

Adult Hepatoblastoma

What do we know?

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الاورام الكبدية في الكبار ماذا نعرف؟

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HEPATOBLASTOMA (HB), DESPITE BEING rare, is the most common malignant hepatic tumour in children. It is debatable whether adult HB occurs as an entity. Several features of the tumours described in adults are strikingly different from HB in the paediatric age group.¹

In the February 2015 issue of SQUMJ, Rabah *et al.* described Oman's experience of HB over the last 21 years.² HB is extremely rare in adults aged between 19 and 84 years, with only 40 reported cases of adult HB from 1958 to 2013.^{3,4} It is presumed that the majority of adult HB cases are, in fact, misdiagnosed hepatocellular carcinoma (HCC), combined HCC-cholangiocarcinoma or carcinosarcoma.⁵

Adult HB usually presents with abdominal pain, weight loss and a palpable liver mass on physical examination. Similarly, Rabah *et al.* found that the main symptom in the Omani children with HB was a palpable mass in the abdomen.² HB affects both hepatic lobes and commonly presents as a unifocal large mass ranging from 5–25 cm.^{3,4}

According to Rougemont *et al.*, liver fibrosis or cirrhosis was diagnosed in 30% of reported adult HB cases, in contrast to paediatric HB which appeared nearly always in children with no primary hepatic disease.¹ The main differential diagnosis of adult HB is HCC.¹ Unfortunately, there are no typical features of HB that are visible on ultrasound, computed tomography (CT) or magnetic resonance imaging (MRI) of the liver. Kishimoto *et al.* suggested that intramural classification is helpful in the diagnosis of HB.⁶ Other radiological features suggestive of HB are hypervascularity and cystic changes. However, none of the previously mentioned radiological features are specific for HB and all of these radiological features can be found in all types of liver tumours.

Postoperative pathological examination is the gold standard for diagnosing HB. HB and HCC usually display remarkable gross, histological and immunohistochemistry similarities.^{1,6} In addition, mixed features of HB and HCC can coexist in one tumour,⁷ and successive occurrence of HB and HCC in the same patient has been described.^{8,9} The use of chemotherapy in HB may lead to architectural and cytological changes resembling HCC.¹⁰ Another differential diagnosis of HB in an adult would be the coexistence of hepatocellular-cholangiocarcinoma with stem cell characteristics. Ishak *et al.* proposed a histological classification that classifies HB into epithelial and mixed epithelial types, which are most common, and the mesenchymal type, which is less common.¹¹

Molecular progress has enabled the recognition of different gene expression patterns in liver tumours.¹² Molecular overlap and coexistence of HB and HCC in adult cases support the hypothesis that these tumours may originate from a common progenitor cell.¹²

Surgical resection, if feasible, remains the main method of HB management. Neoadjuvant chemotherapy has been used to shrink the tumour, prevent intraoperative blood loss and delineate the tumour from the surrounding tissue, therefore facilitating easy resection.¹³ Rabah *et al.* used three to four cycles of neoadjuvant chemotherapy in their study, followed by surgical resection in patients with Pretreatment Extent of Disease stage II, III and IV who were deemed to have resectable disease.²

Postoperative chemotherapy was also used by Rabah *et al.* as two to five cycles of chemotherapy were given to 11 out of 15 patients.² Other local ablative procedures used in the treatment of HCC, such as ethanol ablation and radiofrequency ablation (RFA), have been tried mainly for the treatment of recurrent

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tumours post surgical resection.^{14,15} Randomised trials comparing local ablative therapy, such as RFA, versus surgical resection for a small tumour of less than 3 cm are required to assess the efficacy of such treatments in HB. Other methods used in the treatment of advanced HCC, such as transarterial chemotherapy and transarterial radioembolisation, need to be evaluated, especially in adults with advanced HB who are not surgical candidates.

Based on paediatric experience, liver transplantation has been associated with marked survival improvement, especially in those with an unresectable tumour. One of the 15 children described by Rabah *et al.* had a liver transplantation due to multi-lobar involvement.² Generally, the Milan criteria are used to assess which patients with HCC are suitable for liver transplantation.¹⁶ These criteria, which are also valued for assessing transplantation in HB patients, need further investigation. The survival rate of children who receive transplants due to HB is close to 80%. This percentage is based on results from the International Society of Paediatric Oncology and a review of the global experience of HB and liver transplantation.¹⁷ Rabah *et al.* presented a survival rate of 91% in their study, despite many obstacles faced by the treating paediatricians.² This is comparable to other recently published studies.^{18–20}

HB prognosis has improved since 1995 and this is most likely due to early diagnosis, improvement in surgical techniques, postoperative care and the use of neo-adjuvant and adjuvant chemotherapy.^{1,3}

In conclusion, the results of Rabah *et al.*'s study and the experience gained over many years should guide oncologists in treating patients diagnosed with HB and encourage all parents of such paediatric patients to address their treatment as soon as possible. In addition, the application of paediatric HB protocols in the treatment of adult HB may prove beneficial.

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