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CASE REPORT

A case report of hepatic veno-occlusive disease after ingesting dainties

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Abstract

Hepatic veno-occlusive disease (HVOD) is rarely encountered and easily misjudged as Budd-Chiari syndrome. It is often related to stem cell transplantation in recent years. We report a case of HVOD that is related to ingestion of some palatable local dishes. The diagnosis was confirmed by liver biopsy pathology with specific observation of inflammatory changes and fibrosis of venules intima, dilated sinusoids and central veins. Chronic diarrhea is unique for this case as a result of ingesting harmful stuffs. This case demonstrated that supervision and instruction of food recipe and traditional medicine are crucial, and prompt diagnosis, supportive care and specific treatment are essential to decreasing the morbidity and mortality of HVOD.

Key words: Hepatic veno-occlusive disease; Diagnosis; Management; Biopsy

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INTRODUCTION

Hepatic veno-occlusive disease (HVOD) is a rarely encountered ailment in the literature[1,2]. The establishment of the diagnosis of this condition can be very difficult because there is no specificity in the clinical manifestations and some common findings are similar to Budd-Chiari syndrome (BCS), a peculiar condition with many pitfalls leading to misdiagnosis[3]. Until now, the mechanism of etiology has not been clearly identified and only considered by some authors[2,4] correlated to the non-thrombotic occlusion of the central veins of hepatic lobules, certain inflammatory factors, detoxification of the liver, *etc.* This is a case of HVOD that is related to ingestion of some palatable local dishes.

CASE REPORT

A 17-year-old female was admitted with a complaint of escalating abdominal pain, increased abdominal circumference and recurrent diarrhea. The symptoms started 2 mo ago, mild at first, but now became more and more obvious and bothersome. Physical examination revealed mild jaundice in sclera, no skin petechia, no edema, no superficial varices, and no positive findings in the heart and lungs. The abdomen was distended, no tenderness or rebound pain. The liver and spleen were not palpable. The sign of ascites was positive. Laboratory results showed WBC 6.82 × 10^9/L, Hb 122 g/L, BPC 81.30 × 10^9/L, HbsAg (-), ALT 66 U/L, AST 81 U/L, serum total bilirubin 39.7 μmol/L, direct bilirubin 10.6 μmol/L, and ALB 32.8 g/L. Abdominal paracentesis aspirated clear ascites, with a total cell count of 6400, WBC 5/mL, and ALB 15.3 g/L. CT scan of the abdomen revealed moderate ascites, hepatomegaly, with areas of inhomogeneously reduced density, delayed enhancement of parenchyma, and obscure hepatic veins.

Considering the symptom of repeated diarrhea, the physicians inquired time and again that if she had eaten anything harmful, but nothing was determined. At last, the details of the patient's food consumption found that she had enjoyed a "cuisine recipe for longevity" for a dozen of times. The dish was delicious, the patient admitted, and all the local people thought that it is one of the valuable dainties. The content of the ingredients proved to consist of the seeds of a plant called "wild sesame" by the local people, whereas actually the entity of it is not barbate deadnettle but rattlebox[5], *Crotalaria sessiliflora.*

Venography by catheterization revealed patent inferior vena cava (IVC) and main hepatic veins (HV). Finally, a percutaneous puncture of liver biopsy was completed with the pathological findings of inflammatory changes and fibrosis of venules intima, dilated sinusoids and central veins, and no obvious degeneration of hepatocytes was found. Then the diagnosis of HVOD was established. The patient was given supportive and hepatoprotective treatment. Her symptoms were relieved and she was completely recovered within two weeks.

DISCUSSION

In recent years, HVOD often occurs secondary to stem cell transplantation as the most common regimen-related toxicity[6]. However, herbs or plants are occasionally reported to cause HVOD, especially those used in traditional medicine[7].

The mechanism of liver damage caused by plants has not been elucidated, only a hypothesis of "conflict of plant versus animal or plant-animal interaction" seems more acceptable[8]. The defence system of many plants are used to produce compounds such as alkaloids and polypeptides against the animals that ingest them. Such animals are also self-protected by efflux transporters in the gut and detoxification of the liver, herbivore countermechanism to plant chemical defenses, and multidrug resistance-associated protein isoform, *etc.*

Senecio plants[7,9] as well as *Crotalaria* and *gynura segetum* are reported potentially hepatoxic if consumed over a period of weeks. Poisoning can occur through ingestion of especially the seeds, but also leaves and stems. After ingestion of the plants, the major toxic components pyrrolizidine alkaloids (PAs) are absorbed and converted to highly reactive alkylating pyrroles that cause hepatocellular necrosis, biliary hyperplasia, fibrosis, and hepatocytomegaly.

The clinical manifestations of HVOD fall into the categories of mild, moderate and severe according to its final outcome[4,6,10] or acute, subacute and chronic according to its onset and course. Typical findings include abdominal pain, ascites with elevated ALB, jaundice and hepatomegaly. Chronic diarrhea is unique for this case as a result of ingesting harmful stuffs. Imaging diagnoses including gray-scale US, Doppler US, CT and MRI have been reported as convenient and useful. Venography often reveals patent IVC and main outflow of HV.

Pathology of liver biopsy definitely establishes the diagnosis of HVOD, with the hallmark of fibrous obliteration of terminal hepatic venules and small lobular veins. Both percutaneous and laparoscopic liver biopsies[4] are helpful, and transvenous (transjugular) approach[11] as well. The latter has the advantages of possible hemorrhage to be drained intravascularly and the feasibility of measuring hepatic venous pressure gradients with the upper limit of 10 mmHg for the establishment of HVOD diagnosis, and the higher, the more severe. Unfortunately, this case failed to go through this procedure.

It is very important to differentiate the diagnosis of HVOD from that of BCS[2-4] as both of them present the common signs of abdominal distention, jaundice and ascites with elevated ALB. Several points to identify BCS for this differentiation should be kept in mind: (1) superficial varices of the trunk and lower extremities with edema and pigmentation; (2) stricture or obstruction of IVC and /or HV outflow by venography; (3) thromboses in hepatic venules by liver biopsy pathology.

Supportive care remains the therapy available to date. For some severe cases, ascites must be drained in order to allow sufficient pulmonary ventilation. The drug defibrotide[4,12] has been selected for the treatment of severe HVOD, and in a large, FDA-approved, pivotal, prospective, multi-institutional, global phase Ⅲ trial, seems to have few significant side effects and well-tolerated. Transjugular intrahepatic portosystemic shunting (TIPS) was evaluated[4] for chronic cases with serious obstruction of outlet of main HVs, but should be indicated with discretion. In some severe cases, charcoal hemofiltration[13] has been shown to be effective for adsorbing circulating bilirubin and other protein-bound toxins and for supporting patients in hepatic failure.

This case demonstrated that supervision and instruction of food recipe and traditional medicine are crucial[14], and that prompt diagnosis, supportive care and specific treatment are essential to decreasing the morbidity and mortality of HVOD.

REFERENCES

1 **Bras G**, Hill KR. Veno-occlusive disease of the liver; essential pathology. *Lancet* 1956; **271**: 161-163
2 **Bearman SI**. Veno-occlusive disease of the liver. *Curr Opin Oncol* 2000; **12**: 103-109
3 **Janssen HL**, Garcia-Pagan JC, Elias E, Mentha G, Hadengue A, Valla DC. Budd-Chiari syndrome: a review by an expert panel. *J Hepatol* 2003; **38**: 364-371
4 **Baron F**, Deprez M, Beguin Y. The veno-occlusive disease of the liver. *Haematologica* 1997; **82**: 718-725
5 **Davies TC**. Liver damage caused by extracts of "rattlebox" seeds (Crotalaria). *J S C Med Assoc* 1969; **65**: 201-202
6 **Richardson PG**, Murakami C, Jin Z, Warren D, Momtaz P, Hoppensteadt D, Elias AD, Antin JH, Soiffer R, Spitzer T, Avigan D, Bearman SI, Martin PL, Kurtzberg J, Vredenburgh J, Chen AR, Arai S, Vogelsang G, McDonald GB, Guinan EC. Multi-institutional use of defibrotide in 88 patients after stem cell transplantation with severe veno-occlusive disease and multisystem organ failure: response without significant toxicity in a high-risk population and factors predictive of outcome. *Blood* 2002; **100**: 4337-4343
7 **Tomioka M**, Calvo F, Siguas A, Sanchez L, Nava E, Garcia U, Valdivia M, Reategui E. Hepatic veno-occlusive disease associated with ingestion of Senecio tephrosioides. *Rev Gastroenterol Peru* 1995; **15**: 299-302
8 **Sorensen JS**, Dearing MD. Efflux transporters as a novel herbivore countermechanism to plant chemical defenses. *J Chem Ecol* 2006; **32**: 1181-1196
9 **Liang AH**, Ye ZG. General situation of the toxicity researches on Senecio. *Zhongguo Zhongyao Zazhi* 2006; **31**: 93-97
10 **Yoshimoto K**, Ono N, Okamura T, Sata M. Recent progress in the diagnosis and therapy for veno-occlusive disease of the liver. *Leuk Lymphoma* 2003; **44**: 229-234
11 **Sheela H**, Seela S, Caldwell C, Boyer JL, Jain D. Liver biopsy: evolving role in the new millennium. *J Clin Gastroenterol* 2005; **39**: 603-610
12 **Kornblum N**, Ayyanar K, Benimetskaya L, Richardson P, Iacobelli M, Stein CA. Defibrotide, a polydisperse mixture of single-stranded phosphodiester oligonucleotides with lifesaving activity in severe hepatic veno-occlusive disease: clinical outcomes and potential mechanisms of action. *Oligonucleotides* 2006; **16**: 105-114
13 **Tefferi A**, Kumar S, Wolf RC, Lacy MQ, Inwards DJ, Gloor JM, Albright RC, Kamath PS, Litzow MR. Charcoal hemofiltration for hepatic veno-occlusive disease after hematopoietic stem cell transplantation. *Bone Marrow Transplant* 2001; **28**: 997-999
14 **Zhen LH**. Drug control authorities in China. *Ann Pharmacother* 2004; **38**: 346-350

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