



# Sarcomatoid Carcinoma of the Ampulla of Vater: A Case Report

바터 팽대부에 발생한 원발성 육종성암: 증례 보고

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Sarcomatoid carcinomas are rare malignant tumors comprising a mixture of malignant epithelial and mesenchymal cells. Sarcomatoid carcinomas are found at various sites of the body, including the lungs and the hepatobiliary, urinary, and digestive tracts. However, sarcomatoid carcinoma of the ampulla of Vater (AoV) is rare with only a few reported cases. Here, we present the case of a 60-year-old woman with sarcomatoid carcinoma of the AoV, diagnosed using CT and MRI.

**Index terms** Case Report; Ampulla of Vater; Sarcomatoid Carcinoma; CT; MRI

## INTRODUCTION

Sarcomatoid carcinomas are rare malignant tumors with both carcinomatous and sarcomatous components. These neoplasms have been documented at multiple anatomical sites, including the gastrointestinal tract, lungs, and kidneys, and are histologically described using terminologies such as carcinosarcoma, malignant mixed tumor, and spindle cell, pseudosarcomatous, and metaplastic carcinomas (1). Sarcomatoid carcinoma of the ampulla of Vater (AoV) is rare, and only a few cases have been reported. Here, we present the case of a 60-year-old woman with sarcomatoid carcinoma of the AoV, including detailed CT and MRI findings.

## CASE REPORT

A 60-year-old woman was presented at the emergency department with dyspepsia and jaundice. The patient had no relevant medical history. Laboratory analysis revealed hyperbilirubinemia (total bilirubin level 7.37 mg/dL) and elevated transami-

Received April 23, 2025  
Revised July 5, 2025  
Accepted July 30, 2025  
Published Online January 6, 2026

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nase levels (aspartate aminotransferase, 237 U/L; alanine aminotransferase, 368 U/L).

Multiphase abdominal CT was performed to further evaluate the cause of hyperbilirubinemia. The CT revealed a  $3.0 \times 2.9$  cm-sized enhancing infiltrative soft tissue lesion in the duodenal ampullary region (Fig. 1A). The lesion showed mild heterogeneous enhancement in the arterial phase and mild washout in the portal and delayed phases. (Fig. 1A; lower magnification). Extrahepatic and intrahepatic duct dilatation was also observed. However, the pancreatic duct remained unaffected. No evidence of lymphadenopathy was observed on the CT scan.

The mass was further evaluated using MR cholangiopancreatography (MRCP) and pancreaticobiliary MRI. Axial and sagittal MRI revealed a  $3.0 \times 2.8$  cm-sized ampullary mass with an infiltrative margin, which showed low signal intensity on axial T1 weighted images (T1WIs) and homogeneous mild high signal intensity on coronal T2WI (arrows, Fig. 1B). The mass demonstrated mild heterogeneous enhancement during the arterial phase and mild washout compared to the arterial phase enhancement during portal and delayed contrast-enhanced T1WI (Fig. 1C). It also showed high signal intensity on diffusion-weighted images with a b-value of 1,000 and a low value on the corresponding apparent diffusion coefficient map (Fig. 1D). MRCP showed dilatation of the common bile and upstream intrahepatic ducts, with abrupt narrowing in the distal portion of the common bile duct.

Esophagogastroduodenoscopy revealed a mass in the AoV with an ulcerative protruding appearance (Fig. 1E). An endoscopic biopsy of the visible portion was performed, and the results showed a poorly differentiated carcinoma.

Considering the CT and MRI features, biopsy results, and high prevalence, the patient was clinically diagnosed with adenocarcinoma of the AoV and scheduled for surgical excision.

The patient underwent pancreaticoduodenectomy, and upon gross examination, the tumor showed an irregular ulcerative lesion measuring  $3.0 \times 3.5$  cm in the duodenal ampullary region. The tumor directly invaded the pancreatic head, a finding associated with a less favorable prognosis. Microscopically, the tumor contained two distinct cell components: tubular adenocarcinoma tissue and diffused infiltrative sarcomatous cells. The sarcomatoid component represented approximately 90% of the total tumor volume, whereas the adenocarcinoma component accounted for approximately 10% of the tumor volume. Immunohistochemically, the tumor was diffusely positive for cytokeratin (Fig. 1F) and epithelial membrane antigens. Based on these histopathological features, the tumor was classified as a sarcomatoid carcinoma. The patient received postoperative chemotherapy with fluorouracil and underwent follow-up CT. No evidence of recurrence was observed for one year.

This study was approved by our Institutional Review Board (IRB No. 2025-03-004). Given the retrospective nature of the study, the need for informed consent was waived.

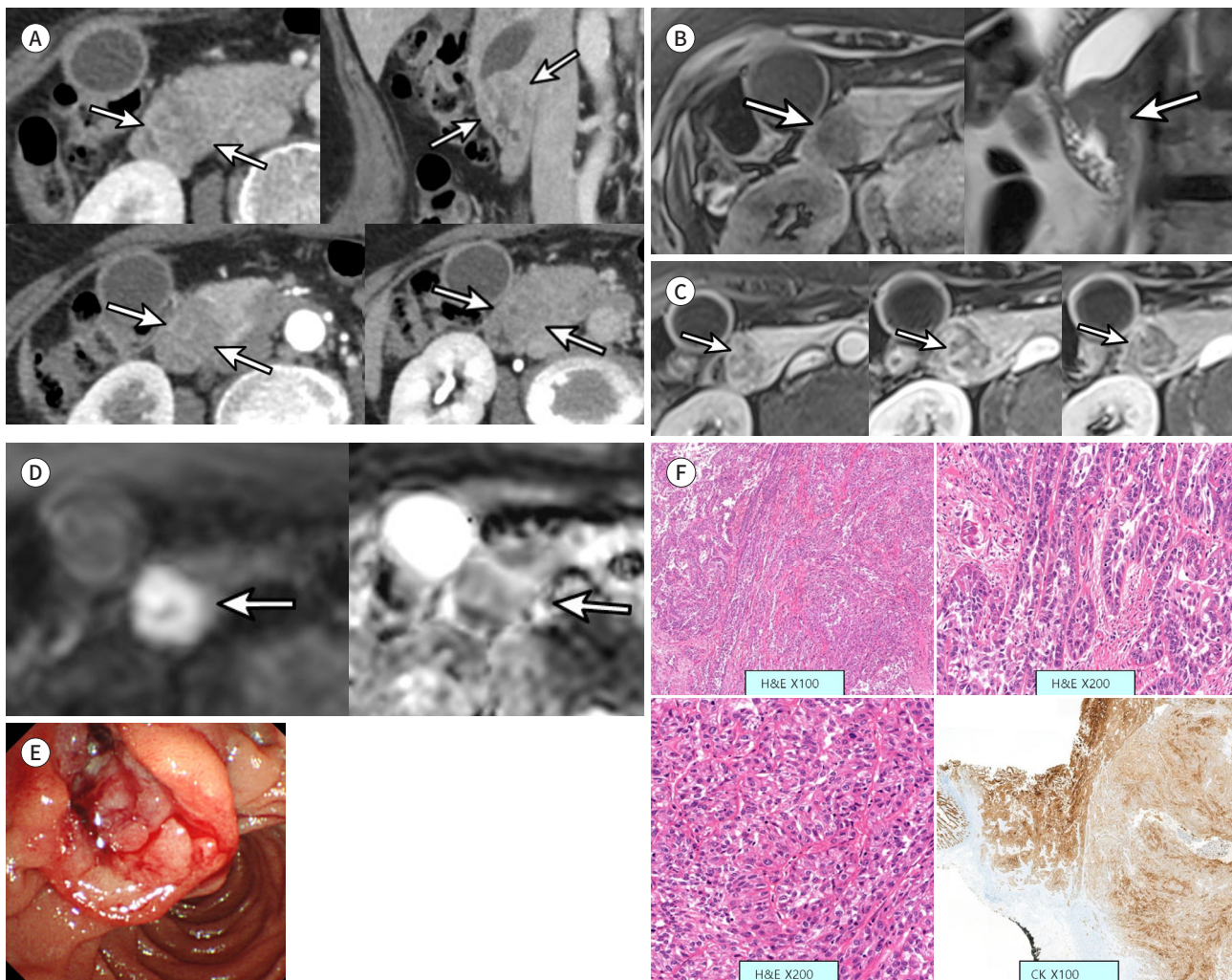
## DISCUSSION

Sarcomatoid carcinomas are rare tumors consisting of both epithelial and mesenchymal elements. They are described by many different names, including carcinosarcomas, metaplastic, spindle cell, and pleomorphic carcinomas. These tumors have been found at various sites in the body, including the digestive tract. The esophagus is the most common site of sar-

comatoid carcinoma in the gastrointestinal tract (1). Published reports of sarcomatoid adenocarcinomas of the AoV are rare. To our knowledge, only a few cases of sarcomatoid carcinoma originating at the AoV have been reported in English literature (2). Because to its rarity, little is known about AoV sarcomatoid carcinomas. Sarcomatoid carcinomas are rarely diagnosed preoperatively based solely on biopsy or radiologic imaging because of their mixed

**Fig. 1.** A 60-year-old woman with sarcomatoid carcinoma of the AoV.

- A.** Contrast-enhanced abdominopelvic CT images. Axial (left upper) and coronal (right upper) CT images reveal an infiltrative soft tissue density lesion (arrows) in the duodenal ampullary region with dilatation of the common bile duct. The lesion shows mild heterogeneous enhancement throughout the arterial phase (left lower) and mild washout through the delayed phases (right lower).
  - B.** Axial and coronal T1 and T2 weighted MRI. These images reveal a periampullary mass with an infiltrative margin, showing low signal intensity on the axial T1WI (arrow, left) and homogeneous mildly high signal intensity on the coronal T2WI (arrow, right).
  - C.** Axial contrast-enhanced T1WI on MRI. The mass exhibits mild, heterogeneous arterial enhancement (arrow, left), mild washout in the portal phase-enhanced T1WI (arrow, middle) and 3-minute delayed image (arrow, right).
  - D.** Axial diffusion-weighted images on MRI. The lesion shows high intensity on diffusion-weighted images with a b-value of 1000 (arrow, left) and mildly low intensity on the corresponding apparent diffusion coefficient map (arrow, right).
  - E.** Esophagogastroduodenoscopy. It reveals a protruding mass with ulceration in the AoV.
  - F.** Microscopic findings of a tumor. The tumor (upper left, H&E stain,  $\times 100$ ) consists of an adenocarcinoma component (upper right, H&E stain,  $\times 200$ ) and sarcomatoid components (lower left, H&E stain,  $\times 200$ ). Immunohistochemical staining shows positive cytokeratin staining in the sarcomatoid and carcinomatous tumor cells (lower right, CK  $\times 100$ ).
- AoV = ampulla of Vater, H&E = hematoxylin-eosin, T1WI = T1-weighted image, T2WI = T2-weighted image



histologic nature, and are mostly revealed after surgical resection (3).

Patients with sarcomatoid carcinoma of the AoV reportedly experience jaundice, abdominal pain, or melena (2). They frequently show metastasis and recurrence after surgery, and the majority of other reported cases have very poor prognoses (4).

Several modalities, including CT, MRI, and US, have been used to differentiate and diagnose sarcomatoid carcinomas of the AoV in previously published case reports. All accessible studies showed CT findings that revealed prominent masses >2 cm at the AoV, typically described as mild, enhanced infiltrative masses (2). Izumi et al. (5) reported the US findings of sarcomatoid carcinoma and described it as a non-homogenous, low-echoic tumor mass. Other publications were not radiologically applicable, with only gross and pathologic findings described in the case reports.

Our case report includes CT and MRI findings with radiological features of a sarcomatoid carcinoma arising from the AoV. CT and MRI showed an infiltrative solid mass with mild heterogeneous enhancement, similar to previous reports of sarcomatoid carcinomas in the AoV (2, 5, 6). MRI findings of sarcomatoid carcinoma show early contrast enhancement and mild washout compared to arterial phase enhancement during portal and delayed contrast-enhanced T1WI. The enhancement pattern is atypical for conventional ampullary adenocarcinoma, which often demonstrates delayed enhancement owing to its desmoplastic stromal components (7). We presumed that this atypical enhancement pattern was related to the presence of hypervascular sarcomatous elements, suggesting a point for distinguishing between sarcomatoid carcinomas and adenocarcinomas.

Another atypical finding was the homogeneous and mildly high signal intensity on T2WI. Aggressive adenocarcinomas are mostly heterogeneous owing to necrosis or cystic degeneration within their complex glandular formations (7). We propose that this homogeneity on T2WI directly correlates with the pathological finding of the tumor, which is predominantly composed of a uniformly dense sarcomatoid component.

AoV sarcomatoid carcinoma presents a significant diagnostic challenge because its imaging features often overlap with those of the more prevalent ampullary adenocarcinomas (3, 8). A few case reports of sarcomatoid carcinomas originating in other sites of the body suggested that differences in contrast enhancement patterns with well-defined internal borders might help differentiate sarcomatoid carcinomas from more common epithelial tumors, although these findings are nonspecific (9, 10). This finding was thought to be due to differences in the proportions of its components. In our case, the mass in the AoV was predominantly composed of sarcomatoid components, and compared with the gross and pathological findings, the enhancement pattern did not allow for differentiation between carcinomatous and sarcomatous areas. Further research should be conducted in more cases using multiple modalities, including contrast-enhanced CT and MRI with enhancement.

Herein, we report a rare case of sarcomatoid carcinoma involving the AoV and describe its imaging features, of which only a few have been reported to date. While its imaging features can overlap with those of adenocarcinoma, the combination of an ampullary mass with early heterogeneous enhancement and mild washout on dynamic contrast-enhanced imaging and homogeneous T2 signal intensity suggests sarcomatoid carcinoma in the differential diagnosis. Our detailed CT and MRI findings provide critical insights into the imaging characteris-

tics of this rare entity, and may aid in distinguishing it from other periampullary malignancies. This case expands the limited radiological literature on AoV sarcomatoid carcinomas and offers a practical reference for clinicians and radiologists encountering similar lesions.

### Author Contributions

Conceptualization, G.J.I., J.S.K.; data curation, G.J.I.; methodology, G.J.I.; project administration, G.J.I.; resources, G.J.I., H.E.M.; supervision, J.S.K.; writing—original draft, G.J.I.; and writing—review & editing, all authors.

### Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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### Funding

None

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## 바터 팽대부에 발생한 원발성 육종성암: 증례 보고

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육종성암은 매우 드물게 발생하는 상피세포와 간엽조직이 혼재하는 악성종양이다. 육종성암은 소화기관, 폐, 비뇨기계 등 다양한 곳에서 생기며, 바터 팽대부에 생기는 경우는 매우 드문 것으로 알려져 있으며 몇 개의 증례로만 보고되었다. 저자들은 한 여성 환자의 바터 팽대부에 생긴 육종성암의 증례와 함께 컴퓨터단층영상과 자기공명영상 소견을 보고하고자 한다.

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