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Resectable hepatoblastoma with tumor thrombus extending into the right atrium after chemotherapy: A case report

Kosuke Endo [a,e,*], Akiko Yokoi [a], Yasuhiko Mishima [a], Akihiko Tamaki [a], Junkichi Takemoto [a], Keiichi Morita [a], Tamaki Iwade [a], Yuichi Okata [a], Hiroaki Fukuzawa [a], Yuko Bitoh [a], Tomomi Hasegawa [d], Makiko Yoshida [b], Yoshinobu Akasaka [c], Hideaki Okajima [e], Yoshihiro Oshima [d], Kosaku Maeda [a], Shinji Uemoto [e]

[a] Department of Pediatric Surgery, Kobe Children's Hospital, 1-1-1 Takakuradai, Suma-ku, Hyogo 654-0081, Japan
[b] Department of Diagnostic Pathology, Kobe Children's Hospital, 1-1-1 Takakuradai, Suma-ku, Hyogo 654-0081, Japan
[c] Department of Radiology, Kobe Children's Hospital, 1-1-1 Takakuradai, Suma-ku, Hyogo 654-0081, Japan
[d] Department of Cardiovascular Surgery, Kobe Children's Hospital, 1-1-1 Takakuradai, Suma-ku, Hyogo 654-0081, Japan
[e] Department of Surgery, Kyoto University, Graduate School of Medicine, 54 Kawahara-cho, Shogoin, Sakyo-ku, Kyoto 606-8507, Japan

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ABSTRACT

Hepatoblastoma with intraatrial tumor thrombus is relatively rare. We report a case of hepatoblastoma with tumor thrombus extending into the right atrium, which responded well to chemotherapy and was resected using extracorporeal circulation. A 4-year-old girl was referred to our hospital because of abdominal distention and tenderness. A computed tomography (CT) scan showed a large tumor occupying the left 3 segments of the liver with tumor thrombus extending into the right atrium. There was also a small intrahepatic metastasis in the right lobe of the liver. She was diagnosed with hepatoblastoma on the basis of the results of open biopsy. Neoadjuvant chemotherapy with an intense CDDP-based regimen was performed. The tumor responded well to chemotherapy, and intrahepatic metastasis became undetectable on CT scan, although the tumor thrombus remained in the right atrium. After 7 courses of chemotherapy, we performed resection using extracorporeal circulation. The postoperative course was uneventful, and adjuvant chemotherapy was started 10 days after the operation. Her serum alpha-fetoprotein (AFP) level decreased to the normal range, and she was free of disease for 1 year after the operation. Tumor resection using extracorporeal circulation can be performed safely and is justified in patients with intraatrial tumor thrombus.

The prognosis of localized hepatoblastoma[1] (HB) has improved over the last few decades by the combination of chemotherapy and operation [1]. Now we are confronted with the challenge of improving the prognosis of high-risk cases such as HB with distant metastasis or local vascular invasion [2]. In the treatment of HB, curative resection greatly improves the prognosis, and recent reports have shown that the rate of curative resection increases with an intense CDDP-based chemotherapy regimen [3,4]. However, there are still cases in which surgical resection is difficult even after chemotherapy, for example, owing to invasion of the main vessels or tumor thrombus extending into the inferior vena cava (IVC) and right atrium. As far as we know, there are only a few reports of HB with tumor thrombus extending into the right atrium resected using extracorporeal circulation. We report a case of a PRETEXT IV HB with the tumor thrombus extending into the right atrium through the left hepatic vein (LHV), which responded well to chemotherapy and was resected using extracorporeal circulation (ECC).

* Corresponding author. Department of Pediatric Surgery, Kobe Children's Hospital, 1-1-1 Takakuradai, Suma-ku, Hyogo 654-0081, Japan. Tel.: +81 78 732 6961; fax: +81 78 735 0910.
E-mail address: kosukee@kuhp.kyoto-u.ac.jp (K. Endo).

[1] AFP: alpha-fetoprotein, CRP: C-reactive protein, CT: computed tomography, ECC: extracorporeal circulation, HB: hepatoblastoma, IVC: inferior vena cava, LHV: left hepatic vein, LTx: liver transplantation, SVC: superior vena cava, WBC: white blood cell.

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1. Case report

A previously healthy 4-year-old girl presented at a local hospital with complaints of fever, abdominal distention, and pain. Physical examination revealed tenderness in the entire abdomen with Blumberg sign, and laboratory data showed a significantly elevated white blood cell (WBC) count and serum C-reactive protein (CRP) levels (33,700/μl and 15.4 mg/dl, respectively). She was diagnosed with acute abdomen and referred to our hospital.

Physical examination revealed a huge tumor palpated around the epigastric fossa. Laboratory data showed an elevated WBC count and serum CRP level. Serum alpha-fetoprotein (AFP) was 305,000 ng/ml. A computed tomography (CT) scan showed a large heterogeneous tumor occupying the left 3 segments of the liver with tumor thrombus extending into the right atrium through the LHV. There was also a small nodule in the right lobe of the liver, which was suspected to be an intrahepatic metastasis (Fig. 1A). No apparent distant metastases were identified by head and chest CT or bone scintigraphy. Open biopsy of the tumor confirmed the diagnosis of HB, mixed epithelial type. The patient was enrolled in the Japanese Study Group for Pediatric Liver Tumor and underwent neoadjuvant chemotherapy with biweekly CDDP and carboplatin/adriamycin according to the regimen of the International Childhood Liver Tumors Strategy Group3-HR under the diagnosis of PRETEXT IV HB. The tumor showed a good response to chemotherapy and shrank significantly, and the intrahepatic metastasis in the right lobe of the liver became undetectable on CT scan (Fig. 1B). No apparent distant metastasis was revealed by chest CT scan. The tumor thrombus also shrank significantly but still existed in the right atrium. The rate of decrease of the serum AFP level lessened gradually, and we judged that the tumor thrombus would not disappear even if we continued chemotherapy. After 7 courses of chemotherapy, we performed the operation.

We performed thoracotomy first and isolated the ascending aorta and superior vena cava (SVC) beforehand in case a pulmonary embolus occurred. Then, an inverted Y-shaped incision was made on the abdomen. Intraoperative ultrasonography revealed that the tumor had shrunk since the last CT scan, and no metastatic lesion was detected in the right lobe of the liver. We considered the tumor resectable by extended left hemihepatectomy with thrombectomy in the right atrium.

The infrahepatic IVC was exposed and encircled with vascular tape for extracorporeal circulation. The liver was mobilized from its ligamentous attachments, and the right hepatic vein was dissected and controlled with vascular tape. Then, we dissected the liver parenchyma using Pringle's maneuver. After finishing the dissection of the liver parenchyma, leaving the left lobe connected to the IVC by only the common trunk of the middle hepatic vein and LHV, we moved to the tumor thrombectomy using ECC. Cardiopulmonary bypass was instituted using cannulation of the ascending aorta, SVC, and infrarenal IVC. A pump sucker was also used during resection of the suprahepatic IVC. The tumor thrombus was free from the right atrium but partly adhered to the wall of the suprahepatic IVC, which was resected with the tumor thrombus and reconstructed with a pericardial membrane patch. The operation time was 14 h, 49 min, and ECC time was 45 min. Total blood loss was 743 ml. The resected tumor was diagnosed as a combined fetal and embryonal type of hepatoblastoma on histopathological examination. Viable cells were found in the tumor thrombus. The postoperative course was uneventful, and adjuvant chemotherapy was started 10 days after the operation. Three courses of adjuvant chemotherapy were completed as scheduled without any significant side effects. The patient's serum AFP level decreased to the normal range (Fig. 2), and CT and MRI revealed no apparent signs of local recurrence in the remnant liver or any distant metastasis. She was free of disease for 1 year after the operation.

2. Discussion

We experienced a case of HB with the tumor thrombus extending into the right atrium. At the first medical examination, the huge tumor occupied the left 3 segments of the liver with tumor thrombus extending into the right atrium through the LHV, and there was intrahepatic metastasis in the right lobe. HB is the most frequent primary malignant liver tumor in children, accounting for almost two thirds of such tumors [5,6]. In many cases, children with

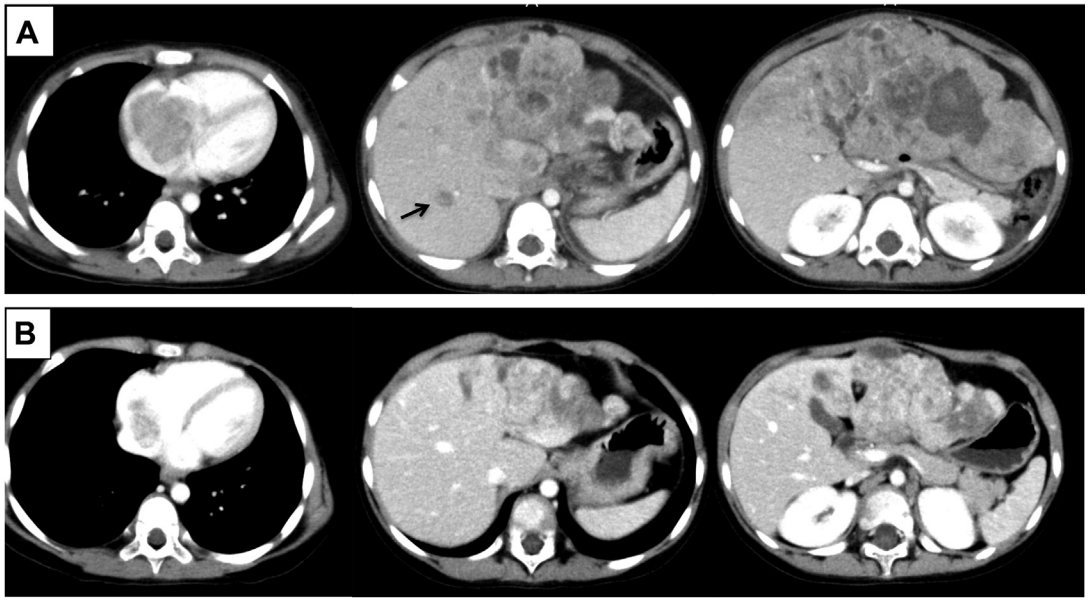

Fig. 1. A) Computed tomography (CT) scan on admission. A huge tumor occupied the left 3 segments of the liver with tumor thrombus extending into the right atrium. There was also a small nodule in the right lobe of the liver, which was suspected to be an intrahepatic metastasis (arrow). B) CT scan after 7 courses of neoadjuvant chemotherapy. The main tumor shrank significantly, and the intrahepatic metastasis in the right lobe of the liver became undetectable. The tumor thrombus still existed in the right atrium.

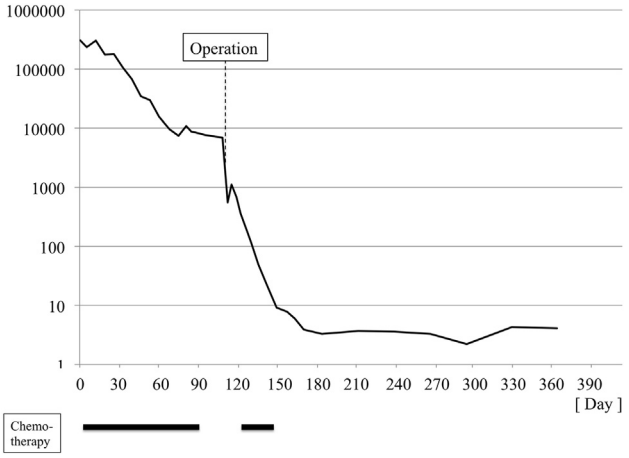

Fig. 2. Serum alpha-fetoprotein (AFP) level. AFP level decreased significantly in response to the chemotherapy and became within normal range after the operation. AFP level remained within the normal range for 1 year after the operation.

HB present with an enlarging abdominal mass, and 70% of the cases are in the advanced stages at diagnosis [5]. However, there are only a few reports of HB with an intraatrial tumor thrombus.

Unlike hepatocellular carcinoma, HB is generally sensitive to chemotherapy, and recently, an intense CDDP-based regimen has been reported to have a high response rate [1–4]. Loh et al. reported 2 cases of HB with tumor thrombus into the right atrium. In both cases, chemotherapy was highly effective, and the tumor thrombus decreased in size up to below the diaphragm, and hepatic resections were performed without ECC [7]. There is even a case report in which a patient with highly advanced HB remained in remission by chemotherapy without any surgical resection [8].

Chemotherapy was highly effective in our case as well. The main tumor shrank significantly, and the intrahepatic metastasis disappeared, although the tumor thrombus still existed in the right atrium. The rate of decrease of the AFP level plateaued gradually. Thus, we decided to perform tumor resection using ECC.

Application of ECC to oncologic surgery is controversial. Hasegawa et al. reported a case of intrapulmonary tumor spread after surgical resection of intrathoracic extension of thyroid cancer with cardiopulmonary bypass [9]. On the other hand, Kauffmann et al. reported that surgery on ECC for advanced non-small cell lung carcinoma resulted in satisfactory results without surgery-induced tumor propagation [10]. In our case, the rate of decrease of the AFP level gradually became slower throughout the course of neo-adjuvant chemotherapy. As surgical resection should be performed before tumors become resistant to chemotherapy, we considered that ECC was inevitable for the resection. There was no evidence of recurrence for 1 year after the operation, which might indicate that the adjuvant chemotherapy was effective for the tumor. We believe that resection on ECC would be justified for cases of HB because the resectability of the tumor has a great impact on the prognosis of this disease.

Using the pump sucker during the operation for a malignancy is uncommon, because it could increase the risk of tumor propagation. However, since in our case, preoperative chemotherapy was highly effective and the tumor thrombus had shrunk significantly, and preoperative echocardiography showed no invasion of the tumor thrombus into the right atrial wall, we thought that we could control the micro-metastasis if we resected the tumor thrombus en block and began adjuvant chemotherapy early after the operation. We made the operative procedure simple by using a pump sucker and ECC with the heart beating, which attributed to the early postoperative recovery and the resultant early initiation of adjuvant chemotherapy. As a result, we believe we could control the insidious micro-metastasis.

In our case, liver transplantation (LTx) might have been another surgical treatment option, because the survival rates after primary LTx are reported to be remarkably higher than those after rescue LTx, incomplete resection with partial hepatectomy, or relapse after a previous partial hepatectomy [11]. However, macroscopic venous invasion was reported to be an independent risk factor of overall survival after primary LTx [11,12]. We refrained from LTx for fear that an insidious micro-metastatic lesion might recur due to the immunosuppressed status after LTx.

3. Conclusions

In summary, we experienced a case of PRETEXT IV HB with tumor thrombus extending into the right atrium. Since complete resection of the tumor is considered essential for the cure of HB, we concluded that tumor resection using ECC could be justified in a patient with HB extending into the right atrium.

Conflict of interest

All authors have no conflict of interest.

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