CASE REPORTS

Carcinosarcoma of the gallbladder: report of an uncommon histological variant and a review of the literature

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Introduction

Gallbladder (GB) malignancies are usually aggressive and the most common histological type is adenocarcinoma. Carcinosarcoma of the GB is a rare histological variant.

Case Report

A 48 year old gentleman presented with abdominal pain of one month duration. Examination of the abdomen revealed a mass in the right hypochondrium without any signs of distant metastases. A computerised tomography (CT) scan of the abdomen showed a heterogenous hypodense mass in the region of GB replacing it with minimal liver infiltration (Figure 1). There was no evidence of enlarged regional nodes or distant metastasis. The serum CA 19-9 was 24.4 U/ml.

Figure 1. CT scan image showing gallbladder mass.

With a presumptive diagnosis of gallbladder cancer, he underwent a laparotomy through a rooftop incision preceded by a laparoscopy which did not reveal peritoneal spread. There was a mass in the GB which was adherent to the transverse mesocolon (Figure 2). He underwent a radical cholecystectomy. A frozen section done from the resected mass was suggestive of a malignant spindle cell neoplasm. Frozen section of the cut edge of the cystic duct was negative.

The histological examination showed a 7.5 x 6.5 x 6 cm tumor in the GB with features of malignant spindle cell sarcoma

(Figure 3). Mitosis was increased (1-2/ HPF) with large areas of necrosis. The tumor was not infiltrating into the liver and the resected liver margin was free of tumor. Eight lymph nodes were harvested and all were negative for metastasis. On immunohistochemistry (IHC), Epithelial Membrane Antigen (EMA), Keratin, Vimentin, Smooth Muscle Actin (SMA), S100-P, CD 56, CD 99 and CK 7 showed a strong positive reaction in the tumor cells. Staining with Ki67 showed that 80% of the cells showed strong nuclear positivity for Ki67. The tumor cells were negative for CK 8, CK 18, CK 20 and CEA, CD 34, CD 31, CD 117 and Calponin. After IHC correlation, the final impression was a high grade spindle cell carcinoma/carcinosarcoma of the gall bladder. Adjuvant chemotherapy was planned for this patient.

Figure 2. Intra-operative picture.

Figure 3. Histopathology microphotograph magnified 40X. (a. Haematoxylin and Eosin stain b. Keratin positivity in squamous carcinoma components c. Strong Vimentin positivity in the sarcoma component d. Strong Cytokeratin-7 positivity).

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Discussion

Carcinosarcoma accounts for less than one percent of all GB neoplasms. The first case was reported in the literature by Landsteiner in 1907 [1]. Subsequently fewer than 100 cases have been reported in the English literature. Histologically it is characterized by the presence of both epithelial and mesenchymal components. Several theories have been proposed to explain the aetio-pathogenesis including a mesenchymal reaction, the collision neoplasm hypothesis, stromal induction/metaplasia model, an embryonic cell rest origin, and the totipotent stem cell hypothesis [2].

The serum alpha-fetoprotein and serum granulocyte colony-stimulating factors may be elevated. The disease is more commonly seen in females and the elderly, with a median age of 68 years.

The prognosis of carcinosarcoma of the GB is poor [3]. In a meta-analysis of 68 cases, the reported 1-year and 5-year survival was $19 \pm 5\%$ and $16 \pm 5\%$ respectively [4]. The factors associated with a good prognosis included a tumor size of less than 5cm, the Japanese race, curative resection and early stage disease [3,4]. Adjuvant chemotherapy has not been shown to improve survival [5].

Conclusion

Surgery is the mainstay in the treatment of Carcinosarcoma of GB. In the absence of large case series and trials, the role of adjuvant therapy is not clear.

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Key Points:

- Surgical resection offers the best chance of cure for carcinosarcoma of gall bladder.
- Currently there are no treatment guidelines and most cases are treated on an individual basis.
- The role of adjuvant treatment is not clear at present.