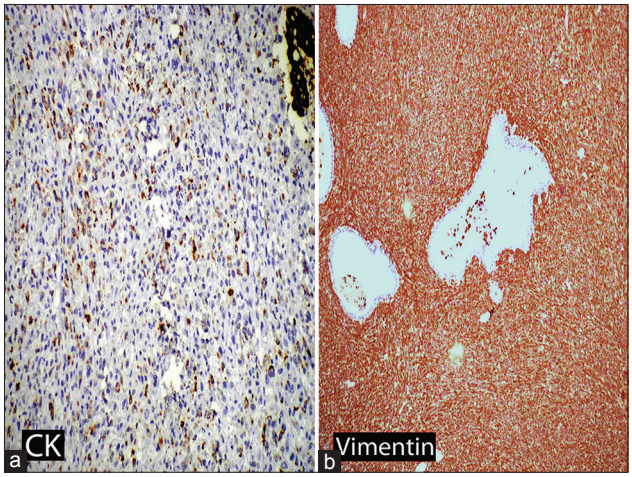

Figure 4: (a) Cytokeratin positivity seen in both glandular and stromal components (immunohistochemistry [IHC], ×100). (b) Strong vimentin positivity in the sarcoma component (IHC, ×100)

was seen in the present case. In about 70% of the cases, it is associated with gallstones.[5] Hence, preoperative differentiation of this tumor from adenocarcinoma is difficult. Park *et al*. found that this tumor presents with a much larger size than adenocarcinoma.[8] Zhang *et al*. have done a meta-analysis of 68 reported cases. They found adenocarcinoma to be the most common epithelial component in 79.2% cases while squamous carcinoma was found only in 9.4% cases, and both components were found in 11.3% cases.[9] The sarcoma component is composed of undifferentiated spindle cells sometimes mixed with heterologous elements like osteosarcoma, chondrosarcoma or a rhabdomyosarcoma.[3] To demonstrate the dual differentiation of this tumor, IHC or electron microscopy is mandatory. The tumor cells of the sarcomatous areas coexpress CK and vimentin and ultrastucturally show desmosome like junctions and aggregates of cytoplasmic intermediate filaments. These findings suggest an epithelial origin of the sarcomatous component.[10,11] The present case showed largely the spindle cell component and foci of adenocarcinoma were focal requiring thorough sampling and diligent search.

Several theories have been proposed to explain the epithelial and mesenchymal origin of these tumors: (1) A mesenchymal reaction (2) A true sarcoma (including the collision theory hypothesis) (3) A malignant proliferation of epithelial origin (including the stromal induction/metaplasia model) (4) Embryonic stem rest origin, and (5) The totipotent stem cell hypothesis.[12] Studies have linked K-ras alteration with disruption of cell cycle regulation to gallbladder carcinogenesis.[5]

The treatment of choice for sarcomatoid carcinoma is surgical excision. Cholecystectomy is sufficient if the tumor is confined to the gallbladder while more advanced disease requires the resection of 2–3 cm of liver tissue combined with lymph node dissection as was done in the present case. Sarcomatoid carcinoma of gallbladder may recur as a liver metastasis, peritoneal dissemination or as a lymph node metastasis. The median survival time is 5.5 months and the longest postsurgery survival time reported is 60 months.[5] No optimal postoperative therapy such as chemotherapy or radiotherapy have been established for this lesion because of its rarity and poor prognosis.[13]

CONCLUSION

Sarcomatoid carcinoma is a very rare type of gallbladder cancer which usually presents as a large polypoidal mass. An extensive sampling for the focal epithelial component is required for diagnosis on histopathology. Both morphology and IHC studies are required for making an accurate diagnosis of this tumor. A complete resection will significantly improve the prognosis in these cases.

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