



Sarcomatoid Cholangiocarcinoma: An Unusual Tumor Posing Diagnostic Challenges

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Abstract

Keywords

- ▶ sarcomatoid
- ▶ cholangiocarcinoma
- ▶ histopathology
- ▶ immunohistochemistry

Sarcomatoid intrahepatic cholangiocarcinoma is an uncommon tumor that accounts for fewer than 1% of hepatobiliary system cancers. Frequently exhibits weight loss, fever, and stomach pain. This case study describes a 72-year-old woman who had been experiencing nausea, vomiting, and stomach pain for the previous 2 months. Histopathology and immunohistochemistry confirmed the diagnosis of sarcomatoid cholangiocarcinoma.

Introduction

An uncommon form of malignant tumor known as sarcomatoid cholangiocarcinoma is a subtype of biliary system cancer that is distinguished by the presence of mesenchymal and epithelial cells that are malignant.¹ With a median survival time of only 6 to 11 months, the prognosis is dismal and the exact pathophysiology remains unclear.² Sarcomatoid carcinoma is more commonly seen in the uterus as a malignant Mullerian tumor, although it is also occasionally discovered in the kidneys, lungs, gastrointestinal tract, and gallbladder. Surgical excision is the main recommended treatment; however, patients still have a poor prognosis due to the challenging diagnosis, frequent metastases to other organs, and invasion of adjacent vasculature. Vimentin has been positively identified in the mesenchymal component and cytokeratins in the epithelial component using immunohistochemistry.² This case study features a 72-year-old female who presented with nausea, vomiting, abdominal pain, and weight loss during the past 2 months. There was a 4 × 4 cm palpable lump in the right hypochondrium. The values of alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), and serum CA 19.9 were all within normal ranges. Sarcomatoid cholangiocarcinoma was iden-

tified by immunohistochemistry and histopathology. After the procedure, the patient was treated symptomatically and given chemotherapy advice.

Case Report

The patient, a 72-year-old woman, complained of weight loss, stomach pain, and vomiting. Contrast-enhanced computed tomography abdomen and pelvis was done and reported as thickened and contracted gallbladder with a large peripherally enhancing hypodense lesion along the fundus extending into the right lobe of the liver suggestive of neoplastic etiology. Few peripherally enhancing lesions were present in the right lobe of the liver with peripancreatic, perigastric lymphadenopathy suggestive of metastasis. Multiple tiny nodular opacities in both lungs were found which was also suggestive of metastasis. Small hiatus hernia with thickening of pylorus of stomach was also noted.

Upper gastrointestinal endoscopy was done and reported as a small sliding hiatus hernia, gastric outlet obstruction, and duodenal stricture likely benign.

All routine hematological investigations were within normal limits. Tumor markers such as AFP, CEA, and CA 19.9 were detected within normal ranges.

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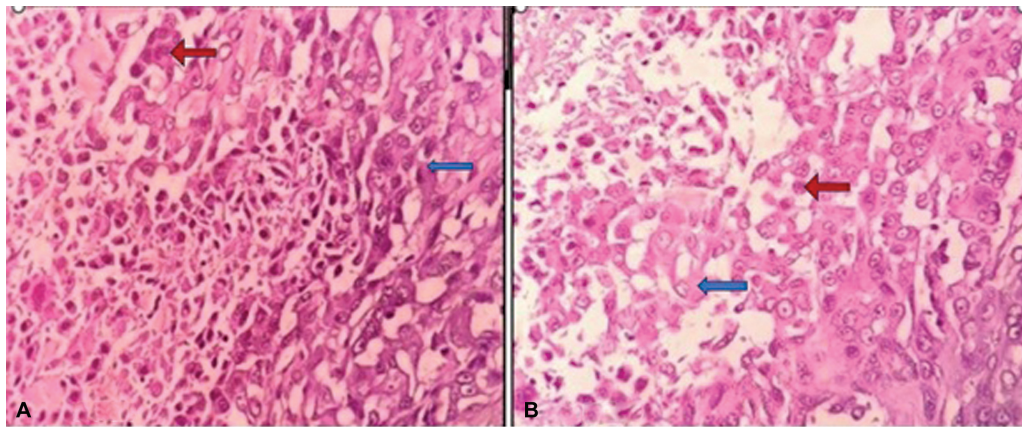


Fig. 1 (A, B) Tumor cells having round to spindled morphology with eosinophilic cytoplasm (marked with blue arrow) and pleomorphic vesicular nuclei (marked with red arrow) (hematoxylin and eosin [H&E], $\times 400$).

Ascitic fluid cytology was done. The smears showed scant cellularity with pleomorphic tumor cells arranged in singles and tiny clusters against a hemorrhagic background. They displayed high nucleus-to-cytoplasm ratio, coarse chromatin, prominent nucleoli, irregular nuclear margins, multinucleation, and moderate cytoplasm. The International Academy of Cytology system of reporting fluid cytology classification was category 5: Malignant effusion favoring poorly differentiated carcinoma.

Multiple biopsies taken from the falciform ligament, stomach, and gallbladder were sent for histopathology. Grossly multiple fibrofatty tissue bits, largest measuring $6 \times 3.5 \times 2$ cm, were found. On the cut surface, multiple pale white nodular lesions were noted, largest measuring $2 \times 1.5 \times 1.5$ cm. The entire cut surface was given for processing. Microscopic sections studied from the tissue sent as falciform ligament, stomach, and gallbladder showed fibrofatty tissue with tumor cells having round to spindled morphology, pale eosinophilic cytoplasm, and marked pleomorphic vesicular nuclei. Extensive areas of coagulative necrosis, many atypical mitoses, and few binucleated and multinucleated tumor giant cells were also seen (\rightarrow Fig. 1A, B). Features were suggestive of malignant undifferentiated tumor. Differential diagnoses were high-grade epithelioid gastrointestinal stromal tumor and poorly differentiated carcinoma. Immunohistochemistry showed tumor cells positive for CK7, CK 19, vimentin, and pan CK (\rightarrow Fig. 2A–D) and negative for CK20, CD117, DOG1, Hepar1, GATA3, TTF1, PAX8, WT1, and P40. Ki67 proliferation was 60%. Features were those of sarcomatoid cholangiocarcinoma.

Discussion

Intrahepatic cholangiocarcinoma is said to be responsible for 4.5% of cases of cholangiocarcinoma and fewer than 1% of hepatobiliary system cancers.³ The World Health Organization defines sarcomatoid intrahepatic cholangiocarcinoma (S-iCCA) as intrahepatic cholangiocarcinoma with sarcomatoid alterations.² It is a very uncommon cancer that exhibits bad prognosis. It is discovered that it is more prevalent in the older age group and female sex.⁴ Symptoms or indicators that

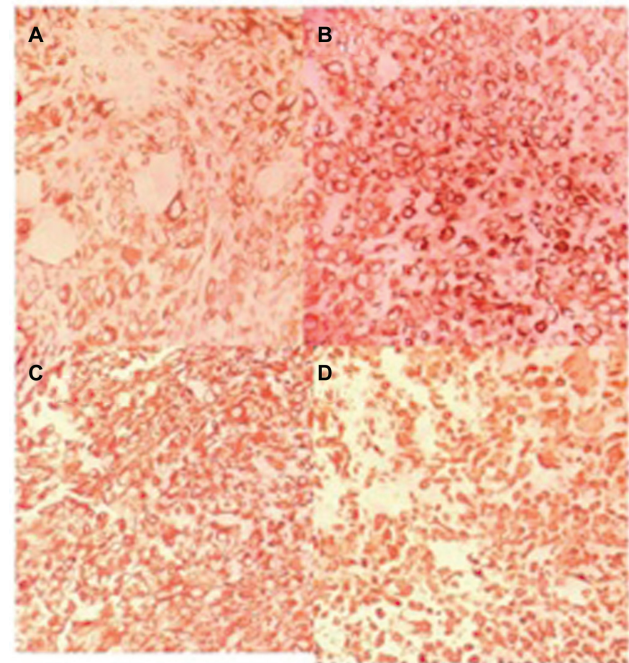


Fig. 2 (A) Tumor cells with CK7 positive, (B) CK19 positive, (C) vimentin positive, and (D) pan CK positive ($\times 400$).

are not identifiable, like fever, abdominal pain, nausea, vomiting, exhaustion, and weight loss, are frequently seen in them. Fever and abdominal pain are the most typical symptoms.⁵ In this instance, the patient complained of weight loss, stomach pain, and vomiting. Only a biopsy can provide a conclusive diagnosis of S-iCCA, and both histopathological and immunohistochemical analyses are necessary.

Histopathological examinations of S-iCCA reveal the coexistence of differentiated, spindle-shaped sarcomatoid cells in bundles or weaves with adenocarcinoma cells. According to immunohistochemistry, S-iCCA cancers are negative for HepPar-1 and positive for the mesenchymal tumor markers vimentin and epithelial cholangiogenic tumor markers CK7 and CK8. HepPar-1, a hepatocyte marker, offers helpful diagnostic information in differentiating hepatocellular carcinoma from cholangiocarcinoma and metastatic liver carcinoma.⁶

In this case, histopathology showed tumor cells having round to spindle morphology, pale eosinophilic cytoplasm, and marked pleomorphic vesicular nuclei. Extensive areas of coagulative necrosis, many atypical mitoses, and few binucleated and multinucleated tumor giant cells were also seen. Immunohistochemistry showed tumor cells positive for epithelial markers like CK7 and CK19 and mesenchymal markers like vimentin and negative for Hepar1. Currently, the only available treatment for patients with S-iCCA is radical liver resection, and there are no pertinent criteria for predicting the prognosis and survival of these patients. Compared with other biliary tumors, S-iCCA is shown to be less sensitive to chemotherapy and radiation, and its overall 1-year survival rate is nearly zero.⁷ Patients with S-iCCA who had surgical resection had a median survival of 11 months, which is similar to that of patients with regular iCCA who did not have surgery (8 months). Patients with S-iCCA who did not have surgery had a 3-month median survival rate. Adjuvant chemotherapy following surgery with cisplatin, doxorubicin, cyclophosphamide, and taxol has been demonstrated in multiple studies to extend survival in patients with sarcomatoid carcinomas. The sole effective treatment for gallbladder sarcomatoid cholangiocarcinoma is surgical resection, and early detection is essential for a better prognosis.⁴ Studies determining the standard of care are insufficient. Some articles recommend gemcitabine, although there is not enough proof to support this.⁸

Conclusion

The sole effective treatment for gallbladder sarcomatoid cholangiocarcinoma is surgical resection, and early detection is essential for a better prognosis. Since no adjuvant therapies have shown to be successful to date, more proof of the

combined adjuvant chemotherapy's benefit for survival is required for pathology-based chemotherapy. After surgery, overall survival is not very good.

Author's Contribution

The manuscript has been read and approved by all the authors, that the requirements for authorship have been met, and each author believes that the manuscript represents honest work.

Conflict of Interest

None declared.

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