

# A Rare Of Malignancy In The Young Patient: Cholangiocarcinoma

Uğur Ergün, Gülhan Zorgör Uçdu, Merve Nur Ak, Erkan Çağlar, Emrah Akay, Eren Altun

**Abstract:** Cholangiocarcinoma (CCA) is one of the primary malignant tumors of the liver caused by biliary epithelium. Approximately 15-25 percent of hepatobiliary malignancies. The incidence is increasing with age and it is generally seen between the ages of 50-70. Many reasons such as infection, biliary stasis and carcinogens are risk factors and the exact cause is unknown. The clinical picture of CCA varies according to the localization of the solitary lesion. Surgical treatment is the only curative method for patients with CCA. It is not very common for cholangiocarcinoma, a malignancy of hepatobiliary origin, to appear at an early age. In this article, we present a case of CCA in a young patient presenting with nausea, weight loss and abdominal pain.

**Index Terms:** Biliary, cholangiocarcinoma, malignant, icterus, pain, patient, young,

## INTRODUCTION

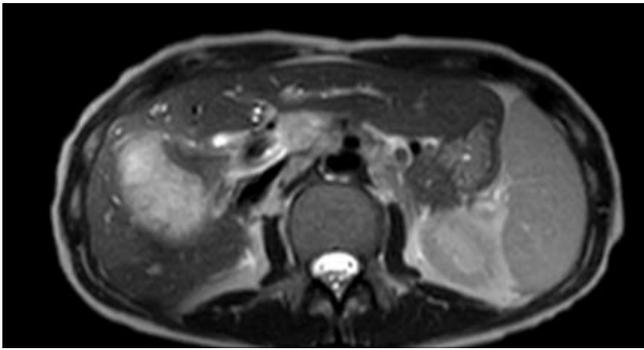
Cholangiocarcinoma (CCA) is a malignant tumor originating from biliary epithelial cells. The incidence and mortality rates of CCA in the world are increasing (1). Usually seen in older men aged between 50 and 70 years. It constitutes 15-25 percent of all hepatobiliary malignancies. Major risk factors; primary sclerosing cholangitis (PSK), choledochal cysts, ulcerative colitis, parasitic diseases of the bile duct and recurrent pyogenic cholangitis. In recent studies, HCV infection has been reported to be a risk for CCA. Although these patients usually present with continuous and painless jaundice, they may also occur with intermittent biliary obstruction due to necrosis and spillage of the tumor and positive blood stains in the stool (2-4). The 5-year survival rate, including cases with early diagnosis of CCA, is around 5-10 %. The 5-year survival rate is 25-30 percent in cases that can be surgically treated. In cases with metastasis, the average life expectancy is not more than 8-12 months, even if any treatment is performed (5). CCA triggered inflammation, environmental and genetic factors predicted to be a stem cell disease, the incidence of malignant malignant disease with a poor prognosis increases with age. Although CCA has an increased incidence with age, it has the potential to occur rarely in young age. In this article, we present a young patient with a diagnosis of cholangiocarcinoma.

## CASE PRESENTATION

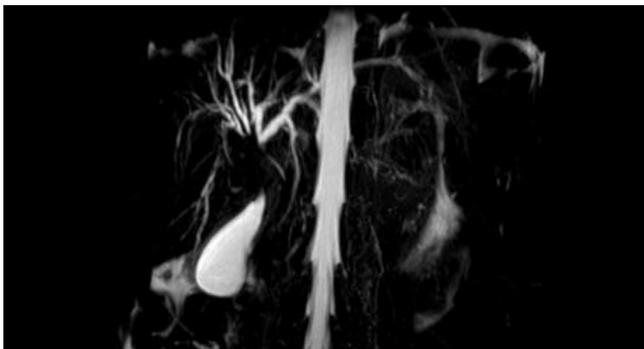
A 34 year old female patient applied to our polyclinic because of nausea, inaccetence, stomach and back pain claims. She has no known additional disease in his background. The patient is not taking any medication. There was no pathologic finding in his physical examination apart from sclera icteric, in the left cervical region lymphadenopathy and hepatomegaly. Blood tests were requested from the patient who had no history of alcohol consumption and any plant use.

Laboratory results; hemoglobin 12,4 g/dl, lökosit: 19.650/mm<sup>3</sup>, platelet 259.000/mm<sup>3</sup>, alanine aminotransferase (ALT): 293 IU/L, aspartate aminotransferase (AST): 214 IU/L, alkalen fosfat (ALP): 698 IU/L, gamma glutamil transferase (GGT): 413 IU/L, total bilirubin: 6,49 mg/dL, direct bilirubin: 4,53 mg/dL, albumin 3,9 g/dL, lactate dehydrogenase (LDH) 344 U/L, sendimentasyon 37 mm/hour, C-reactive protein (CRP) 10 mg/L, was apart from this normal. In the hepatobiliary ultrasonography (USG) was viewed liver segment 5 and 8 level sizes near to 47x48 mm, smoothly limited, lobed contoured, peripheral liver parenchyma according to mild hyperkotic appearance viewing and extensions to the power plant, a solid lesion with arterial flow was observed in the internal echocardiograph. It was also reported that the right branch of the portal vein with the right lobe vascular structures of the liver was secondary compressive view of the lesune. Cancer antigen 19.9 (CA 19.9), carcinoembryonic antigen (CEA) and alpha-fetoprotein (AFP) including tumor markers were within normal limits. Magnetic resonance cholangiopancreatography (MRCP) revealed an incisive lesion extending to the hilar region of approximately 53x34 mm with unclear borders in the liver segment 5-8. There were several satellites lesion adjacent to the lesion neighborhood and there were pathologic intensities surrounding the main vascular structures and the choledo. Prominence in all segments of intrahepatic bile ducts, dilatation of bile ducts in the area near the central section, biliary tract with infiltration and at the confluence level of the main hepatic biliary tract segment has been reported as not monitored (image 1,2) In the foreground, the patient was considered to have cholangiocellular carcinoma. At the same time, the esophagogastroduodenoscopy and colonoscopy examination of the patient was also common. The patient was consulted for histopathological diagnosis in the interventional radiology department. A USG guided tru-cut biopsy was performed in a heterogeneous hypoechoic mass of approximately 5.5 cm in diameter located near the liver right lobe. The pathological report was concluded in favor of CCA with the present morphological and immunohistochemically findings (image 3). A consent form was receipt from the patient.

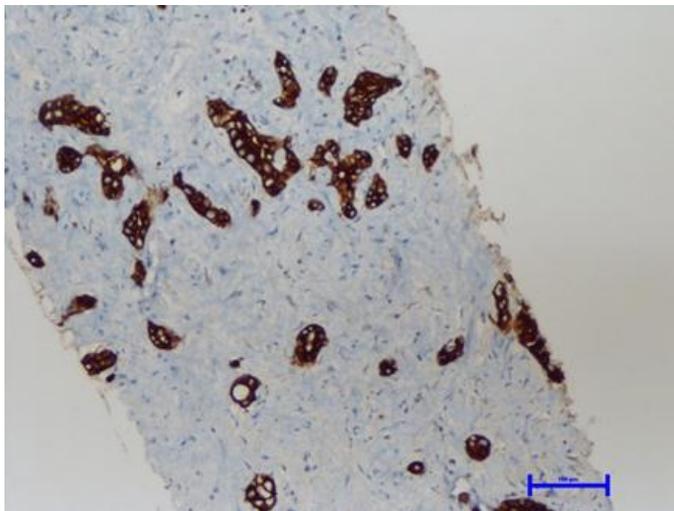
- Uğur Ergün, Balıkesir University Faculty of Medicine, Department of Internal Medicine, Balıkesir, Turkey, [mbalkes10@gmail.com](mailto:mbalkes10@gmail.com)
- Erkan Çağlar, Balıkesir University Faculty of Medicine, Department of Gastroenterology, Balıkesir, Turkey



**Image 1.** In T2 weighted TSE image, there is hyperintense mass with indistinct borders which is located at right lobe of liver. Also hyperintense pathological signal consistent with tumoral infiltration is seen in portal hilar region.



**Image 2.** 3D MIP image of MRCP shows no fluid signal in right and left main hepatic duct, common hepatic duct and common biliary duct, which is thought to be due to tumoral infiltration. All peripheral intrahepatic biliary ducts are dilated due to central obstruction.



**Image 3.** Atypical cells with prominent hyperchromatic nuclei in fibrotic stroma in histopathological sections have glandular neoplastic structures. In immunohistochemical studies, CK7, LMWCK, CK19 CEA positive, ttf-1, CK20, hepatocyte negative, Ki67 50-60% positive.

## CONCLUSION

CCA divided into two groups as intrahepatic and extrahepatic. The extrahepatic CCA is divided into two subgroups as perihilar and distal. Each subgroup has different epidemiologic, pathogenetic and therapy characteristics. Approximately 50% of cases are localized in the bifurcation of the CCA extrahepatic bile duct and are referred to as klatskin tumor. Main symptoms detected in these patients are; acedia, pain, nausea, throwing up, obstructive jaundice and can vary up to sepsis, there is no specific diagnostic laboratory blood test for diagnosis yet (6-8). Symptom findings and laboratory values may vary depending on the localization of the tumor. Diagnosis of CCA, USG, high-resolution/spiral computed tomography (CT) magnetic resonance imaging (MRI), MRCP, endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), endoscopic ultrasonography (EUS), miniprob USG, positron emission tomography (PET), endomicroscopy histology and cytology, are used methods. MRCP, MRI and CT are frequently used in the present to show the tumor in the CCA. PET is useful in demonstrating tumor involvement and metastasis. Aspartate aminotransferase (AST), alanin aminotransferase (ALT), are usually normal, acute obstruction or cholangitis can be found higher. Generally, cholestatic enzymes are high. In our case, the localization of the mass was in the perihilar region and the liver enzymes were elevated due to obstruction in the biliary duct. Detection of tissue, we are using percutaneous fine needle aspiration biopsy, brush, cytological examination of curettage and saffron methods (3). In our case, tru-cut biopsy was performed for the diagnosis of solitary lesion histopathology. The pathology report was interpreted in favor of CCA. In perihilar CCA cases are usually used bismuth-corlette classification. Our case was classified as bismuth type IIIb according to MRCP analysis. It was over thought about the treatment options of the patient diagnosed as perihilar type IIIb CCA. Treatment options varies according to localization. A surgical cure can be provided in a small number of these patients. Palliative procedures such as biliary drainage can be applied if the tumor cannot be resected (3). The patient was diagnosed as perihilar CCA at an early age without any metastatic disease and was referred to a higher center for curative surgical procedure or liver transplantation. As a result, it is an adenomatous malignant disease with a poor prognosis originates from the origin of CCA biliary origin. With age, the incidence is increasing and is usually peaking at the age of 65 years. Treatment varies according to the localization of the mass and the chance of cure is quite low. In our report of case it is important to have a 34 years old female patient diagnosed with CCA. Although the pathological causes underlying CCA are not really known and this disease can rarely occur at a young age. It should be kept in mind that in the early diagnosis of patients with cholestatic enzyme elevation in the hepatobiliary area, patients with solitary lesions may have a diagnosis of CCA.

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