Research Article

Integrative Gastroenterology and Hepatology

Is Budd-Chiari Syndrome Associated to Alcoholic Related Liver Cirrhosis

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Abstract

Introduction and aim: The Budd-Chiari Syndrome (BCS) is redefined as hepatic vein outflow tract obstruction with a very low incidence. We aim to analyze the etiology and clinical character of BCS in Hebei area of North China.

Material and methods: The diagnosis of BCS and alcoholic related liver cirrhosis (Alcohol-LC) are according to the guidelines of American Association for the Study of Liver Diseases (AASLD), while the diagnosis of hepatitis B virus related liver cirrhosis (HBV-LC) is according to the guidelines of European Association for the Study of the Liver (EASL). BCS patients including inferior vena cava block (IVC), hepatic vein block (HV) and inferior vena cava combining with hepatic vein block (IVC/HV) are involved in this analysis.

Results: The subtype's distributions of this disease are more frequent for IVC patients compared with HV and IVC/HV patients. The subsequent analysis shows that the incidence of BCS is more predisposed to Alcohol-LC than HBV-LC ($p < 0.001$).

Conclusion: BCS seem to be associated with Alcohol-LC compared with that of HBV-LC.

Keywords: Diagnosis, Types, Clinical features, Etiology, Incidence

Introduction

Budd-Chiari syndrome (BCS) was redefined as obstruction of hepatic vein outflow tract. Three cases of symptomatic hepatic vein occlusion were first reported by Budd in 1845. Since then, there have been more hepatic vein occlusions with inferior vena cava lesions [1-3].

BCS is a rare disease in Western countries; however, the incidence is relatively high in some Asian regions [4]. According to the location of obstruction, the disease is usually divided into three types: hepatic veins block

(HV type), inferior vena cava block (IVC type), and IVC/HV combination block (IVC/HV type) [2,5]. In Western countries, HV block is a more common subtype with clear etiology such as oral contraceptives, pregnancy, myeloproliferative disorders (MPD), paroxysmal nocturnal haemoglobinuria (PNH), and Behcet diseases (BD) [3], while IVC or IVC/HV combined block has almost no hypercoagulability of blood, which seems to be more common in Asia, and the cause of the disease is interpreted as congenital abnormal development, thrombosis and inflammation [6-9].

Clinical manifestations of BCS include abdominal pain, splenomegaly, hepatomegaly, ascites, gastrointestinal bleeding, and abdominal wall varicosis. The diagnosis of BCS relies primarily on imaging examinations, including Color Doppler Ultrasound (CDUS), Computed Tomography (CT), Magnetic Resonance Imaging (MRI) and digital subtraction angiography (DSA) [10]. Currently, the treatment of BCS mainly includes interventional therapy and surgical treatment, but simple drug therapy is less used [11-14]. We retrospectively investigated the clinical features and etiology of BCS in the Hebei area of North China to find the risk factors for the disease.

Materials and Methods

Patients

Thirty two patients from the Hebei area were diagnosed with BCS at the Fourth Hospital of Hebei Medical University according to the guidelines of American Association for the Study of Liver Diseases (AASLD) from January 2005 to May 2017 were retrospectively analyzed [15]. The clinical data analysis included general characteristics, signs and symptoms, laboratory tests, imaging characteristics, accompanying diseases and treatment method. The diagnosis of alcohol related liver cirrhosis (Alcohol-LC) was based on a history of significant alcohol intake, clinical evidence of cirrhosis of the liver and supporting laboratory abnormalities, elimination of other liver disease [16]. The diagnosis of hepatitis B virus related liver cirrhosis (HBV-LC) was based on histological or clinical evidence of cirrhosis and etiology of HBV infection [17]. The analysis of the patient's data was approved by The Ethical Committee of the hospital.

Statistical analysis

The chi-square test and Fisher exact method were used for categorical comparison with the SPSS 21.0 statistical software package (SPSS Company, Chicago, IL), and $P < 0.05$ was considered as the statistically significant threshold.

Results

Clinical features of BCS patients in the Hebei area

A total of 32 patients including 14 males (43.75%) and 18 females (56.25%) with the average age of 45.64 ± 18.23 years old in males and 45.33 ± 16.66 in females were involved in the investigation. There was no statistical difference in the distribution frequency of age and gender (data not shown). As shown in figure 1A, the incidence of

IVC type (74%, 24/32) was higher than that of HV type (6%, 1/32) and IVC/HV type (21%, 7/32). The chi-test was used to compare the distribution frequency of clinical features such as abdominal pain, splenomegaly, hepatomegaly, ascites and gastrointestinal bleeding in these three subtypes. No statistical difference was found (Figure 1B). Laboratory data for transaminase, total bilirubin, prothrombin time (PT) and activation of partial thrombin time (APTT) was also compared in these three subtypes, without statistical difference (Figure 1C).

Figure 1: The distribution of BCS subtypes and their association with clinical data. A: The distribution of BCS subtypes for IVC, HV and IVC/HV; B: The distribution frequency for clinical characteristics among three BCS subtypes; C: The laboratory data including transaminase, total bilirubin, PT and APTT was also compared among these three BCS subtypes.

Association of Alcohol-LC with BCS in the Hebei area

Among 32 BCS patients, 26 patients combined with liver cirrhosis. As shown in figure 2, one case was accompanied with HBV-LC whereas five cases accompanied with Alcohol-LC. Since HBV infection is the main cause for liver diseases in China, we investigated whether BCS is associated with Alcohol-LC. There are 610 resident patients of HBV-LC and 137 resident Alcohol-LC patients from January 2005 to May 2017 in our hospital. As shown in table 1, the incidence of BCS was more predisposed to Alcohol-LC than HBV-LC ($p < 0.001$).

Figure 2: The accompanying liver disease for BCS.

Table 1: The resident patients of HBV-LC and Alcohol-LC from January 2005 to May 2017 in the Fourth Hospital of Hebei Medical University.

	Accompanying BCS, n	Without BCS, n	P-value
Patients			0.00
HBV-LC	1	609	
Alcohol-LC	5	132	
BCS: Budd-Chiari syndrome.			
HBV-LC: hepatitis B virus related liver cirrhosis.			
Alcohol-LC: alcoholic related liver cirrhosis.			

Discussion

We found that IVC type was the predominant type in Hebei area of North China, which was different with the previous reports that the incidence of IVC/HV subtype was higher in China [18,19]. These findings indicate that the predominant types of BCS may have regional differences in China based on genetic background and environmental factors.

We collected 32 BCS patients including one patient accompanied with HBV-LC whereas 5 patients accompanied with Alcohol-LC. The accompanied liver diseases of the Alcohol-LC seem to occupy higher proportion than that of HBV-LC by our first analysis. Since China belongs to the highest HBV endemic areas, we compared the distribution frequency of accompanied liver diseases of BCS and found that the incidence of BCS

was more associated with Alcohol-LC compared with that of HBV-LC patients. Drinking has been reported to exhibit both favorable and unfavorable effects on metabolism, lipid profile, blood coagulation and fibrinolysis, blood pressure and vascular tone depending on the amount of alcohol consumed and the way of drunk [20]. Long-term alcohol administration could induce pathological changes of basilar arteries in brain tissue including endothelial exfoliation, inner elastic lamina fragmentation and thickening of tunica media and adventitia, which might be responsible for cerebral embolism, thrombosis and ischaemia with coagulation cascade and contractility of the cerebral vessels involved in the process [21,22]. In addition, acute ingestion of large quantities of alcohol is known to be able to trigger acute myocardial infarction [23]. The underlying mechanisms of alcohol induced artery embolism have yet to be demonstrated, through coronary vasospasm, decreased fibrinolysis and an enhanced prothrombotic state was supposed. As for vein, a prospective study showed that alcohol could increase blood pressure in the portal vein so as to inducing portal vein regurgitating and blood clots [24]. Portal venous reflux is a special feature of alcoholic cirrhosis and is rarely found in viral cirrhosis [25]. In addition, chronic alcohol ingestion activity NADPH oxidase activity to induce vascular wall damage, followed by penetration of liquid and macromolecules to trigger production of collagen, elastin, and basement membrane-like substances [26,27]. All of the above mechanisms may contribute to alcohol induced BCS. Our data in one center with small sample size indicated that BCS was more predisposed to Alcohol-LC than HBV-LC, but the finding need to be validated in more sample size from different areas.

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Consent

All procedures were supervised and approved by the hospital's Ethic Committee.

Conflicts of Interest

All authors declare that they have no conflicts of interest.

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