## MINIREVIEWS

<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
</tr>
</thead>
</table>
| 2696 | Standardization of critical care management of non-critically ill patients with COVID-19  
| 2703 | Mediastinal lymphadenopathy in COVID-19: A review of literature  
Taweesedt PT, Surani S |
| 2711 | Polycystic ovary syndrome: Pathways and mechanisms for possible increased susceptibility to COVID-19  
Ilias I, Goulas S, Zabulienne L |

## ORIGINAL ARTICLE

### Clinical and Translational Research

<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
</tr>
</thead>
</table>
| 2721 | Circulating tumor cells with epithelial-mesenchymal transition markers as potential biomarkers for the diagnosis of lung cancer  
Jiang SS, Mao CG, Feng YG, Jiang B, Tao SL, Tan QY, Deng B |

### Retrospective Study

<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
</tr>
</thead>
</table>
| 2731 | Management and implementation strategies of pre-screening triage in children during coronavirus disease 2019 pandemic in Guangzhou, China  
| 2739 | Clinicopathological features of superficial CD34-positive fibroblastic tumor  
Ding L, Xu WJ, Tao XY, Zhang L, Cai ZG |
| 2751 | Application of a rapid exchange extension catheter technique in type B2/C nonocclusive coronary intervention via a transradial approach  
Wang HC, Lu W, Gao ZH, Xie YN, Hao J, Liu JM |

## SYSTEMATIC REVIEWS

<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
</tr>
</thead>
</table>
| 2763 | Paradoxical relationship between proton pump inhibitors and COVID-19: A systematic review and meta-analysis  
Zippi M, Fiorino S, Budriesi R, Micucci M, Corazza I, Pica R, de Biase D, Gallo CG, Hong W |

## META-ANALYSIS

<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
</tr>
</thead>
</table>
| 2778 | Predictive risk factors for recollapse of cemented vertebrae after percutaneous vertebroplasty: A meta-analysis  
<table>
<thead>
<tr>
<th>Contents</th>
</tr>
</thead>
<tbody>
<tr>
<td>CASE REPORT</td>
</tr>
</tbody>
</table>
| 2791 | Malignant pheochromocytoma with cerebral and skull metastasis: A case report and literature review  
Chen JC, Zhuang DZ, Luo C, Chen WQ |
| 2801 | Unresectable esophageal cancer treated with multiple chemotherapies in combination with chemoradiotherapy: A case report  
| 2811 | Role of positron emission tomography in primary carcinoma ex pleomorphic adenoma of the bronchus: A case report  
Yang CH, Liu NT, Huang TW |
| 2816 | Positive reverse transcription-polymerase chain reaction assay results in patients recovered from COVID-19: Report of two cases  
Huang KX, He C, Yang YL, Huang D, Jiang ZX, Li BG, Liu H |
| 2823 | Laryngeal myxoma: A case report  
| 2830 | Prostate stromal tumor with prostatic cysts after transurethral resection of the prostate: A case report  
| 2838 | Intramuscular hematoma in rhabdomyolysis patients treated with low-molecular-weight heparin: Report of two cases  
Yuan SY, Xie KF, Yang J |
| 2845 | Partial response to Chinese patent medicine Kangliu pill for adult glioblastoma: A case report and review of the literature  
| 2854 | Behcet’s disease manifesting as esophageal variceal bleeding: A case report  
Xie WX, Jiang HT, Shi GQ, Yang LN, Wang H |
| 2862 | Successful endoscopic surgery for emphysematous pyelonephritis in a non-diabetic patient with autosomal dominant polycystic kidney disease: A case report  
Jiang Y, Lo R, Lu ZQ, Cheng XB, Xiong L, Luo BF |
| 2868 | Robotic assisted removal of pelvic splenosis fifty-six years after splenectomy: A case report  
Tognarelli A, Faggioni L, Erba AP, Faviana P, Durante J, Manassero F, Selli C |
| 2874 | Pulmonary alveolar proteinosis complicated with nocardiosis: A case report and review of the literature  
Wu XK, Lin Q |
| 2884 | Detection of EGFR-SEPT14 fusion in cell-free DNA of a patient with advanced gastric cancer: A case report  
Kim B, Kim Y, Park I, Cho JY, Lee KA |
<table>
<thead>
<tr>
<th>Page</th>
<th>Title</th>
<th>Authors</th>
</tr>
</thead>
<tbody>
<tr>
<td>2899</td>
<td>Torsades de pointes episode in a woman with high-grade fever and inflammatory activation: A case report</td>
<td>Qiu H, Li HW, Zhang SH, Zhou XG, Li WP</td>
</tr>
<tr>
<td>2916</td>
<td>Allogeneic hematopoietic stem cell transplantation in a 3-year-old boy with congenital pyruvate kinase deficiency: A case report</td>
<td>Ma ZY, Yang X</td>
</tr>
<tr>
<td>2930</td>
<td>Sclerosing polycystic adenosis of the submandibular gland: Two case reports</td>
<td>Wu L, Wang Y, Hu CY, Huang CM</td>
</tr>
<tr>
<td>2937</td>
<td>Budd-Chiari syndrome associated with liver cirrhosis: A case report</td>
<td>Ye QB, Huang QF, Luo YC, Wen YL, Chen ZK, Wei AL</td>
</tr>
</tbody>
</table>
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Budd-Chiari syndrome associated with liver cirrhosis: A case report

Qiao-Bo Ye, Qin-Feng Huang, Yao-Chang Luo, Yi-Lei Wen, Zi-Kun Chen, Ai-Ling Wei

Abstract

BACKGROUND
Budd-Chiari syndrome (BCS) is a rare heterogeneous liver disease characterized by obstruction of the hepatic venous outflow tract. The incidence of BCS is so low that it is difficult to detect in general practice and difficult to include within the scope of routine diagnosis. The clinical manifestations of BCS are not specific; hence, BCS tends to be misdiagnosed.

CASE SUMMARY
We report the case of a 33-year-old Chinese woman who presented with progressive distension in the upper abdomen. She was initially misdiagnosed with liver cirrhosis (LC) due to abnormalities on an upper abdominal computed tomography scan. Although she was taking standard anti-cirrhosis therapy, her symptoms did not improve. Magnetic resonance imaging showed caudate lobe hypertrophy; and dilated lumbar and hemiazygos veins. Venography revealed membranous obstruction of the inferior vena cava owing to congenital vascular malformation. A definitive diagnosis of BCS was made. Balloon angioplasty was performed to recanalize the obstructed inferior vena cava and the patient’s
CONCLUSION
BCS lacks specific clinical features and can eventually lead to LC. Clinicians and radiologists must carefully differentiate BCS from LC. Correct diagnosis and timely treatment are vital to the patient's health.

Key Words: Budd-Chiari syndrome; Liver cirrhosis; Venography; Balloon angioplasty; Inferior vena cava; Case report

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INTRODUCTION
Budd-Chiari syndrome (BCS) is a rare heterogeneous liver disease characterized by obstruction of the hepatic venous outflow tract, which may occur anywhere from the small hepatic veins (HVs) to the inferior vena cava (IVC) and right atrium[1]. According to the location of the obstruction, BCS can be classified as involving the small HVs, large HVs, IVC, or arbitrary combinations of these[2]. BCS can eventually lead to sinusoidal congestion, portal hypertension, liver cell injury, centrilobular fibrosis, and ultimately, cirrhosis[3]. The incidence rate of BCS varies among countries. In China, the annual incidence rate of BCS in the five areas with the highest prevalence is estimated to be 0.88 per million[4]. The rate of liver cirrhosis (LC) caused by BCS is approximately 4%[5]. Due to its low incidence, BCS is difficult to detect in general practice and to include within the scope of routine diagnosis. Moreover, the clinical manifestations of BCS are abdominal pain, hepatomegaly, and ascites, which are also frequently seen in LC; thus, BCS tends to be misdiagnosed.

Here, we report a case of BCS associated with cirrhosis, which was initially misdiagnosed.

CASE PRESENTATION

Chief complaints
A 33-year-old Chinese woman was admitted to our medical institution on May 21, 2018, owing to progressive distension in the upper abdomen.

History of present illness
Two weeks before admission, she was diagnosed with LC, portal hypertension and splenomegaly, based on an upper abdominal computed tomography (CT) scan at another hospital. Although she was taking prescribed medication that exerted effects such as anti-hepatic fibrosis, inhibition of gastric acid secretion, and protection of the stomach, her symptoms did not improve. She developed progressive distension in the upper abdomen with sour regurgitation. There was no nausea, vomiting, diarrhea, or symptoms were completely resolved.
abdominal pain.

**History of past illness**
The patient had a history of thrombocytopenia going back more than 10 years and she had undergone surgery for an ovarian cyst on the left side in 2011.

**Personal and family history**
No special personal and family history.

**Physical examination**
Physical examination revealed dark discoloration and mild tenderness in the left lower abdomen; other examinations were normal.

**Laboratory examinations**
Complete blood cell count showed a reduced white blood cell count 3.1 × 10^9/L (normal range 3.5-9.5 × 10^9/L) and platelet count 74 × 10^9/L (normal range 125-350 × 10^9/L). Liver and renal functions, coagulation, and tumor markers were normal. Serum electrolytes were within the normal range. The levels of protein C, protein S, immunoglobulin (Ig) G, IgA, and IgM were also within normal limits. Serology for hepatitis B surface antigen, hepatitis C antibody, anticardiolipin antibodies, and lupus anticoagulant was negative. No other obvious abnormalities were discovered.

**Imaging examinations**
Gastroscopy showed mild esophageal varices. Magnetic resonance imaging (MRI) revealed caudate lobe hypertrophy, cirrhosis, and dilated lumbar and hemiazygos veins. Dilated azygos veins and narrowed IVC were present. Hypersplenotrophy and dilated veins in the lower esophagus and surrounding the hilus lienalis were also observed.

**Pathological examination**
To confirm the diagnosis of BCS, liver biopsy was performed under CT guidance. Histochemical staining (hematoxylin-eosin and Masson trichrome) showed hepatocyte degeneration, bridging fibrosis, sinusoidal dilatation, and areas of fibrous tissue with substantial hyperplasia.

**FINAL DIAGNOSIS**
The definitive diagnosis in this patient was BCS, compensated LC, and thrombopenia.

**TREATMENT**
Venography of the HVs and IVC via the femoral vein and internal jugular vein revealed complete occlusion of the IVC with the formation of numerous collateral branches. Balloon angioplasty was performed to recanalize the obstructed IVC on June 19, 2018. The administration of anti-hepatic fibrosis medication was continued. Three months after balloon angioplasty, the patient again presented with upper abdominal distension and pain and was readmitted to our hospital. MRI showed HV stenosis with ectopic tissue. The obstructed IVC was treated with balloon dilation angioplasty on October 23, 2018.

**OUTCOME AND FOLLOW-UP**
Six months after balloon angioplasty, the patient’s MRI revealed that dilation of the lumbar and hemiazygos veins and caudate lobe hypertrophy were improved. In addition, venography of the IVC combined with hepatic venous pressure measurement was performed. The results showed that the pressure was 12 mmHg. Furthermore, liver function tests were normal.
**DISCUSSION**

BCS is a relatively rare vascular disorder among liver diseases. It is characterized by obstruction of the hepatic venous outflow tract and may lead to congestion in the liver. Chronic hepatic congestion can eventually result in liver fibrosis, cirrhosis, and hepatocellular carcinoma. BCS is divided into four classifications involving small HVs, large HVs, the IVC, and arbitrary combinations of these according to the venous stenosis site. The distribution of BCS varies geographically. IVC with or without HV obstruction is predominant in Asia, whereas HV obstruction predominates in Western countries\[6]. In terms of age and sex, BCS tends to be more common in females than males and to mostly appear at the age of 20-39 years\[7].

A number of causes of BCS have been identified. According to etiology, BCS can be divided into primary BCS and secondary BCS. Risk factors for the former include...
hypercoagulable states and thrombosis, which may be caused by myeloproliferative disorders, antiphospholipid syndrome, oral contraceptive use, pregnancy, hyperhomocysteinemia, Behçet disease, protein C deficiency, and protein S deficiency, among others. Risk factors for secondary BCS are predominantly tumoral invasion, abscesses, and cysts[9]. Vascular thrombosis is the most common element resulting in obstruction of the HV system, as reported in a long-term follow-up study[8]. However, more potential risk factors or causes of BCS should be investigated clinically.

Classical clinical manifestations of BCS include abdominal pain, ascites, hepatomegaly, jaundice, and leg swelling[10,11]. Up to 20% of patients with BCS are asymptomatic[12]. In our report, the main complaint in this patient was upper abdominal distension and pain. Nevertheless, this finding was not specific; these symptoms or other classical clinical manifestations may be observed in progressive liver diseases. Hence, in addition to symptoms and signs, diagnostic techniques and the skill of the physician are important in the diagnosis of BCS.

There are various auxiliary examinations for diagnosing BCS. In some cases, serum aminotransferases and bilirubin can be obviously increased, and albumin is decreased. However, laboratory findings may also show that liver function or other indicators are normal. Under these circumstances, radiological methods are helpful for further diagnosis. Doppler ultrasound, CT, and MRI are the main diagnostic methods. Although the sensitivity of ultrasound is as high as 87%/13, BCS can be excluded relatively easily using ultrasound in comparison with CT/MRI, which can reveal abnormal changes in vessels of the liver. CT is limited owing to its uncertain results in nearly 50% of cases[14]. In fact, most patients with BCS can be diagnosed if the radiologist and clinician carefully examine the imaging features of BCS. MRI along with intravenous gadolinium injection has advantages in the visualization of HVs, IVC, large intrahepatic or comma-shaped collateral vessels, and spider web networks. Thus, MRI is a reasonable choice for the diagnosis of BCS.

Liver biopsy is not a routine diagnostic requirement. However, when imaging does not show the obstruction of venous outflow or BCS is suspected, liver biopsy is required. Liver biopsy may demonstrate liver cell loss, congestion, and fibrosis. In addition, liver biopsy is helpful for differentiating between BCS and veno-occlusive disease. It should be noted that congestion can be found in constrictive pericarditis and cardiac failure, and fibrosis can indicate other diseases, such as diabetes. These similar clinical characteristics should be identified.

Venography can be considered as a diagnostic procedure when the diagnosis of BCS remains unclear. At the same time, valuable information, including the degree of thrombosis, assessment of HVs, and caval pressures, can be provided by venography, to help in the choice of optimal treatment. The portal vein can also be evaluated in this procedure. Venous pressure measurements are conducive to treatment options under various circumstances. For example, to relieve cirrhosis, dilation of HV stenosis is performed to reduce pressure of the HV. Transjugular intrahepatic portosystemic shunt (TIPS) is performed to decrease pressure of the portal vein, which is conducive to the regression of ascites and can effectively prevent bleeding via embolization of varicose collateral vessels in the same sitting. Furthermore, venous pressure measurements are helpful for assessing the patient’s condition in postoperative follow-up.

BCS has a standard management and appraisal system based on guidelines. Specifically, the first-line therapy is medical treatment, which is administered to patients with a hypercoagulable state, portal hypertension, basic diseases, or related complications. The second-line therapy is angioplasty or stenting, which is appropriate for patients with short-length stenosis after medical therapy failure. The next step in management is TIPS, which is suitable for patients who do not respond to medical, angioplasty, or stenting treatment. Liver transplant is the last step in management and has strict requirements, being suitable only for patients with LC, advanced liver dysfunction, or fulminant hepatic failure[15]. It has been reported that the long-term survival rate of liver transplant is approximately 84%/16.

In our patient, the causes and classical clinical features of BCS were not recognized. BCS was diagnosed by MRI findings, which showed a narrowed IVC, caudate lobe hypertrophy, and dilated lumbar vein,azygos veins, and hemiazygos veins. Cavography confirmed IVC obstruction in this patient. Short-length stenosis is found in approximately 60% of patients with obstructive IVC[17]. In theory, patients with segmental or focal block of the HV outflow tract are suitable for recanalization. Recanalization of the IVC with balloon angioplasty can improve the clinical presentation. The short-term effect is satisfactory, but the restenosis rate is as high as 50% at 2 years after angioplasty[18]. Therapy using stents could prolong the long-term patency rates[19]. Stent implantation should be considered for patients with an
unsatisfactory curative effect or recurrent restenosis. However, the disadvantage of stent treatment should not be underestimated. The clinician should choose the best therapy according to the patient’s condition.

The ultimate therapeutic purpose of various clinical treatment methods is to remove the diseased HV/IVC; restore normal blood flow; relieve portal hypertension, stasis cirrhosis, or IVC hypertension syndrome; improve the quality of life of patients; and prolong survival. Balloon angioplasty is one of the procedures used to recanalize the obstruction of the hepatic venous outflow tract. However, the occurrence of postoperative restenosis has serious effects on the long-term curative effects. In our report, with regard to the possible cause of the first three-month re-stenosis, we concluded that the following causes were possible. First, the diameter of the balloon catheter used in balloon angioplasty was not optimal, and the medical community has not yet set a standard for the size of the balloon in balloon angioplasty. Second, inductive vascular repair after balloon angioplasty resulted in thickening of the intima and proliferation of vascular smooth muscle cells in the media. To prevent recurrence, we should maximize treatment efficacy. Specifically, selection of the balloon diameter is key. It affects the clinical efficacy and the recurrence of postoperative lesions. Selection of the appropriate balloon diameter should be fully evaluated in preoperative planning. During the operation, the surgeon should fully dilate the stenosis or occlusion of the IVC, tear the septum, and loosen the thickened venous wall and extravascular fibrous connective tissue to improve the efficacy and reduce the postoperative recurrence rate.

One point that should be noted is that LC was secondary to BCS in this case, as the common causes of cirrhosis had been ruled out. Owing to its rarity as a cause of LC, a diagnosis of BCS may be overlooked by inexperienced clinicians. A detailed medical history, clinical presentations, physical examination, and imaging findings or liver biopsy should be used to make a diagnosis. Moreover, BCS should be differentiated from other similar diseases, such as congestive hepatopathy. Correct diagnosis will avoid missing the optimal therapy time and prolonging the condition. Theoretically, chronic liver congestion in patients with BCS is the main factor leading to the development of LC. Greater attention is needed for the timely treatment of BCS before its progression to LC. 

CONCLUSION

We describe a case of BCS, in which the obstruction site was the IVC. BCS is relatively easy to misdiagnose or overlook owing to its rarity. Clinicians and radiologists must carefully differentiate BCS from LC. Particularly when patients with LC present with unknown causes and normal liver function, the possibility of BCS should be considered. Correct diagnosis and timely treatment can affect the therapeutic efficacy. Furthermore, recanalization before the formation of hepatocirrhosis is vital to prevent or reverse hepatic fibrosis.

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REFERENCES


