Clinical Case Report

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Combined resection of the right liver lobe and retrohepatic inferior vena cava to treat hepatic alveolar echinococcosis

A case report

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Abstract

Rationale: Hepatic alveolar echinococcosis (HAE) is a potentially fatal and chronically progressive infestation that is caused by the multivesicular metacestode of *Echinococcus multilocularis* (EM). HAE behaves like a malignant tumor and has been referred to as "worm cancer." The main treatment method for HAE is surgical resection.

Patient concerns: We present a 41-year-old Tibetan alveolar echinococcosis (AE) patient with AE lesions invading the right liver lobe and retrohepatic inferior vena cava (RHIVC).

Diagnoses: The patient was diagnosed with HAE based on results obtained from ultrasound examination, computed tomography, liver 3-dimensional reconstruction, serology tests, clinical presentation, and surgical exploration. The final pathology report confirmed the diagnosis as HAE.

Interventions: A radical surgery that combined resection of the liver and RHIVC was performed successfully.

Outcomes: The patient had an uneventful postoperative recovery and a good prognosis.

Lessons: When lesions of the liver significantly violate the RHIVC, resecting the RHIVC without reconstruction may be considered if possible.

Abbreviations: CT = computed tomography, EM = *Echinococcus multilocularis*, HAE = hepatic alveolar echinococcosis, IVC = inferior vena cava, RHIVC = retrohepatic inferior vena cava.

Keywords: alveolar echinococcosis, retrohepatic inferior vena cava, surgical resection

1. Introduction

Hepatic alveolar echinococcosis (HAE) is a chronic zoonotic parasitic disease in which humans are mainly infected by the metacestodes of *Echinococcus multiloculari* (EM). HAE damages liver function through direct erosion and mechanical oppression, finally leading to liver failure. HAE diagnosis mainly depends on epidemiological evidence, clinical presentation, serology test results, radiological examination, and nucleic acid detection.[1] At present, radical resection is the preferred means to treat HAE. The relationship between the location of the lesions, vessels, and bile duct is an important factor influencing radical resection. Therefore, comprehensive preoperative assessment can significantly improve radical resection rates and reduce postoperative complications. The present study reports the case of a patient with complicated HAE who underwent combined resection of the liver and retrohepatic inferior vena cava (RHIVC) and provides a reference for the treatment of similar cases in the future.

2. Case presentation

2.1. Preoperative evaluation

A 41-year-old Tibetan man from the pastoral area of Qinghai Province, who led a long-term nomadic life, presented with upper abdominal pain. B-ultrasound showed a large lesion located in the entire right lobe. The preliminary diagnosis was HAE. Laboratory tests showed the following: hemoglobin: 115 g/L, albumin: 29.8 g/L, globulin: 52.0 g/L, albumin to globulin ratio: 0.6, cholinesterase: 3674 U/L, and normal bilirubin. An assay to detect immunoglobulin G targeting *Echinococcus* was positive, although other tests, including kidney function, blood sugar, and hepatitis A, B, C, and E, gave normal results. Liver computed tomography (CT) (Figs. 1A, B and 2) showed lesions that were located mainly in the right liver lobe with a maximum diameter of 14 × 13 cm, which were diagnosed as HAE. The RHIVC was

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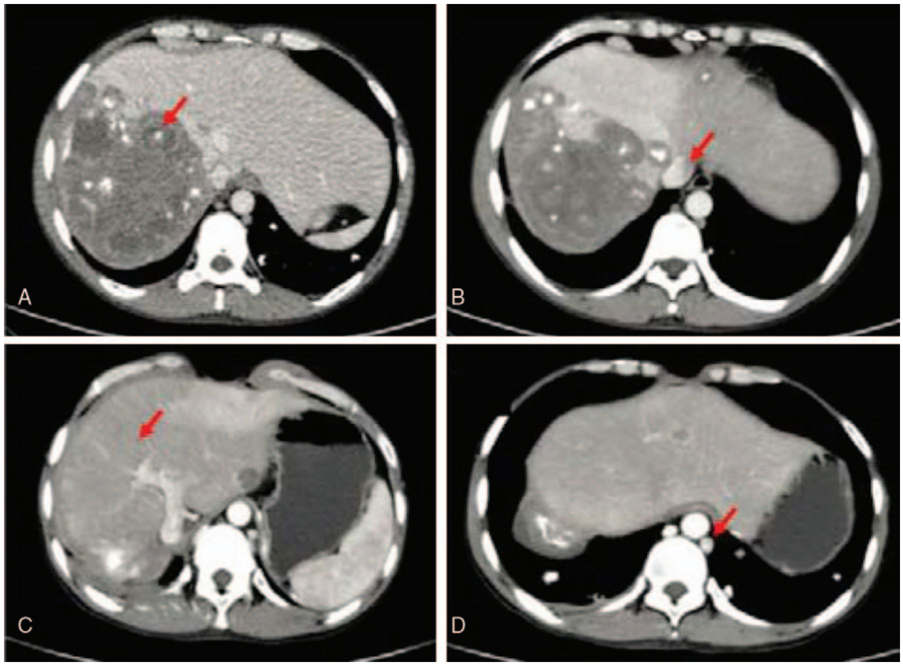

Figure 1. A CT scan of the upper abdomen of an HAE patient. (A) The red arrow shows an alveolar hydatid lesion. (B) The red arrow shows the violations of the IVC. (C) The red arrow shows the left liver after surgery. (D) The red arrow shows an apparently odd vein after surgery. CT=computed tomography, HAE=hepatic alveolar echinococcosis, IVC=inferior vena cava.

oppressed and violated. The right hepatic vein and right portal vein were unclear. The remaining liver volume is shown in Fig. 3. After careful preoperative discussion and obtaining consent from the patient's family, right-sided hepatolobectomy combined with RHIVC resection was performed.

2.2. Surgery process

Surgery was performed under general anesthesia. A thoraco-abdominal J-shaped incision was chosen to provide excellent exposure. After careful surgical exploration, we identified large (15 cm), indurated lesions with a marked upward extension involving the right hepatic vein and the right hemidiaphragm and with a prominent downward extension involving the inferior vena cava (IVC). The right hepatic vein was not present. The confluence of the right hepatic vein was also involved in the lesions. The umbilical and azygos veins were opened and widened in a compensatory fashion. Because the lesion had severely eroded the right hemidiaphragm, the eroded right hemidiaphragm was partly cut. After cholecystectomy, the first portal structures were carefully isolated, and the hepatic pedicles of the right side, including the hepatic artery, portal vein, and bile duct, were ligated. The right lobe of the liver was resected using a cavitron ultrasonic surgical aspirator along the ischemic line on the liver surface. The RHIVC was severely eroded and flattened and was almost blocked. The RHIVC and lesion tissues were separated with difficulty. Some lesions were left on the surface of the RHIVC. The following considerations applied: Pre-operative CT was used to assess the compensatory opening and widening of the azygos vein, and the IVC was almost occluded (Figs, 1A, B and 2); Intraoperative

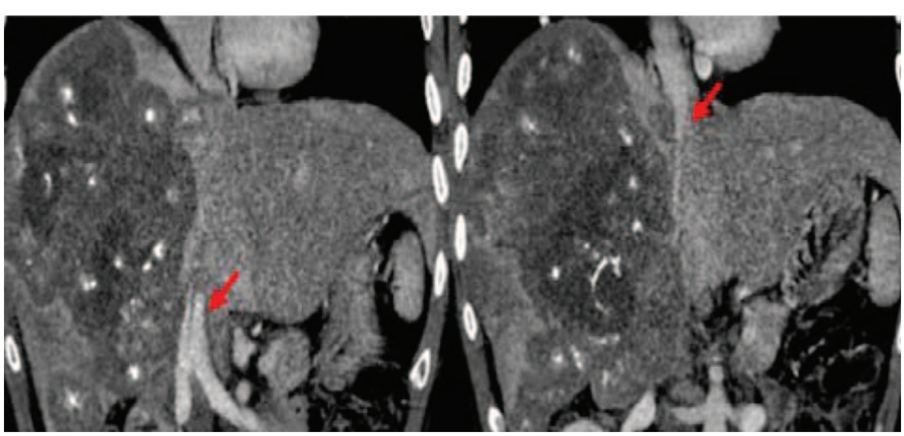

Figure 2. A CT scan of the upper abdomen of an HAE patient. Red arrows show that the alveolar hydatid lesions are involved the IVC. CT=computed tomography, HAE=hepatic alveolar echinococcosis, IVC=inferior vena cava.

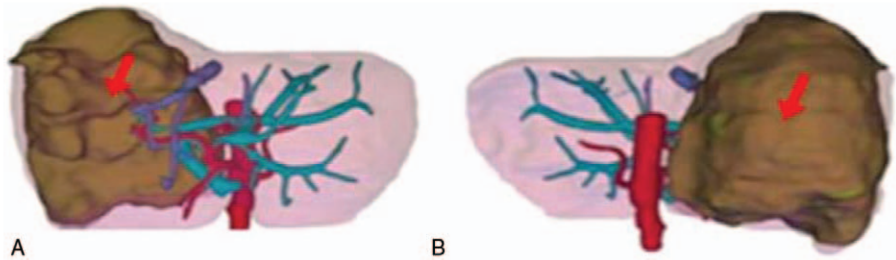

Figure 3. (A) Anterior view (3-dimensional liver reconstruction). (B) Dorsal view (3-dimensional liver reconstruction). Three-dimensional reconstruction of the liver. Liver volume (cm^3): 3785.79. The total volume of the nodule (cm^3): 1488.53. The remaining liver volume (cm^3): 2297.26. All nodules/liver volume ratio: 39.32%. Liver volume excluding the liver nodules and vascular volume (cm^3): 2246.73.

investigation found that the RHIVC was almost eroded, and the azygos vein and umbilical vein were opened and widened in a compensatory mode; If the lesions were left in the RHIVC, the lesions would have recurred in a short time after operation and might have completely violated the RHIVC, resulting in loss of blood return through the IVC, thus affecting the efficacy of surgery; If the RHIVC were to be reconstructed using an artificial graft or autogenous vein, the long-term application of anticoagulant drugs after the operation would have affected the quality of life of the patient; and Reconstruction surgery of the RHIVC can cause greater trauma, longer surgery time, and increase the risk of postoperative thrombosis, thereby affecting the patient's postoperative recovery. Based on these considerations, we decided to cut the RHIVC without reconstruction. The infrahepatic vena cava located at 1 cm on the left renal vein was cut and ligated. After removing the infrahepatic vena cava, the IVC was separated from the retroperitoneum upward and to the suprahepatic vena cava located 1 cm under the confluence with the right hepatic vein. After repairing the right hemidiaphragm, thoracic close drainage was performed. In total, the surgery lasted for 4 hours. The patient was transfused with 5 units of erythrocyte suspension and 1000 mL of fresh-frozen plasma. Blood loss of approximately 3000 mL was sustained, and the urine volume was approximately 1900 mL.

2.3. Postoperative observation

After surgery, the patient exhibited persistent low fever and high white blood cell counts. Hence, antiinfection therapy was provided, and the temperature and white blood cell counts returned to normal at 12 days after the operation. B-ultrasound indicated that the patients suffered from severe pleural effusion at 6 days after the operation, and the bilateral thoracic drainage was retained for 5 days. A total of 1500 mL of light yellow liquid was drawn, and the bilateral pleural effusion was eliminated at 11 days after the operation, at which time the thoracic drainage tubes were removed. In addition, the patient suffered from lower-limb and scrotal edema after the surgery and was given treatment to ameliorate low blood albumin and diuretic treatment; magnesium sulfate was applied to the scrotum. The edema disappeared at 14 days after the operation, and liver function returned to normal on postoperative day 8. The patient was finally discharged on postoperative day 20. The final pathology report diagnosed HAE (Fig. 4). CT images obtained after a 9-month follow-up period are shown in Fig. 1C, D. No severe postoperative complications occurred.

3. Discussion

HAE is among the most dangerous zoonotic diseases worldwide and has a high incidence in northwestern China,[2] especially in pastoral areas.[3] HAE is a larval tapeworm infection that occurs in humans after the ingestion of foods or water that have been contaminated with the larval form of EM.[4] The liver is the most commonly invaded organ. HAE is characterized by chronic progressive hepatic damage that is caused by the continuous proliferation of the larval stage (metacestode) of EM. Pathologically comparable to a slow-growing tumor, the disease is characterized by its invasiveness, exogenous growth, and

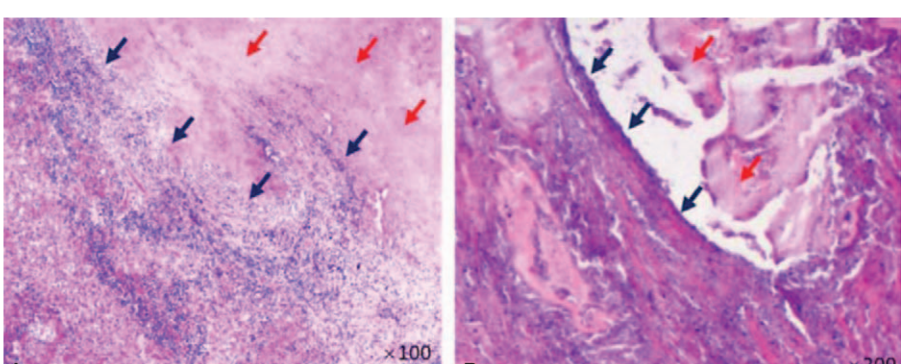

Figure 4. Pathological slices of hepatic alveoli were stained with hematoxylin and eosin (HE). (A) Blue arrows indicate the edge of the lesion with hydatid disease, showing a large degree of inflammatory cell infiltration. The red arrows are placed in the middle of necrotic tissue lesions (×100). (B) Black arrows indicate hydatid germinal epithelium; red arrows indicate necrotic tissue. Cavity formation is clearly visible (×200).

spreading to other organs (lung, brain, and others) via dissemination to surrounding structures, the lymphatic system and vessels. For this reason, HAE has been referred to as "worm cancer." The Chinese government has worked to prevent and treat HAE. The diagnosis of HAE is mainly based on epidemiological evidence, clinical presentation, serology tests, and radiological examinations. According to the latest recommendations of the WHO-Informal Working Group on Echinococcosis (WHO-IWGE) for the management of human alveolar echinococcosis (AE),[5] radical resection (R0-resection) remains the most effective HAE treatment at present. In addition, other treatment methods may be used, including the use of drugs (albendazole), palliative surgery, and liver transplantation.[6] At clinical presentation, most HAE patients show an advanced stage of disease for which radical surgery is no longer as effective as at earlier stages. Although liver transplantation can result in a good prognosis for many types of advanced-stage liver disease, it is difficult to perform due to a lack of donors, high cost, and serious posttransplant complications.

In our case, ultrasound examination, CT, and liver 3-dimensional reconstruction techniques were used to preoperatively evaluate the patient. The WHO-IWGE PNM classification system is similar to the tumor TNM classification, where "P" refers to the extent of parasite localization inside the liver, "N" establishes the involvement of neighboring organs, and "M" evaluates the absence (M0) or presence (M1) of distant metastasis. Our case was at the P4N1M0 phase. The RHIVC was invaded by AE and could not be separated; therefore, it was necessary to remove the RHIVC to achieve R0 resection. Autogenous veins and artificial vessels have been used to reconstruct the RHIVC in patients with lesions infringing the RHIVC.[7] The present study reports the use of a right-side hepatectomy combined with RHIVC resection to treat end-stage HAE at our center. This approach was taken for the following reasons. First, the lesion occupied the right liver lobe and violated the RHIVC, which was almost completely blocked. Chronic obliteration of the RHIVC leads to the establishment of rich umbilical and azygos vein collateral circulation. The anatomical feature of a compensatory azygos vein enables RHIVC resection without serious hemodynamic complications. Second, because the RHIVC was invaded and had become narrow, a thrombus was easily formed. Finally, if the lesion was not completely resected, recurrence was likely. However, the following considerations were applied. First, if the azygos vein, umbilical vein, and other veins can be established as rich collateral circulation veins for patients with lesions that completely violate the RHIVC, perhaps we could use partial resection of the RHIVC as in conventional surgery. Second, we debated whether we could preestablish a vein for compensatory collateral circulation during the preoperative preparation for liver transplantation; if so, we could use the collateral circulation in place of the IVC during liver transplantation. Although the surgery was successful in alleviating the suffering of the patient, prolonging his life, and improving the survival time, some deficiencies remained. For example, inadequate preoperative assessment was available regarding the RHIVC occlusion and the confirmation of the existence of a rich collateral circulation. To overcome this problem, IVC angiography and blood vessel color Doppler ultrasound can be used to accurately assess the size, location, degree of obstruction of the violating IVC lesion, and the establishment of a compensatory collateral circulation before surgery. This approach can be very helpful when selecting the operation mode, assessing the difficulty of the surgery, and treating postoperative complications.

In general, when lesions of the liver significantly violate the RHIVC, autogenous veins and artificial vessels can be used to reconstruct the violated RHIVC in routine operations. The unique and special feature of our case was excision of the RHIVC without reconstruction. The advantages of the current approach are as follows: shortened operation time, decreased complications, less operative trauma, less difficult surgery, lower cost of medicine, and the lack of a need for the application of anticoagulant drugs. In summary, our patient with advanced-stage HAE underwent right liver lobe combined with RHIVC resection without reconstruction; this treatment was effective, and the case provides a reference for future complicated HAE surgeries.

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References

[1] Filippou D, Tselepis D, Filippou G, et al. Advances in liver echinococcosis: diagnosis and treatment. Clin Gastroenterol Hepatol 2007;5:152–9.

[2] Torgerson PR, Keller K, Magnotta M, et al. The global burden of alveolar chinococcosis. PLoS Negl Trop Dis 2010;4:e722.

[3] Tunger O. [Epidemiology of cystic echinococcosis in the world]. Acta Parasitol Turcica 2013;37:47–52.

[4] Nunnari G, Pinzone MR, Gruttadauria S, et al. Hepatic echinococcosis: clinical and therapeutic aspects. World J Gastroenterol 2012;18: 1448–58.

[5] Brunetti E, Kern P, Vuitton DA. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans. Acta Trop 2010;114:1–6.

[6] Sozuer E, Akyuz M, Akbulut S. Open surgery for hepatic hydatid disease. Int Surg 2014;99:764–49.

[7] Hu H, Huang B, Zhao J, et al. Liver autotransplantation and retrohepatic vena cava reconstruction for alveolar echinococcosis. J Surg Res 2017; 210:169–76.