Case Series of Hepatoblastoma

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Abstract
Background: In the past 20 years, a dramatic improvement in the prognosis of patients with hepatoblastoma (HB) has been achieved by combining surgery and chemotherapy in several national and international trials.

Materials and Methods: Four children (3 girl and 1 boy) aged 34 days, 4 months, 13 months, and 1 year with mean age of 6.53 (±5.15) months were presented to our center from 2002 to 2009 with abdominal mass and abdominal distention. Their abdominal ultrasonography revealed liver mass, and pathological diagnosis was hepatoblastoma. They were treated with a combination of surgery and chemotherapy.

Results: Patients with non-metastatic HB were treated at Shahid Sadoughi Hospital of Yazd. A complete resection of primary tumor was achieved in all of them. The median follow-up of the patients was 38.25 (± 29.02) month and all of them (100%) remained alive with complete remission.

Conclusion: Complete resection with chemotherapy can improve prognosis of hepatoblastoma patients.

Keywords: Hepatoblastoma, Chemotherapy, Cisplatin, doxorubicin.

Introduction
Hepatoblastoma (HB) is a rare malignant liver tumor which occurs almost exclusively in childhood. It accounts for approximately 1% of all pediatric cancers. There is general agreement that complete surgical resection is the cornerstone of treatment for children with HB and the only way for eventual cure. Chemotherapy has been shown to improve survival of the patients.

Here, we describe 4 patients with HB who were treated by chemotherapy after surgery and discuss the role of chemotherapy in the treatment of HB.

Cases
We report 4 infants (3 girls and 1 boy) with non-metastatic HB referred to us from 2002 to 2009 with abdominal mass and/or distention. The median age at presentation was 6.53 months (± 5.15) (range; 34 days to 13 months).

Case 1
At 4 months of age, a boy was presented with an abdominal mass. Abdominal computed tomography (CT) scan showed the tumor involved the left lobe of the liver and serum alpha-fetoprotein (AFP) level was elevated to 1210 ng/ml. Liver function tests (LFT) were elevated, too. On surgery, tumor and left lobe of the liver were resected completely. The pathological diagnosis was hepatoblastoma of epithelial type. The patient was treated with 6 courses of adjuvant chemotherapy including doxorubicin and cisplatin. Serum AFP level decreased. He has been followed monthly and is well 4 years after the date of diagnosis.

Case 2
An 8-month-old girl was found to have an abdominal mass without tenderness. Abdominal ultrasonography revealed a mass in the left lobe of the liver with heterogeneous echogenicity measuring 85*77 cm. Abdominal CT scan also showed a heterogeneous mass in the left lobe of liver. Serum AFP level was elevated but alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels were normal. In surgery, the mass and left lobe of the liver was completely resected. Pathological
diagnosis was pure epithelial type hepatoblastoma. The patient was treated with 6 courses of adjuvant chemotherapy including doxorubicin and cisplatin. She has been well 4.5 years after the diagnosis.

Case 3
A 13-month-old girl was presented with an abdominal mass in right upper quadrant without tenderness. She complained of intermittent vomiting and anorexia too. Her abdominal CT scan revealed a hypodense mass in the right lobe of liver. Spleen and kidney were normal. The mass was removed completely by surgery and no metastases were seen. Pathological diagnosis was pure epithelial type hepatoblastoma. The patient was treated with 7 courses of adjuvant chemotherapy including doxorubicin and cisplatin. She has been well 30 months after diagnosis.

Case 4
A 34-day-old neonate girl was presented with abdominal distention. Her abdominal sonography showed severe hepatomegaly. Spleen and kidney were normal. LFT was normal too, but serum AFP level was elevated. The mass was removed completely by surgery and the pathological diagnosis was hepatoblasma. The patient was treated with 6 courses of adjuvant chemotherapy including doxorubicin and cisplatin. The child is alive and well 2 years after the diagnosis.

Chemotherapy and follow up
Initial treatment was with doxorubicin 25mg/m2/day for 3 days as continuous infusion, cisplatin 20mg/m2/day for 5 days as continuous infusion, days 0-4 (6 cycles of chemotherapy). For patients less than 10 kg, chemotherapy was administered on the basis of weight of the patient as follows: doxorubicin 0.83mg/kg/day for 3 days, and cisplatin 0.66mg/kg/day (6 cycles of chemotherapy). Cardiac function was monitored by two-dimension mode echocardiography. After completion of treatment, patients were followed monthly by physical examination and measurement of serum AFP levels. At the time of this report, the median duration of follow-up was 38.25 months (± 29.02) (range; 6 months to 5 years). All of the children are doing well with complete remission.

Discussion
Hepatoblastoma (HB) is the most common primary malignant tumor of the liver in children. Complete surgical resection is still the mainstay of treatment for HB. Recently, it has been reported that cisplatin-based chemotherapy along with doxorubicin is effective in patients with HB. Survival of the affected children was approximately 25% before the development and use of effective chemotherapy. Recently, postoperative adjuvant chemotherapy has improved the survival of patients whose tumors could be completely resected, on the other hand, complete surgical excision has been possible in patients who had unresectable tumors before preoperative neoadjuvant chemotherapy.

References