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Successfully Surgical Treatment of Lung Metastatic Hepatoblastoma: A Rare Case Report

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Abstract

Background

Hepatoblastoma is a common liver malignancy in children and commonly presents with primary tumors. In hepatoblastoma, lung is the most common place to metastasis. Chemotherapy have led to many improvements in the local control of hepatoblastoma. A main goal of treatment for hepatoblastoma is to achieve complete tumor resection.

Case Presentation

The patient was a 2.5 years old boy with abdominal distention and abdominal pain. Abdominal and pelvic ultrasound and thoracic and abdominal CT was performed for the patient and the results of them showed a large and hyperecho mass in the liver and several nodular lesions in lung segments. After doing some other tests, the diagnosis for the patient was hepatoblastoma. After chemothetapy the primary tumor was removed by surgery. Follow-up by CT scan after second chemotherapy showed that the lesions in the liver were removed, but lung masses were still unchanged and after second surgery, lung masses were removed too. The outcome has been favorable with no recurrence as of 20 months after the operation.

Conclusion

In our case, the patient did not respond to chemotherapy and as main treatment, surgery was carried out, that shows its importance in the treatment of hepatoblastoma.

Key Words: Children, Hepatoblastoma, Lung metastasis, Surgical Treatment.

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1- INTRODUCTION

Hepatoblastoma (HB) is one of the most common liver malignancies in children (1), and it is considered as a rare tumor for approximately 1% of all pediatric malignancies. Hepatoblastoma commonly presents with primary tumors that are not amenable to resection. Improvements in neo-adjuvant chemotherapy have led to successes in the local control of the tumor (2). At presentation, 50% of the tumors are considered unresectable, yet half of these can be rendered resectable with modern preoperative chemotherapy (3).

In some studies, in the management of hepatoblastoma, complete tumor resection is a crucial step on the route to cure. Therefore, a main goal of treatment of hepatoblastoma, is to achieve complete tumor resection (4). In hepatoblastoma, most common place to metastasis, is the lung (5, 6). Historically, management of pulmonary metastases has been approached in a variety of fashions integrating chemotherapy and surgical resection (7).

The presence of lung metastases is not an absolute contraindication for partial liver resection or liver transplant because many of lung metastases respond to chemotherapy very well and can disappear completely, or become resectable by compeleting of pre-operative chemotherapy (4). So, in this case report study, we describe the case of a 2.5 years old boy with hepatoblastoma that is resistant to chemotherapy and became treated by surgical resection.

2- CASE REPORT

We present the case of a 2.5 years old male patient with abdominal distention and abdominal pain. He did not report other associated symptoms followed by abdominal pain such as loss of appetite, weight loss, vomiting and fever. The patient's history of about a year, showed that, sometimes generalized itchy papular lesions were appear on the child's body that with diagnosis of allergy, was treated with antihistamines. He is the first child of his family and there was not any certain disease in history of the patient and his parents. At the beginning that the patient visited with abdominal pain, abdominal and pelvic ultrasound was requested for the patient. The primary results of ultrasound showed a large and hyperechoic mass in the liver. After that, thoracic and abdominal CT scan was also performed for the patient. The results showed a large-sized 75×71×61 mm mass in the right lobe of the liver (**Figure.1**).

Also, several nodular lesions with a diameter of 8mm were observed in lung segments, which can be suggestive as metastatic involvement. Ascites and pleural effusion were not observed. All patient’s hematological tests were normal. But the level of Alpha-fetoprotein in AFP test, was 415 that is higher than normal range. Also Beta-Human Chorionic Gonadotropin (βHCG) test was negative. Finally, bone marrow sampling was performed for the patient. According to the results of laboratory and diagnostic tests, the final diagnosis of the patient was hepatoblastoma, and 6 course of chemotherapy were begun for patient.

During chemotherapy, follow-up measuring of the level of AFP and re-imaging was performed. The results of the tests showed that the patient was not respond to chemotherapy and likewise, AFP level was high, and tumor size did not change. After completing the cycles of chemotherapy, the primary tumor was removed by liver lobectomy surgery (**Figure.2**). Pathology results after surgery confirmed hepatoblastoma, and after resection of the mass from liver, patient’s AFP level was checked that in this test a significant reduce from 415 to 219 ng/mL was observed.

The second chemotherapy was begun again in 6 sessions. Follow-up CT scan after second course of chemotherapy showed that the liver was in normal size and space-occupying lesions in the liver were removed. But lung masses in the upper and anterior segments of the left uper lobe (LUL), left lower lobe (LLL) was still unchanged compared with previous imaging. This lack of response to second chemotherapy caused that the second surgery for resection of pulmonary nodules to be performed (**Figure.3**). Postoperative pathology results also indicated metastatic hepatoblastoma. After 5 days after the second surgery, imaging and laboratory tests were performed for the patient again, that all of them was normal and the patient's AFP level reached to 21 ng/mL. In addition, the periodic follow-up of patient was performed in intervals of every two months to 20 months, and the results of all tests was normal.

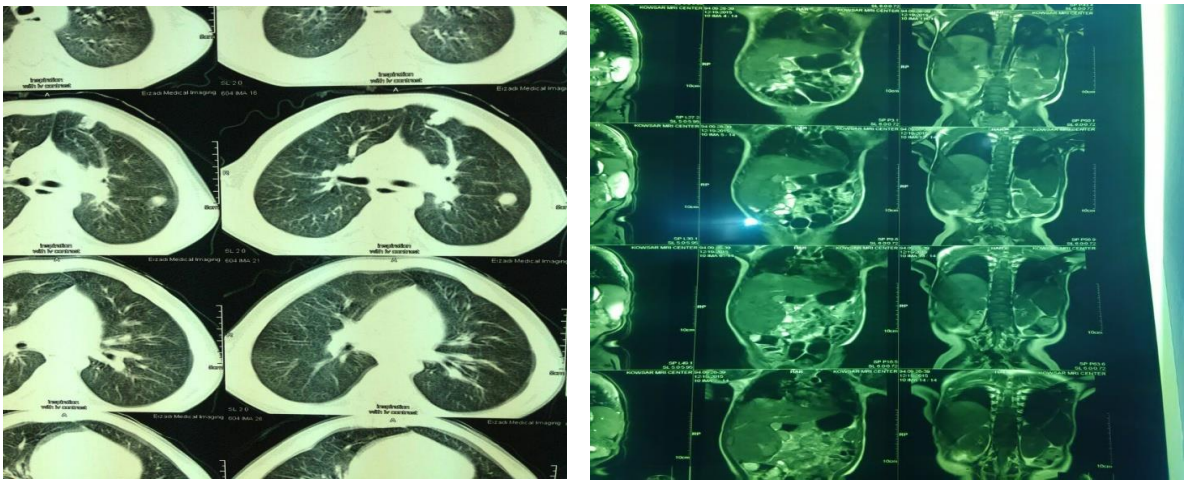

Fig.1: CT scan showing mass in the liver and lung.

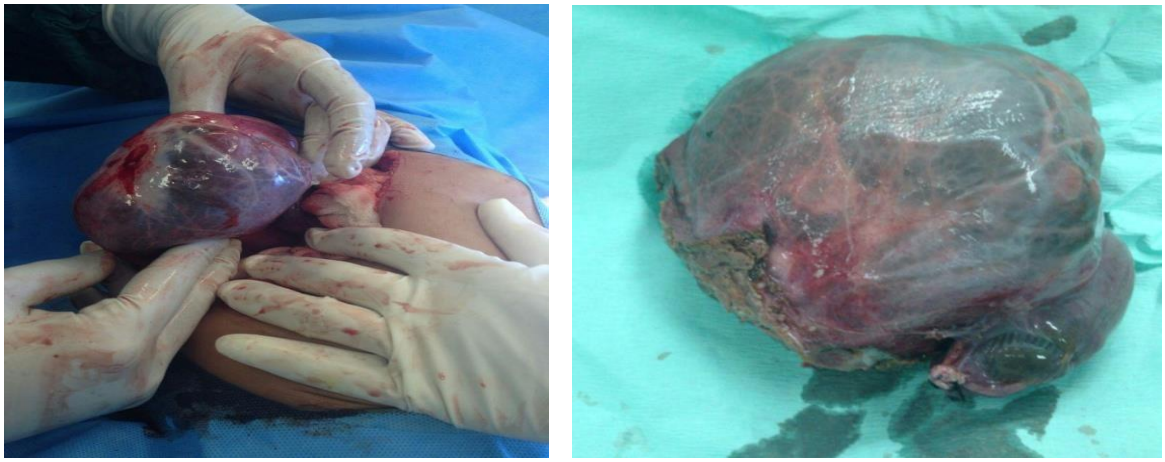

Fig.2: Remove tumor of liver with lobectomy surgery.

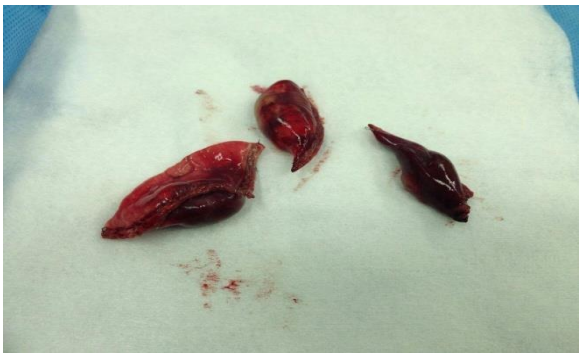

Fig.3: Metastatic nodules of lung.

3- DISCUSSION

It has been proven that Hepatoblastoma is related to Beckwith-Wiedemann syndrome, hemi-hypertrophy and Prader–Willi syndrome. Moreover, six cases of hepatoblastoma in children with trisomy 18 have been published since 1987, when Dasouki and Barr reported the first case, which was presumed to be hepatoblastoma (8). Out of all childhood tumors, excluding leukemia and lymphoma, hepatic tumors form about 0.5-2% in frequency. The most common primary malignant liver neoplasm in children is hepatoblastoma (9). Hepatoblastoma, clinically and biologically, is a heterogeneous disease. In addition, children with metastatic disease, have poorer outcomes than patients with localized disease (10).

Two thirds of cases occur before 2 years old of age and 90% of the cases are below 5 years old. Males are affected two times more compared to females (11). Our presented patient was a 2.5 years old boy. High levels of serum AFP in a child between 6 months and 3 years old of age is diagnostic for hepatoblastoma. High levels of AFP in hepatoblastoma, may suggest massive tumoral extensions, and /or presence of metastasis signify an unfavorable prognosis (9).

In our case, we considered AFP level, and the diagnosis was confirmed by very high level of AFP, and by the physical examination, abdominal ultrasound and chest X-ray findings. So, routine follow-up examinations after each part of initial treatment were done. Hepatoblastoma develops more frequently in the right hepatic lobe (12). The left hepatic lobe receives oxygenated blood totally from the umbilical vein, while the right lobe is irrigated with blood from the portal vein, with lower oxygen saturations. The low blood pressure of the oxygen could favor the embryonic differentiation of the hepatoblastoma in certain conditions that is more common in the right hepatic lobe (13). Our patient showed a left lobe hepatoblastoma, which is less common.

The International Society of Pediatric Oncology Liver Tumor Study Group launched its first prospective trial (SIOPEL-1) with the intention of treating all patients with preoperative chemotherapy for the following reasons:

1) The experience of individual surgeons. Compared with the resection of HBs at the time of diagnosis, most surgeons agreed that operating on tumors that had become smaller after chemotherapy was easier, and, hence, safer, because the tumor became better defined, less friable, and less prone to bleeding.

2) Visible metastases were present at the time of diagnosis in 20% of patients; these and micro metastases would be exposed to chemotherapy earlier.

In other studies, these patients usually, but not consistently, were given preoperative chemotherapy as well (3). It was desirable to establish a multidisciplinary approach at the onset with the objective of standardizing the selection and clinical grouping of patients, including use of a uniform staging system. For this reason, a pretreatment extent of disease (PRETEXT) grouping system was designed specifically for patients with liver tumors (the predictive value of this system will be discussed in more detail elsewhere). Standardization is particularly important in an international and multi-institutional study of such a rare pediatric tumor. The published overall results of this completed study support the strategy chosen (14-16); but in our case, the patient did not respond to chemotherapy before surgery and tumor size did not change.

A complete surgical resection is the cornerstone of treatment (17), and before the 1980s, only surgical treatment alone, was directed toward thisaim. Towu in his study demonstrated that, in the 1980s, it became evident that cisplatin based chemotherapy often dramatically decreased the size of the primary tumor and allowed some previously unresectable tumors to be excised successfully. Surgical excision after this type of chemotherapy improved survival figures by more than 50% (3).

Busweiler and colleagues (2016) examined therapeutic manners of 103 children with hepatoblastoma between 1990 and 2013; 92% of these patients underwent partial hepatectomy surgery and were cured (18). Our case had a complete excision of the tumor with partial hepatectomy, based on histological examination. A prospective trial by the International Society of Pediatric Oncology Liver Tumor Study Group noted visible metastases in almost 20% of patients presenting with hepatoblastoma (19). Several studies have confirmed the presence of metastatic lung hepatoblastoma (20-23). Pulmonary surgery is frequently used for the treatment of metastasis in children. However, hepatoblastoma is a relatively rare tumor, the use of metastasectomy in this tumor remains poorly characterized. Matsunaga et al., highlighted the importance of complete resection of the hepatic primary tumors in patients with distant metastases (24). In the presented case, pulmonary metastatic lesions were with high level of AFP. The post-surgical pulmonary metastasectomy evaluation of AFP level was favorable and this showed that the surgery was successful.

4- CONCLUSION

Eventually, although preoperative chemotherapy applied as initial treatment in pediatrics hepatoblastome, but in our case, the patient did not respond to chemotherapy and surgery as main treatment was carried out. As well as, the outcome has been favorable with no recurrence as of 20 months after the operation.

5- CONFLICT OF INTEREST: None.

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