



# HEPATOBLASTOMA WITH TUMOR EXTENSION INTO RIGHT ATRIUM

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## Introduction

Hepatoblastoma is the commonest malignant liver tumor in children. The prognosis has improved over the years with the combination treatment of chemotherapy and surgery. Complete surgical resection has always been the cornerstone of cure. However, the extension of the tumor thrombus into the right atrium poses a significant technical challenge for complete resection. In this report, we share our experiences in handling this extremely rare condition.

## Case Report

A case of a 2 year-old girl who was diagnosed to have PRETEXT II Hepatoblastoma with a tumour thrombus extending into the right atrium. The main tumour responded well to SIOPEL 4 HR chemotherapy but the tumour thrombus did not regress. The technical difficulties for a curative surgical resection is compounded by the non-availability of on-site cardiothoracic services. Surgical excision was then decided to be done in a same setting with the cardiothoracic team in their centre. The surgery was started with a median sternotomy and sling of both pulmonary arteries in anticipation of a pulmonary embolism, followed by a roof top incision for a right hepatectomy. Tumour was seen at segment VII and VIII, closely adhered to inferior vena cava (IVC) and diaphragm requiring resection of part of the right hemidiaphragm along with the tumour. Following liver resection, the excised tumour was left connected to the IVC and cardiopulmonary bypass was instituted for open thrombectomy from the right atrium. The right hepatic vein was then isolated and venotomy was done to identify the thrombus. Thrombectomy was performed and then the tumour was completely removed en-bloc. Bleeding was encountered from the raw areas following reinstatement of cardiopulmonary bypass but managed to be secured. There were no immediate postoperative complications and she recovered well from the surgery. Histopathological examination reported as hepatoblastoma with chemotherapy changes and calcified thrombus seen without residual viable malignant cells. Alpha fetoprotein was normalized after surgery. She was well upon follow up with no evidence of recurrence.

## Discussion

Secondary cardiac tumours following tumour extension into the right atrium via the inferior vena cava (IVC) are extremely rare in children. Besides hepatoblastoma, few other solid tumours that were reported to have intracardiac tumour extension included Wilms tumour, neuroblastoma and hepatocellular carcinoma<sup>1</sup>. In managing hepatoblastoma, complete resection of the tumour carries the best prognosis and, with the good response to chemotherapy, numbers of resectable hepatoblastoma has been increased with decreasing morbidity of surgery<sup>2,3</sup>. However, cases of hepatoblastoma with intra-atrial tumour extension which persist even after chemotherapy, has posed to be a great challenge to the managing surgical team who aims for a complete resection. Combination of liver and cardiac surgery is required, aiming for "en-bloc" resection of both liver tumour together with its intra-atrial tumour extension without rupturing or detaching it<sup>4</sup>. We have successfully achieved this goal; we started with hepatectomy and prior to opening up the IVC, cardiopulmonary bypass was instituted, followed by cardiorespiratory arrest. This enables the opening of the right atrium and IVC for complete "en-bloc" resection. The surgery was successful and we would like to advocate using this similar approach for this group of patients with intra-atrial tumour extension.



Figure 1: Series of CT images pre-chemotherapy (left) and post chemotherapy (right) showed a significant reduction of liver tumor size after chemotherapy



Figure 2: Echocardiogram images demonstrated the presence of the tumor thrombus in the right atrium

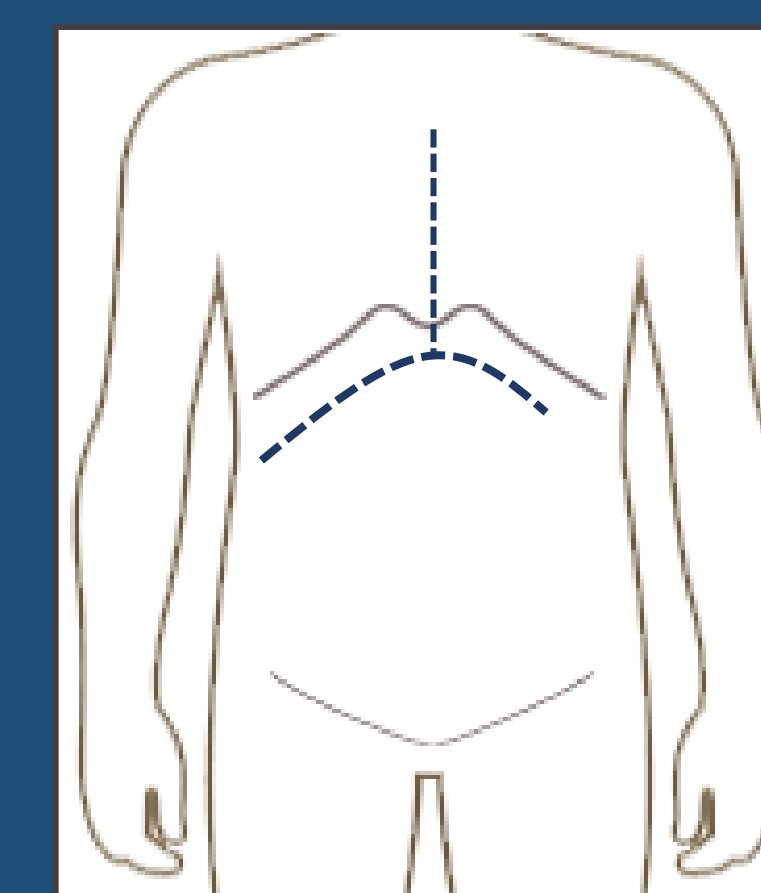


Figure 3: Line of incision

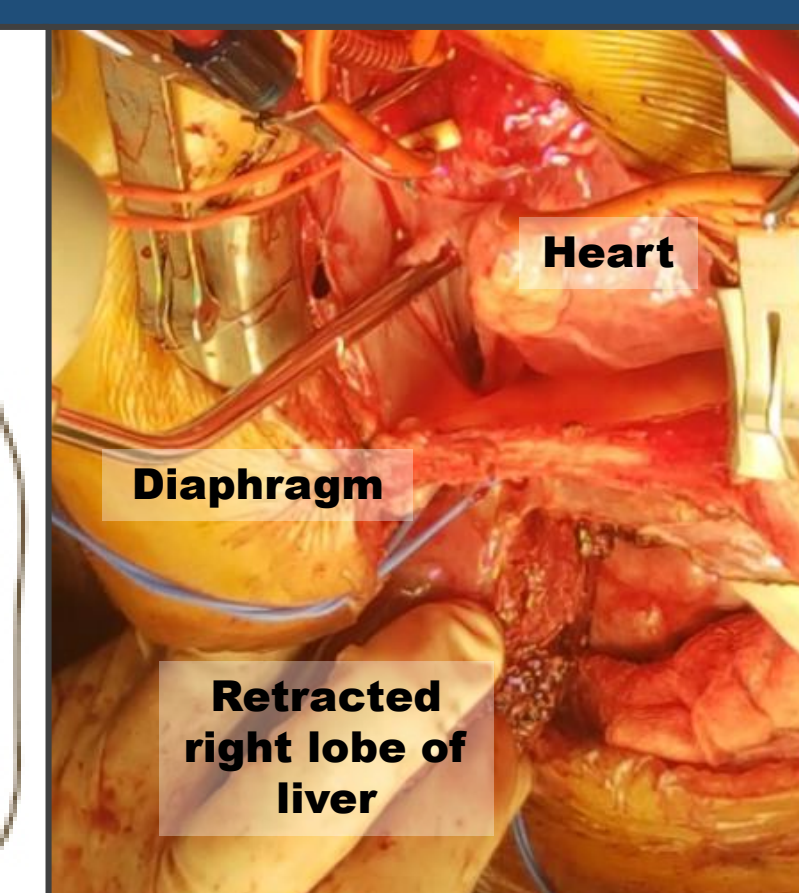


Figure 4: Combination of median sternotomy and laparotomy

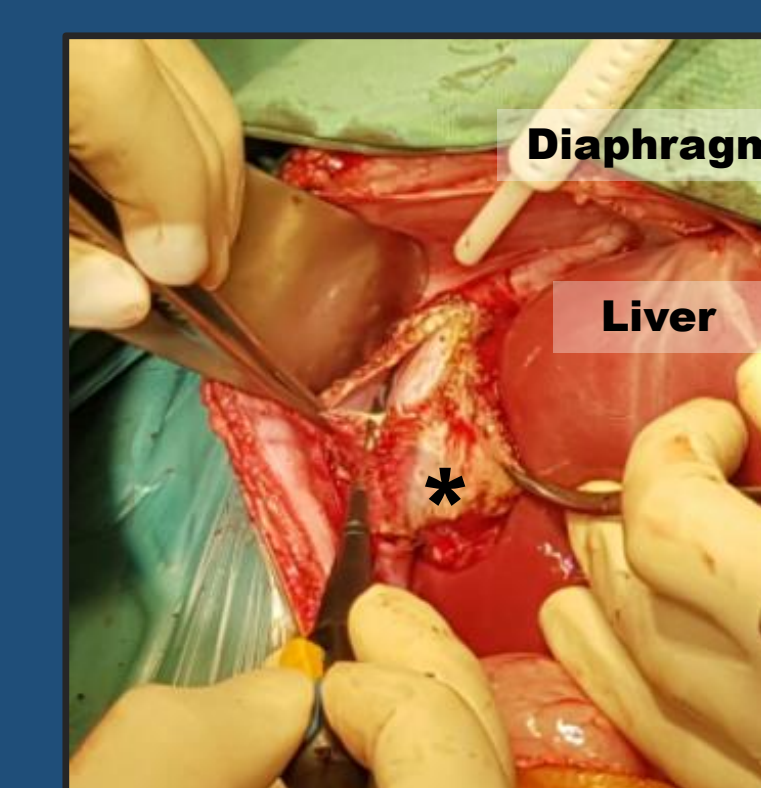


Figure 5 (Abdominal view): The liver tumor (\*) closely adhered to the right hemidiaphragm

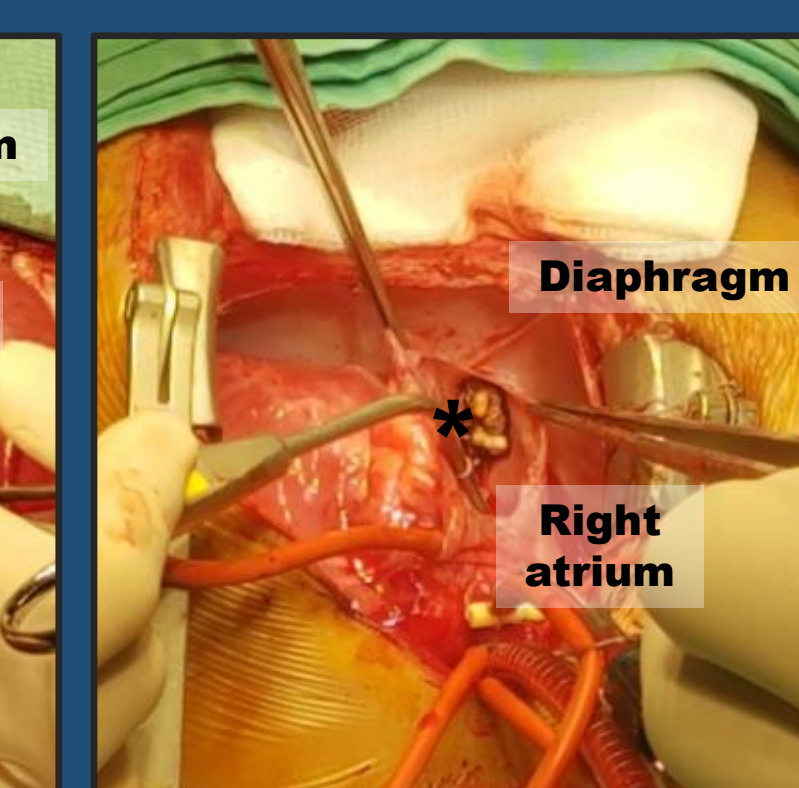


Figure 6 (Thoracic view): Tumor thrombus (\*) seen in the right atrium

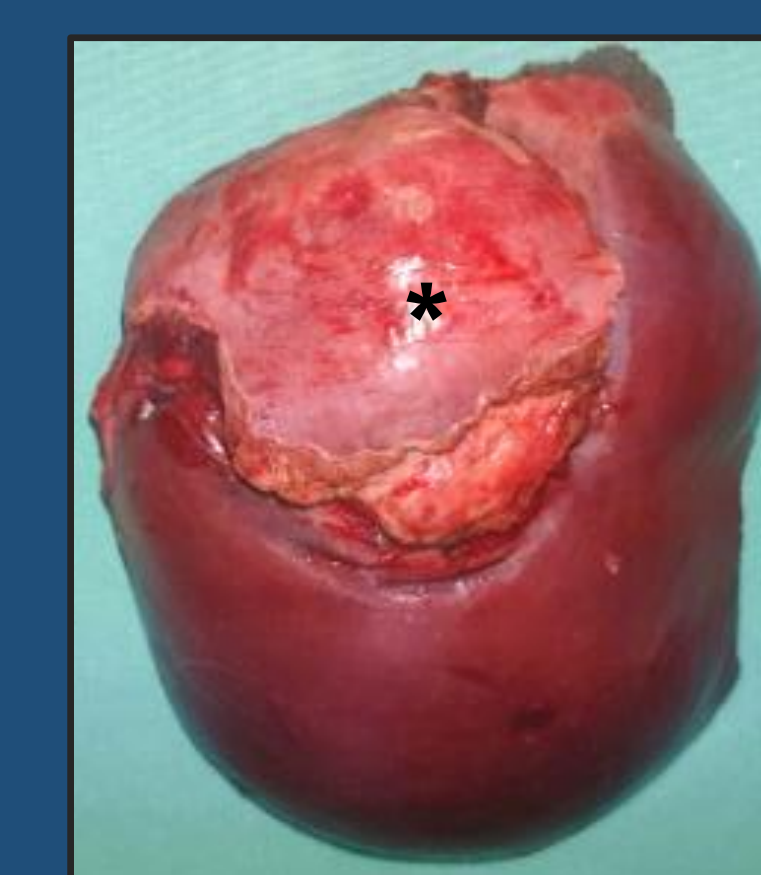


Figure 7 (Anterior surface): Excised right lobe of the liver along with the tumor (\*)

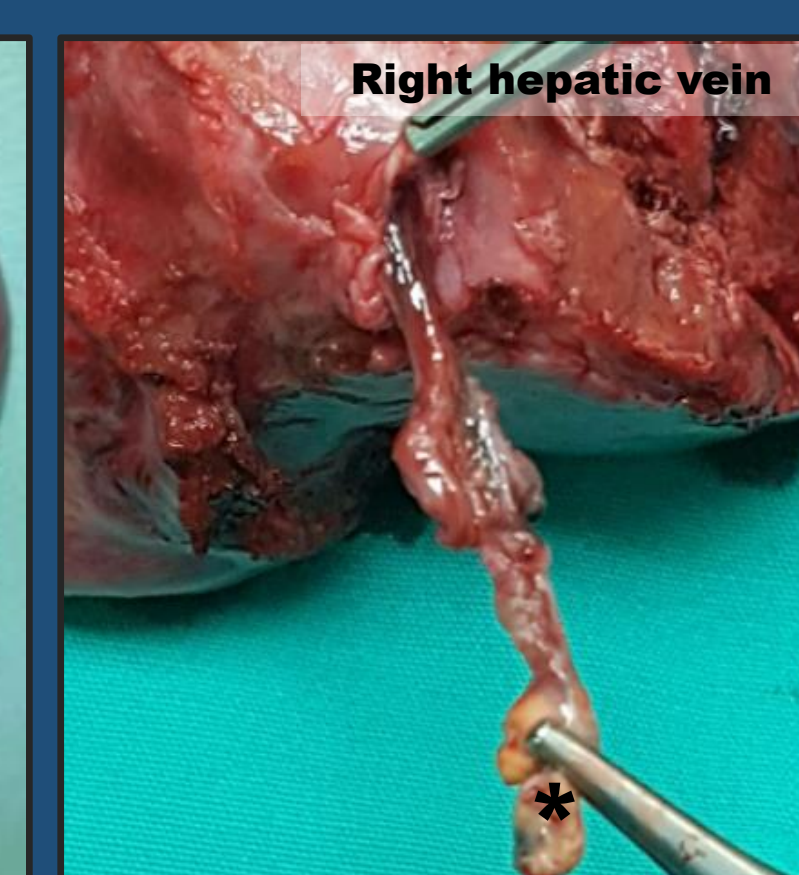


Figure 8 (Posterior surface): Tumor thrombus (\*) within the right hepatic vein

## References

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