A liver carcinosarcoma is a malignant tumor containing a mixture of carcinomatous and sarcomatous elements (1). Liver carcinosarcomas are classified as two types including sarcomatoid hepatocellular carcinoma (HCC) and cholangiocarcinoma. A sarcomatoid HCC is a carcinoma that most resembles a sarcoma found in the liver. Only a few cases of intrahepatic sarcomatoid cholangiocarcinomas have been reported (1–6). To the best of our knowledge, an intrahepatic sarcomatous cholangiocarcinoma with portal vein thrombosis had not been reported. We report here the first case of a sarcomatoid cholangiocarcinoma with portal vein thrombosis in the English language literature.

**Case Report**

A 53-year-old man was admitted to our hospital with a liver mass. CT and MR images showed the presence of a multilobulated mass with hemorrhagic and necrotic foci in the left hepatic lobe. In addition, thrombosis of the left portal vein was noted. The patient underwent surgery for total removal of the mass. A pathological examination revealed the presence of an intrahepatic sarcomatoid cholangiocarcinoma with portal vein thrombosis. The patient underwent a left hepatectomy and chemotherapy. However, the disease recurred with metastases after five months. The patient underwent surgery and chemotherapy for the recurred tumor.

**Index words**: Liver neoplasms
Thrombosis
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vein that had the same enhancement pattern as compared to the mass (Fig. 1).

As depicted on MR imaging, the mass was observed as hypointense on a T1-weighted image and was observed as hyperintense on a T2-weighted image (Fig. 2). Several hemorrhagic foci were hyperintense within the mass as seen on a T1-weighted image. For contrast-enhanced MR imaging, thick contrast enhancement around the tumor as seen on an arterial phase image and concentric filling of contrast material was seen on portal and delayed phase images. The tumor thrombi of the left portal vein were also noted as hypointense as seen on a T1-weighted image and were noted as hyperintense as seen on a T2-weighted image. The tumor thrombi had thin rim-like enhancement as seen on an arterial phase image and concentric filling of contrast media as seen on portal and delayed phase images. There was no intrahepatic bile duct dilatation. The preoperative differential diagnosis was cholangiocarcinoma based on the enhancement pattern. However, we could not rule out a hepatocellular carcinoma as the patient had underlying liver cirrhosis and portal vein thrombosis.

The patient underwent a left hepatectomy with a portal vein thrombectomy. The cut surface showed the presence of a multilobulated, white to yellowish solid mass that measured 5.5 × 4.5 cm at the greatest dimension. Several small areas of necrosis and hemorrhage were detected in the intrahepatic tumor mass. In addition, tumor thrombi were present within the left portal vein. The surrounding parenchyma showed cirrhotic change.

A histological examination of the tumor showed a malignant neoplasm with poorly differentiated adenocarcinoma and sarcomatous components. An immunohistochemical examination of the sarcomatous components showed positive staining for vimentin, cytokeratin and low molecular cytokeratin (Fig. 3). Based on these histological and immunohistochemical findings, the tumor was diagnosed as a cholangiocarcinoma with sarcomatous changes.

Fig. 1. A mass in the left hepatic lobe is shown on abdominal dynamic CT (A) arterial, (B) portal and (C) delayed phase images. The mass has thick peripheral enhancement as shown on an arterial phase image and concentric filling of contrast media as shown on portal and delayed phase images. D. As shown on a portal phase image, tumor thrombi within the left portal vein are noted [arrow].
Fig. 2. A. An axial T1-weighted gradient echo image shows a well-defined hypointense mass with a focal hyperintense lesion. 
B. An axial T2-weighted single shot fast spin-echo image reveals the hyperintense mass. 
C. An axial T1-weighted image obtained during the arterial phase shows peripheral ring-like enhancement. 
D. An axial T1-weighted image obtained during the delayed phase demonstrates concentric filling of contrast media. 
E. An axial T1-weighted image obtained during the delayed phase demonstrates tumor thrombi with thin rim-like enhancement in the left portal vein (arrow).

Fig. 3. A. The histological appearance of the sarcomatoid cholangiocarcinoma is shown. Hematoxylin and eosin staining (×200) shows the cholangiocarcinoma mixed with bundles of atypical spindle cells. 
B. The appearance of the sarcomatoid cholangiocarcinoma is shown after immunohistochemical staining. The sarcomatous component (arrows) is stained for low molecular cytokeratin (×400).
The patient underwent a left hepatectomy and chemotherapy. However, the disease recurred with metastases after five months. The patient underwent surgery and chemotherapy for the recurrent tumor.

**Discussion**

Most sarcomatoid carcinomas in the liver are thought to be sarcomatoid HCCs. An intrahepatic sarcomatoid cholangiocarcinoma is extremely rare [1–6]. Radiological findings for intrahepatic sarcomatoid cholangiocarcinomas have not been established.

As depicted on dynamic CT and MR imaging, intrahepatic cholangiocarcinomas have irregular rim-like, band-like contrast enhancement around the tumor as seen on early phase images, with progressive and concentric filling of contrast material as seen on late phase images. The findings for the present case were similar to the findings for the enhancement pattern of a cholangiocarcinoma. The first presumptive diagnosis was an intrahepatic cholangiocarcinoma. However, portal vein thrombosis is not a common manifestation in cholangiocarcinomas.

The etiologies of portal vein thrombosis are liver cirrhosis, liver malignancy including HCC, metastases, pancreatic carcinoma and cholangiocarcinoma, pancreatitis, a hypercoagulable state and idiopathy [7]. Portal vein thrombosis in patients with liver cirrhosis is usually associated with the presence of an HCC. Most HCCs show early enhancement on hepatic arterial phase images and rapid washout on portal and delayed phase images. In our case, the enhancement pattern was different from an HCC. Portal vein thrombosis and liver cirrhosis are suggestive findings of an HCC. It was difficult to make the differential diagnosis between a cholangiocarcinoma and HCC in this case.

It has been reported that the prognosis for an intrahepatic sarcomatoid cholangiocarcinoma is worse as compared to an intrahepatic cholangiocarcinoma [8]. The factors that account for the poor prognosis may be ascribed to the remarkable intrahepatic development, especially the high potential of the sarcomatous component to metastasize to a variety of regions [5, 8]. The effectiveness of chemotherapy and radiation therapy for intrahepatic sarcomatoid cholangiocarcinomas has not been well investigated well and remains unclear.

In conclusion, we have reported the first case of a sarcomatoid cholangiocarcinoma with portal vein thrombosis. A sarcomatoid carcinoma should be included in the etiology of portal vein thrombosis.

**References**

문맥혈전을 동반한 간내육종형담관암:
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임재훈 ∙ 김진용 ∙ 허숙희 ∙ 정용연 ∙ 강형근

53세 남자가 간의 종괴를 주소로 내원하였다. CT와 MRI에서 간 좌엽의 출혈과 정상부위를 포함하는 대엽성의 종괴가 있었으며, 좌측 문맥에 혈전이 관찰되었다. 수술을 시행하였으며, 문맥 혈전을 동반한 육종형 담관암으로 진단되었다. 환자는 좌엽 절제술을 시행 받고 나서 항암 치료를 받았다. 그러나 5개월 후 병변이 재발하였고 전이되어 재수술과 항암 치료를 받았다.