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Pediatric Hepatoblastoma: A Case Report and Review of the Literature

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Abstract

Introduction and Objective: Hepatoblastoma is the most frequent malignant embryonal liver tumor, and its incidence is increasing. Children under 3 years of age are particularly affected. The etiology is largely unknown, but a genetic predisposition exists. This case highlights the clinical and imaging characteristics and management strategies. **Case Report:** We report the case of an 18 months old male admitted for exploration of an abdominal mass and was diagnosed with hepatoblastoma. **Conclusion:** Hepatoblastoma is an aggressive tumor that begins locally and eventually metastasizes to the brain, lungs, lymph nodes, and other organs. Tumor stage at presentation and resectability are the most significant prognostic factors.

Keywords

Hepatoblastoma, Liver, Children, Alpha-Fetoprotein

1. Introduction

In children, hepatoblastoma (HB) is the most prevalent primary liver tumor [1]. Though it accounts for only 1% of childhood malignancies, the incidence of this tumor is 1.5 cases/million population annually, and it has been rising over the past 30 years. Its incidence has increased by as much as 2.7% per year [2].

The frequency of diagnosis peaks in infancy and rapidly drops in the years that follow, with few cases reported after the age of 5. This tumor is usually sporadic; however, it can exhibit symptoms of a syndrome [3]. It often manifests as an abdominal mass with elevated alpha-fetoprotein levels. In general, distant metastasis takes place in the lungs. The two main groups of HB are mesenchymal and epithelial types. For the diagnosis of various HB, histomorphology and im-

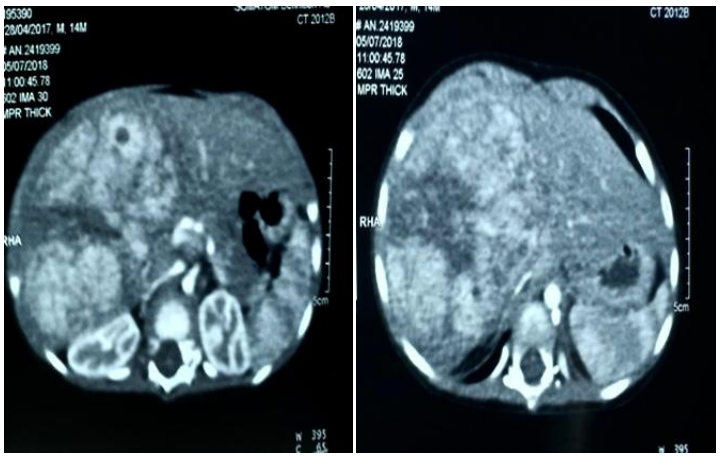
munohistochemistry are crucial [3].

Chemotherapy is a significant factor in boosting the number of tumors that can be surgically removed. Surgical resection remains the primary treatment. For those with tumors that are surgically resectable, the prognosis is favourable. The purpose of this report is to review our experience with children diagnosed with Hepatoblastoma.

2. Clinical Observation

We report the case of an 18 months male, who was born weighing 3600 grams after a 37-week of amenorrhea. The child was admitted for the management of an abdominal distension linked to postprandial vomiting, all of which occurred in the context of an alteration of the general state characterised by anorexia, asthenia, and weight loss. The patient's clinical examination revealed a distended but soupy abdomen, the presence of a palpable mass that was dependent on a right hypochondrium that was not excessively painful on palpation, and no other abnormalities that could be detected during the examination. An abdominal X-ray was performed to identify a suspected hepatic mass; additional testing using Abdominal CT (**Figure 1**) revealed a voluminous, macrolobulated, well-limited hepatic mass hypodense, consisting of four contiguous segments that were rehaussed after a contrast product injection. The segments measure 115 × 63 mm and extend 82 mm, with invasion of the portal branch.

On a biological level, thrombocytosis of 1,000,000/mm^3 and an eleveated level of alpha-fetoprotein (20,000 ng/ml) were observed in the hemogram. Due to the strong suspicion of a malignant cause, hepatic biopsy was required, with studies on immunohistochemistry and anatomopathology are being conducted to identify morphological features that are consistent with Hepatoblastoma. An extension assessment was performed based on a cerebral-cervical-thoracic-abdominal and pelvic CT scan without secondary localization, and it turned up nothing abnormal.



(a) (b)

Figure 1. Scan images of Hepatoblastoma.

The patient was assigned to hepatoblastoma group III according to PRETEXT classification, and was placed under neoadjuvant chemotherapy based on a cisplatine and carboplatine-doxorubicine every 15 days in accordance with SIOPEL protocol. After seven rounds of neoadjuvant chemotherapy administered, the control imaging observed a reduction of around 40% in the hepatic tumor's size in comparison to its initial size.

The patient had surgery, including a hepatectomy involving segments 5 - 8, a cholecystectomy anterograde, and a piece of segments 1 and 4 with the right branch portal. The initial tumor was controlled, and two more rounds of chemotherapy completed the course of treatment.

The evolution was characterized by the installation of a jaundice with septic state signs at J 10 post-operatively, for which imaging revealed the presence of a biliome at the level of the hepatectomy tranche. For this reason, the child was placed under appropriate intravenous antibiotics and monitored in the operating room for potential drainage.

Throughout the clinical evaluation process: child with jaundice, an abdominal distension with splenomegaly, asthenic and anorexic. According to the biological tests, the AFP rate was negative at 1.52 ng/ml, the prothrombin time (PT) was 60%, platelet count was at 45,000/mm^3 with elevated transaminases. A radiological examination revealed signs of portal hypertension, including homogenous splenomegaly and peri-gastric varices. A fibroscopy oesogastroduodenal (FOGD) reveled a esophageal varices with treatment based on avlocardyl, vitamin K and ur-sodeoxychol acid.

The child was admitted to the hospital multiple times to treat a hemorragic syndrome involving gingivitis and hemorrhagic seizures. He received multiple transfusions including platelet-rich plasma, globulaires, fresh frozen plasma, albumin perfusions, intravenous vitamin K, and an indication of possible hepatic graft.

3. Discussion

An uncommon malignant liver embryonal tumor known as hepatoblastoma (HB) typically affects young infants, with a median diagnosis age of 16 months. Hepatoblastoma is the most frequent type of liver cancer in children, accounting for 1% of all new cancer diagnoses in this age group. Hepatoblastoma is thought to affect 0.5 to 1.5 cases out of every million children annually [4]. It is the young child's tumor. In the primary series described in the literature, the typical age at diagnosis ranges from 12 to 21 months, and more than 80% of cases happen before the age of two. Although occurrences in teens and young adults have also been reported, some cases are diagnosed prenatally [5] correlation between HB and developmental syndromes like Beckwith-Wiedemann Syndrome (BWS) and Familial Adenomatous Polyposis (FAP) [6]. That was absent in our case report, It's also a significant risk factor that may be present in up to 10% of cases and is currently driving the recommendation that all children with hepatoblastoma [7].

Risk factors like prematurity and extremely low birth weight (<1500 g) have been described [7].

There are histological subtypes of hepatoblastoma. These include a variety of transitional, small- and large-cell undifferentiated tumor forms; tumors with mixed (epithelial and mesenchymal characteristics); and fully epithelial tumors with pure foetal or mixed foetal/embryonal histology [8]. This varied tumor spectrum suggests that HB has a developmental origin and may be responsible for the variance in their clinical behaviour. It also seems to represent different patterns of embryonal hepatic development and maturation [8].

Abdominal mass or distension is the most typical symptom. Due to tumor distension or subsequent anemia, some children present with stomach discomfort, widespread weariness, and appetite loss. Severe anemia, peritoneal irritation symptoms, and vomiting are common presentations in children with burst tumors [9]. In some instances, the tumor secretes β-human chorionic gonadotropin (β HCG), which causes early puberty and virilization. The most significant clinical indicator of HB is serum alpha-fetoprotein (AFP), which is also the primary indicator of malignant transformation, response to therapy, and recurrence [9].

In our study, AFP value was found to be elevated and was highly advantageous and significant as it could be used for follow up. In addition to confirming the mass's intrahepatic location, imaging must also reveal the tumor's operability and pinpoint any secondary locations [7]. A big mass in the liver is typically shown on abdominal ultrasonography; occasionally, the tumor also has satellite lesions and areas of internal bleeding. Multiphase computed tomography (CT) or magnetic resonance imaging (MRI) is the most helpful diagnostic modality. A malignant liver tumor is strongly suggested by helical CT findings of hypervascular lesions in the liver with delayed contrast excretion.

The International Childhood Liver Tumor Strategy Group (SIOPEL) developed the PRETEXT approach to stage and stratify liver tumors based on risk [10]. Prior to starting medication, PRETEXT is used to characterize the extent of the tumor. It is the foundation for risk stratification in the ongoing SIOPEL hepatoblastoma investigations and has strong interobserver repeatability and prognostic relevance in children with the disease. Even though PRETEXT is not their primary staging approach, the majority of other study groups now use it to explain imaging abnormalities at diagnosis [10].

Although some researchers agree that a biopsy may not be required for young children (6 months to 3 years) with an extremely high AFP level [5], histological diagnosis of a tumor specimen is crucial. Furthermore, delaying a biopsy theoretically lowers the risks of tumor seeding or dissemination. Children's liver tumors should only be treated when a biopsy specimen has been definitively diagnosed, unless there are immediate life-threatening situations, such as tumor invasion of the right atrium or tumor rupture, according to the Japanese Study Group for Pediatric Liver Tumors (JPLT) [9]. When determining the dosage of

chemotherapy and the possibility of surgery, segmental evaluation of the tumor's size and connection to the major hepatic arteries is crucial [9].

Combining surgery and chemotherapy could increase the percentage of patients with hepatoblastoma who have tumor-free survival to 75% of all cases. For possibly resectable (SR, standard risk) cancers, this percentage rises to 90%. High risk (HR) hepatoblastomas nevertheless have a poor prognosis despite having substantial vascular invasion, extrahepatic extension, multifocally disseminated development in the liver, and metastases [11]. This is because these tumors frequently quickly develop resistance against cytotoxic medicines. Each treatment cycle was followed by an assessment of acute toxicity. During follow-up, assessment of ototoxicity and cardiac toxicity was also necessary. Prior to beginning treatment, it was necessary to establish baseline cardiac, renal, hepatic, and audiological parameters that were normal [12].

Since the 1990s, patients with HB generally had a markedly better prognosis because of advanced surgical methods and platinum-based chemotherapy. Between 50% and 100% of people worldwide will survive five years following HB, depending on the severity of the disease and the histological subtype. Almost every patient who has a tumor surgically removed during the diagnosing stage has a long-lasting remission. For the majority of patients, chemotherapy is typically sufficient to treat distant metastases [13].

4. Conclusion

Hepatoblastoma is the most common malignant liver tumor in children. A measurement of Alfa fetoprotein and imaging are employed in the diagnosis. Chemotherapy and surgery are the main forms of treatment; the prognosis is good if it is completely cured.

Consent

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images.

Author Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflict of interest.

References

[1] Herzog, C.E., Andrassy, R.J. and Eftekhari, F. (2000) Childhood Cancers: Hepatoblastoma. *The Oncologist*, **5**, 445-453. https://doi.org/10.1634/theoncologist.5-6-445

[2] Kahla, J.A., Siegel, D.A., Dai, S.F., Lupo, P.J., Foster, J.H., Scheurer, M.E. and Heczey, A.A. (2022) Incidence and 5-Year Survival of Children and Adolescents with Hepatoblastoma in the United States. *Pediatric Blood Cancer*, **69**, e29763.

https://doi.org/10.1002/pbc.29763

[3] Bharti, S., Bharti, J.N., Sinha, A. and Yadav, T. (2021) Common and Rare Histological Variants of Hepatoblastoma in Children: A Pathological Diagnosis and Review of the Literature. *Gastrointest Tumors*, **8**, 41-46. https://doi.org/10.1159/000512236

[4] Lucas, B., Ravishankar, S. and Pateva, I. (2021) Pediatric Primary Hepatic Tumors: Diagnostic Considerations. *Diagnostics* (*Basel*), **11**, 333. https://doi.org/10.3390/diagnostics11020333

[5] Brugières, L., Branchereau, S. and Laithier, V. (2012) Paediatric Malignant Liver Tumours. *Bulletin du Cancer*, **99**, 219-228. https://doi.org/10.1684/bdc.2011.1539

[6] Heck, J.E., Meyers, T.J., Lombardi, C., Park, A.S., Cockburn, M., Reynolds, P. and Ritz, B. (2013) Case-Control Study of Birth Characteristics and the Risk of Hepatoblastoma. *Cancer Epidemiology*, **37**, 390-395. https://doi.org/10.1016/j.canep.2013.03.004

[7] Purcell, R., Childs, M., Maibach, R., Miles, C., Turner, C., Zimmermann, A., Czauderna, P. and Sullivan, M. (2012) Potential Biomarkers for Hepatoblastoma: Results from the SIOPEL-3 Study. *European Journal of Cancer*, **48**, 1853-1859. https://doi.org/10.1016/j.ejca.2011.10.019

[8] Taque, S., Buendia, M.A., Fabre, M., Branchereau, S., Pariente, D., Bruneau, B. and Brugières, L. (2013) What We Have to Know about Paediatrics Hepatoblastoma. *Revue d'Oncologie Hématologie Pédiatrique*, **1**, 111-119. https://doi.org/10.1016/j.oncohp.2013.10.003

[9] Hiyama, E. (2014) Pediatric Hepatoblastoma: Diagnosis and Treatment. *Translational Pediatrics*, **3**, 293-299.

[10] Roebuck, D.J., Aronson, D., Clapuyt, P., Czauderna, P., de Ville de Goyet, J., Gauthier, F., MacKinlay, G., Maibach, R., McHugh, K., Olsen, Y.E., Otte, J.-B., Pariente, D., Plaschkes, J., Childs, M. and Perilongo, G. (2007) 2005 PRETEXT: A Revised Staging System for Primary Malignant Liver Tumours of Childhood Developed by the SIOPEL Group. *Pediatric Radiology*, **37**, 123-132. https://doi.org/10.1007/s00247-006-0361-5

[11] Häberle, B., Bode, U. and von Schweinitz, D. (2003) Differentiated Treatment Protocols for High- and Standard-Risk Hepatoblastoma—An Interim Report of the German Liver Tumor Study HB99. *Klinische Padiatrie*, **215**, 159-165.

[12] Zsiros, J., Brugieres, L., Brock, P., Roebuck, D., Maibach, R., Zimmermann, A., *et al.* (2013) Dose-Dense Cisplatin-Based Chemotherapy and Surgery for Children with High-Risk Hepatoblastoma (SIOPEL-4): A Prospective, Single-Arm, Feasibility Study. *The Lancet Oncology*, **14**, 834-842. https://doi.org/10.1016/S1470-2045(13)70272-9

[13] Pihlajoki, M., Eloranta, K., Nousiainen, R., Väyryne, V., Soini, T., Kyrönlahti, A., Parkkila, S., Kanerva, J., Wilson, D.B., Pakarinen, M.P. and Heikinheimo, M. (2023) Biology of Childhcood Hepatoblastoma and the Search for Novel Treatments. *Advances in Biological Regulation*, **91**, 100997. https://doi.org/10.1016/j.jbior.2023.100997