

RECURRENCE OF HEPATOBLASTOMA WITH METASTASES TO THE ANTERIOR ABDOMINAL WALL IN A CHILD AFTER 5 YEARS SURGICAL RESECTION. A CASE REPORT WITH LITERATURE REVIEW.



HAZLINA MOHD KHALID¹, SHUNMUGAM RAJAH¹, ZAKARIA ZAHARI², ASHOK KRISHNAN¹, SITI HAJAR OMAR³

¹DEPARTMENT OF PAEDIATRIC SURGERY, SABAH WOMEN AND CHILDREN'S HOSPITAL, MALAYSIA

²DEPARTMENT OF PAEDIATRIC SURGERY, HOSPITAL KUALA LUMPUR, MALAYSIA

³DEPARTMENT OF RADIOLOGY, SABAH WOMEN AND CHILDREN'S HOSPITAL, MALAYSIA.

INTRODUCTION.

Hepatoblastoma (HB) is the most common primary malignant liver tumor in children under the age of five, accounting for 1% of childhood malignancies. Hepatoblastoma is the third most common abdominal neoplasm in children after neuroblastoma and nephroblastoma. It is more common in boys and has a incidence of 1 to 1.5 cases per million population. In children diagnosed with hepatoblastoma (HB), the lungs are the most common site of metastasis at both initial presentation and relapse. Abdominal wall metastases is almost unheard of.

We report a case of child who diagnosed to have recurrent hepatoblastoma with metastases to the anterior abdominal wall after five years surgical resection.

We report a case of recurrent hepatoblastoma heralded by rising alpha-feto protein (AFP) level and anterior abdominal nodule after five years of right hepatectomy.

MATERIAL and METHOD

Retrospective review of medical record a child who had recurrent hepatoblastoma with metastatic nodule at the abdominal wall who was treated in our department from January 2011 till December 2016. The clinical presentation, surgical procedure undertaken, radiology and histopathology findings were evaluated.

CASE REPORT

A 3-year old girl was admitted in one the district hospitals with the complaint of abdominal distension and discomfort for 2 weeks associated with intermittent episodes of fever. She had a palpable, non-tender, firm liver and was treated as a case of enteric fever. She had completed a course of antibiotics. During her follow-up noted she had persistent hepatomegaly. Abdominal ultrasonography (USG) noted a huge heterogenous lesion occupying the right lobe of liver measuring 8.7cm x 6.5 x 11cm. AFP was 35,350 ng/mL. She was referred to our centre for further management. Computed Tomography (CT) chest, abdomen and pelvis was performed and revealed multiple lung nodules and large heterogenous mass measuring 12.8cm x 10.6cm occupying right lobe of liver with cystic area within [FIG 1]

USG guided trucut biopsy of the liver lesion was done and revealed a hepatoblastoma of pure fetal type. Neoadjuvant chemotherapy; Cisplatin + Doxorubicin according to SIOPEL 3 High Risk Protocol was initiated. After completed 6 courses, repeated CT thorax and abdominopelvis showed a marked reduction in tumour size and no lung nodules. [FIG 2] AFP level was 1568 ng/ml.

Right hepatectomy was performed [FIG 3]. No perioperative complication. Histopathology showed mixed fetal and embryonal type of hepatoblastoma. There was no vascular invasion with negative resection margin. She completed another 9 courses of chemotherapy. AFP level showed gradual decrease, from 1240ng/ml to 3.6 ng/ml two months after surgery.

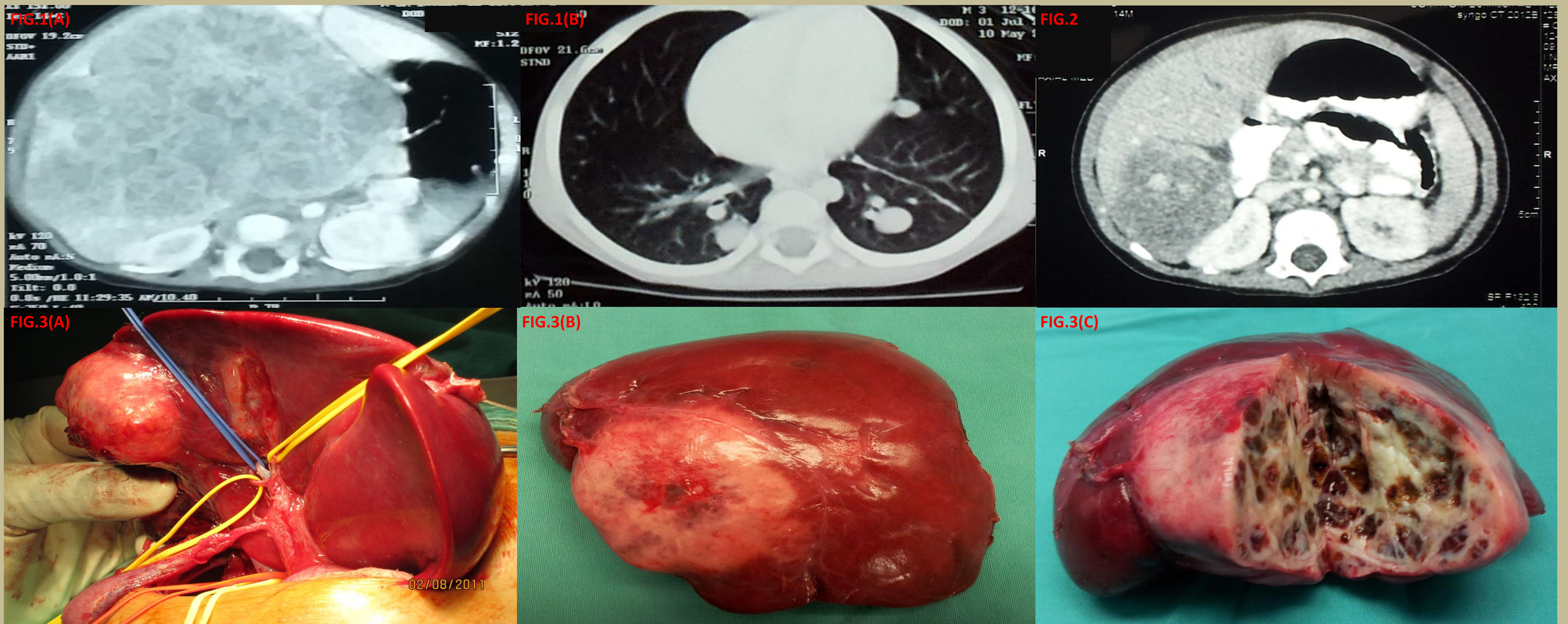


FIG. 1(A) & 1(B) :CT showed large heterogenous liver mass involving right lobe measuring 8.7cm x 6.5 x 11cm with some areas of necrosis and multiple lung nodules suggestive of metastases.

FIG. 2 : Repeated CT after receiving neoadjuvant chemotherapy showing significant reduction in tumour size and clearance of lung nodules.

FIG. 3(A, B & C) : Intraoperative found tumor occupied in the segment VI/VII. Right hepatic lobectomy was performed. On cut section noted some areas of necrosis. Histology revealed mixed fetal and embryonal type.

She was well during this 5-years follow-up with the AFP level remain < 5 ng/ml and follow up USG abdomen and CT scan showed no lesion in the liver, abdomen or lung till in May 2016 noted elevation in AFP; 77.3 ng/ml with further increase to 432 ng/ml and discovered small nodule at the anterior abdominal wall measuring 4cm x 3cm. CT scan revealed a suspicious soft tissue lesion of anterior abdominal wall muscle measuring 3.5cm x 2.0cm x 3.5cm. No evidence of local or lung metastases. [FIG 4]. We proceeded with an excision of the nodule/lesion. Histopathology showed a metastatic lesion which confirmed recurrence. She was started on IRINOTECAN and remains under our care with latest AFP serum level is 8.9 ng/ml and normal USG.

DISCUSSION.

Approximately 90% of HB cases occur in patients under 5 years of age and two thirds of the cases occurs in the first 2 years of life as in our child. They can present with abdominal swelling and hepatomegaly; or some cases are asymptomatic and present in an advanced stage. The diagnosis is established according to clinical (age), radiological imaging, serum (very high AFP level) and histological criteria. Our case was delayed in diagnosis due to mistreated as enteric fever.

HB commonly presents with primary tumours not amenable to resection. Approximately 20% of patients present with metastatic disease, most frequently in the lung. It is considered as high-risk disease and has poor prognosis, with an overall survival of 50 – 60%.

Surgical resection is the cornerstone of treatment for patient with HB. The Society of Paediatric Oncology Liver Tumor Study Group launched its first prospective trial (SIOPEL-1) in early 90's with the intention to treat all patients with preoperative chemotherapy and delayed surgical resection. This study revealed that neoadjuvant chemotherapy (combination cisplatin and doxorubicin) seems to make tumour resection easier and a greatly improved prognosis. As illustrate in our case, the preoperative chemotherapy downstage the tumour and make it feasible for resection. Resection of a positive margin does not necessarily have to be performed, because postoperative chemotherapy showed good results. With this current treatment most centers now achieve 3-year survival rate approaching 80%.

The AFP level monitoring represents a valuable marker of the response to the preoperative chemotherapy, in the evaluation of the excision result and for the precocious diagnosis of the HB relapse. The complete excision of the HB determines the decrease of the AFP serum level, which will be normalized after 4-6 weeks. Our patient had 4 years postoperative normal values of AFP, confirming complete tumor resection. The persistent or secondary increase of the AFP value suggests a residual tumor, metastases or a relapse. The AFP serum level increases very much before the radiology imaging demonstration of the tumor relapse as shown in our case.

Local recurrence or pulmonary metastases are the most common site in relapse case of HB but abdominal wall metastases is almost unheard of. This case report exhibits the possibility of metastases to sites other than the lung and demonstrate the need of more vigilant follow up in such cases despite a 4 year disease free period. The USG monitoring and AFP measuring in dynamics are compulsory

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There is no COI to be disclosed

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