ISSN 2689-8268 | Volume 5

American Journal of Surgery and Clinical Case Reports

Case Report Open Access

Eosinophilic Cholangitis Along with Obstructive Jaundice and Liver Damage: A Case Report and Review of the Literature

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Received: 09 Jun 2022
Accepted: 21 Jun 2022
Published: 27 Jun 2022
J Short Name: AJSCCR

Citation:

Zhang Y, Tang S. Eosinophilic Cholangitis Along with Obstructive Jaundice and Liver Damage: A Case Report and Review of the Literature. Ame J Surg Clin Case Rep. 2022; 5(1): 1-4

Keywords:

Eosinophilic cholangitis; Eosinophil's infiltration; Obstructive jaundice; Bile duct dilatation

Abbreviations:

CT: Computed tomography; EC: Eosinophilic cholangitis; EUS: Endoscopic ultrasonography; MRCP: Magnetic resonance cholangiopancreatography.

1. Abstract

Eosinophilic Cholangitis (EC) is a rare benign self-limiting disease but easily misdiagnosed. Its main features are infiltration of eosinophils in the wall of the biliary tract, extrahepatic bile duct stenosis and obstructive jaundice. We describe the case of a 29-year-old female with EC who initially complained of right epigastralgia, jaundice, nausea and vomiting. We analyzed the clinical presentation, imaging diagnosis results, histopathology and the treatment options for EC (including surgical resection of the bile duct occupying, managed with Roux-en-Y and symptomatic treatment), compared with the few other cases reported in the literature. This case highlights that clinician should be on the alert for the rare case of EC along with obstructive jaundice and liver damage. There is no consensus among experts on its diagnosis and treatment. This case has a certain reference value for the diagnosis and treatment of EC.

2. Introduction

Eosinophilic Cholangitis (EC) is a rare benign self-limiting disease characterized by infiltration of eosinophils in the wall of the biliary tract, sclerosing cholangitis, extrahepatic biliary strictures, and obstructive jaundice [1]. Although the etiology of eosinophilic cholangitis is unclear, underlying illnesses include eosinophilic gastroenteritis, cholelithiasis, hypereosinophilic syndrome, parasitic infection and allergy [2]. It is also occasionally accompanied by several complications sometimes. Because the lesion can affect any section or all of the biliary system, the severity and prognosis are highly variable. Liver damage or fibrosis may occur in EC patients with rapid disease progression, and liver transplantation may be required finally [3]. Here, we present a case of EC with liver damage that was effectively treated by biliary resection and Roux-en-Y surgery, as well as symptomatic therapy.

3. Case Report

A 29-year-old female was admitted to our hospital after suffering from right epigastralgia for 9 days duration and obstructive jaundice with nausea and vomiting for 1-week duration. The patient's vital signs were steady at the time of admission. A physical examination indicated moderate jaundice in the skin and sclera. In addition, the clinical abdominal examination revealed a positive Murphy's sign and right epigastric pain, as well as an old surgical scar in the right lower quadrant that was around 8-cm long (healed with no drainage). She had an appendectomy four years ago for acute appendicitis. She did not have any additional medical history, such as hypertension and diabetes.

Laboratory tests revealed a significant increase in transaminases and biliary enzymes (Table 1, Prior to admission) and abdominal ultrasonography revealed a weak echo in the middle and upper segment of the common bile duct, as well as intrahepatic bile duct expansion, gallbladder narrowing and cystic wall thickening. The patient was transported from a nearby hospital to ours for additional treatment. Re-examination of blood biochemistry and routine revealed significant liver damage and a high level of eosinophilia (Table 1, Prior to surgery). Hepatitis A, B, C and E indicators, as well as autoimmune hepatitis antibodies, were all negative. The structure of porta hepatis was disordered, the wall of the upper seg-

ment of the common bile duct was thickened and narrowed, and the intrahepatic bile duct was dilated, according to an abdominal Computed Tomography (CT) scan (Figure 1). The blockage was found in the upper portion of the common bile duct, and a new organism could not be ruled out. Subsequently, a Magnetic Resonance Cholangiopancreatography (MRCP) was preformed to confirm the abdominal CT diagnosis. MRCP demonstrated aberrant gallbladder morphology as well as biliary dilatation, which was consistent with CT (Figure 2). Furthermore, Endoscopic Ultrasonography (EUS) showed solid space occupied by the submucosa of the common hepatic duct blocked the lumen, suggesting the lesion broke through the outer membrane of the bile duct, infiltrated into the triangle of the gallbladder, and blocked the cystic duct (Figure 3). At present, the diagnosis of obstructive jaundice was clear, but we wondered about the cause of the obstructive jaundice.

According to the above results of MRCP and EUS, the possibility of choledocholithiasis was excluded. We highly suspected that the biliary obstruction was caused by a neoplasm. We timely communicated with patients and their families to explain the patient's condition. According to the opinions of surgical consultation, the patient strongly demanded surgical treatment.

The patient underwent surgical resection of the gallbladder and the occupied bile duct, and was managed with Roux-en-Y meanwhile. During the operation, the size of the confluence of the upper middle common bile duct and cystic duct was about 2.0 × 2.0 cm, which was tough and poorly mobile. Besides, the expansion of the left and right hepatic ducts could be seen above the occupying area. A pathological biopsy showed no evidence of malignancy. In low power magnification, the remaining bile duct was irregular in shape without bile duct epithelial atypia. The subepithelial area displayed periductal fibrosis and a pronounced diffuse inflammatory cellular infiltrate (Figure 4A). In high magnification, a large number of eosinophils infiltrated the gallbladder wall, resulting in the formation of a local abscess, and lymph node hyperplasia with more eosinophils infiltrated the porta hepatis. The pronounced inflammatory cellular infiltrate was comprised almost entirely of eosinophils, identified by their pathognomonic cytoplasmic granules and bilobed nuclei (Figure 4B). Further retrospective review showed peripheral eosinophils (Table 1).

According to the pathological features and the benign clinical course, we diagnosed the patient with eosinophilic cholangitis and obstructive jaundice with liver damage. Postoperatively, epigastralgia and obstructive jaundice were gradually relieved with symptomatic treatment. One week later, the levels of serum bilirubin and transaminase were close to normal and eosinophils in the peripheral blood also decreased significantly (Table 1, Post-surgery). At 16-month follow-up, she remained asymptomatic with normal liver function tests, and eosinophils in her peripheral blood (0.22×109/L) had returned to a normal level.

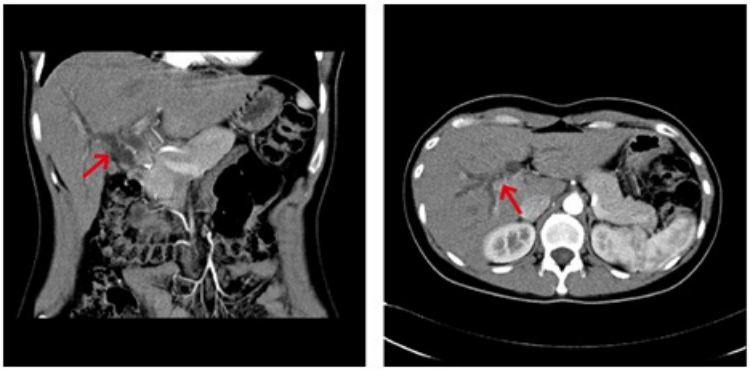

Figure 1: Computed tomography (CT) demonstrating biliary obstruction. The wall of the upper segment of the common bile duct was thickened and narrowed, and the intrahepatic bile duct was dilated (red arrows).

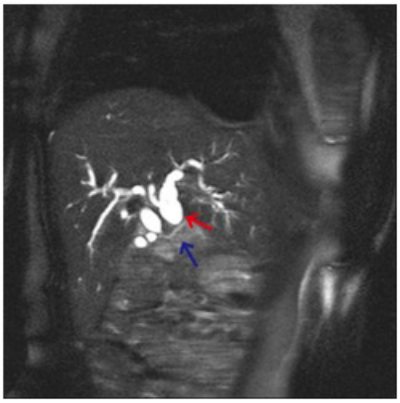

Figure 2: Magnetic resonance cholangiopancreatography (MRCP) exhibited structural disorder in the hepatic portal area with abnormal bile duct obstruction (blue arrow, abnormal occupying; and red arrow, biliary dilatation).

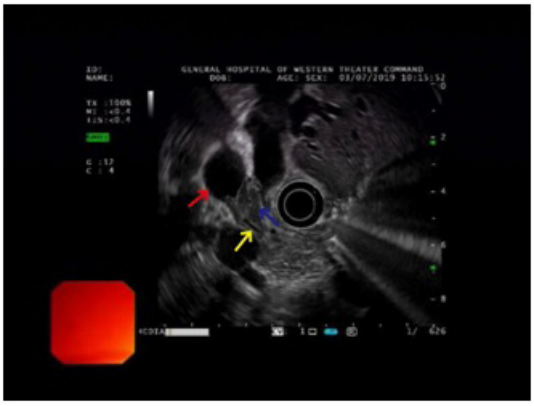

Figure 3: Endoscopic ultrasonography (EUS) revealed dilatation of the common hepatic duct (red arrow), an occupying lesion (blue arrow) and stenosis of the common bile duct (yellow arrow).

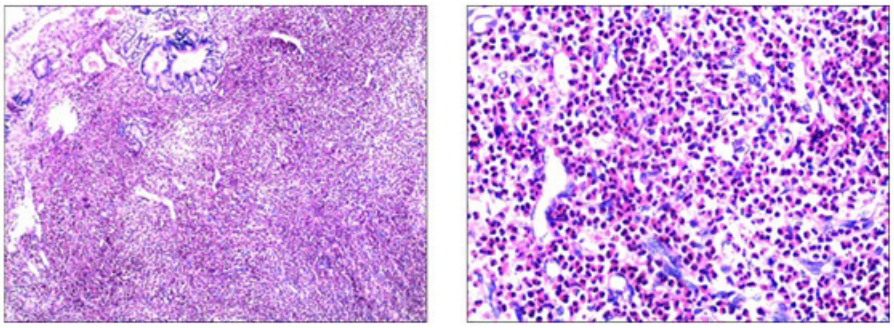

Figure 4: Histopathological examination showing high eosinophilic infiltration of the gallbladder wall with eosinophilic abscess formation (A). Diffuse infiltration of eosinophils in a high magnification pathological section (B).

Table 1: Laboratory values

Testing items	Results			References
	Prior to admission	Pre-surgery	Post-surgery	
ALT (U/L)	583	254.3	68.7	7.0~45.0
AST (U/L)	195.2	127.2	35.6	13.0~40.0
ALP (U/L)	200.5	245.4	81.5	35.0~100.0
GGT(U/L)	460.1	512.2	105.4	7.0~45.0
TBIL (μmol/L)	77.2	102.5	35.1	5.0~28.0
DBIL (μmol/L)	59.4	59	18.6	0~7.0
IBIL (μmol/L)	17.8	43.5	16.5	3.0~23.0
TBA (μmol/L)	259.3	136.1	13.4	0~2.0
Eo# ($\times 10^9$/L)	—	1.1	0.56	0.02~0.52

ALT, Alanine Aminotransfease; AST, Aspartate Transaminase; ALP, Alkaline Phosphatase; GGT, Gamma Glutamyltranspeptidase; TBIL, Total Bilirubin; DBIL, Direct Bilirubin; IBIL, Indirect Bilirubin; TBA, Total Bile Acid; Eo#, absolute eosinophil count.

4. Discussion

EC is a rare disorder of the biliary tract, which was first reported by Leegaard in 1980 [4]. At present, the pathophysiology of EC is still unclear. Most EC patients showed marked peripheral blood eosinophilia, which is associated with several biological conditions, such as allergy, parasitic infection, connective tissue disorder, hematopathy, malignant tumors, and immune deficiency status. The pathogenesis of EC may theoretically involve an atopic mechanism. This mechanism is supported by the increased levels of IgE and IL-5 in EC, and IL-5 can exert antiapoptotic effects on eosinophilia [5]. Recently, some reports have shown leukotriene and IL-16 can mediate eosinophilia recruitment [6]. Therefore, most studies consider that the pathogenesis of EC may be related to an allergy mechanism, but its pathogenesis and pathophysiology still need to be clarified.

In EC, transmural eosinophilic infiltration of the biliary tract may lead to biliary stricture and obstructive jaundice [7]. The diagnosis of EC is still a formidable challenge as it may be confused with cholangiocarcinoma in clinical manifestation and imaging features. It is very important to distinguish benign and malignant biliary stricture because of the complexity of differential diagnosis. Previous studies have shown that the majority of strictures are secondary to malignancy (pancreatic adenocarcinoma and cholangiocarcinoma), with up to about 30% due to benign pathologies. Tumor markers are useful in distinguishing between benign and malignant biliary obstruction but are usually unable to determine the exact cause of a biliary stricture [8]. Here, we tested the tumor markers in the peripheral blood of the patient and they were all

within the normal range.

Prior to making the diagnosis of EC, other benign pathologies of biliary strictures should be ruled out, such as inflammatory stenosis secondary to common bile duct stones, immune-mediated inflammation (primary sclerosing cholangitis, IgG4-related sclerosing cholangitis, mast cell cholangitis), infection (parasitic, recurrent pyogenic cholangitis, tuberculosis), radiation therapy and cystic fibrosis [4]. The laboratory values of the patient showed normal autoimmune markers (myeloperoxidase antibody, anti-smooth muscle antibody and anti-mitochondrial antibody), IgG4 levels and no sign of infection. Although the presence of peripheral eosinophilia may be a clue to the diagnosis of EC, the sensitivity or specificity is variable. With the advent of ERCP, EUS and MRCP, accuracy in diagnosing the cause of biliary stricture has been improved, but the definitive diagnosis needs to be confirmed by pathological analysis of the bile duct lesions.

Therefore, it is often necessary to carry out a surgical operation to take pathological tissue biopsy for a clear diagnosis. However, this kind of invasive examination greatly increases the patients' safety risks. At present, no clear diagnosis standard of EC has been established. Matsumoto proposed a triad diagnosis standard, including wall thickening or stenosis of the biliary system, elevated eosinophil infiltration in the pathological tissue, and reversibility of biliary abnormalities following steroid treatment or without treatment [9].

Currently, the treatment of EC is mainly based on case report experiences and no expert consensus. As EC is a benign disease, some scholars believe a trial of corticosteroids should be considered before invasive treatment. Recently, a case report showed an EC patient got better with biochemical analysis of her eosinophilia and liver function test by oral budesonide treatment. Although a diagnostic test for glucocorticoids can be attempted, surgical resection of the extrahepatic bile duct and Roux-en-Y may be required as histopathological evidence before EC is often impossible [10].

5. Conclusion

In our case report, the patient was treated with a surgical resection of the gallbladder and the occupying extrahepatic bile duct along with a Roux-en-Y hepaticojejunostomy. Although the pathogenesis of EC is still unclear, it is suggested that etiology treatment and biliary obstruction relief are the main treatment guidelines through a retrospective study of cases.

6. Funding

This work was supported by grants from the National Natural Science Foundation of China (82000530) and the Basic Medical Research Projects of the General Hospital of Western Theater Command, China (2021-XZYG-B24).

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