

Adulthood hepatoblastoma

To the Editor,

Hepatoblastoma is one of the most common malignant hepatic tumors to be diagnosed in children. On the other hand, it is rare and associated with poor prognosis in adults (1). Here, we present a case of an adult who was diagnosed with hepatoblastoma, and has been followed up for 5 years, during which time she remained alive.

An 18-year-old female patient was admitted to our clinic following detection of hepatocellular carcinoma in her liver biopsy. Alpha-fetoprotein (AFP) was 4646 ng/mL and other tumor markers and laboratory parameters were within the normal ranges. In a thoraco-abdominal computerized tomography examination, a 4.5x5.5 cm lesion was detected in the liver (Figure 1). A hepatic left segmentectomy was then performed and histopathological examination revealed an epithelial hepatoblastoma (fetal embryonic) (Figure 2). After the operation, 6 cycles of cisplatin and adriamycin chemotherapy were administered. Within the first year of follow-up, a recurrent lesion was detected in the liver. The patient underwent mass excision and the subsequently received 6 cycles of vincristine, cisplatin and 5-FU chemotherapy following the operation. In the second year of follow-up, a recurrent lesion in the liver and a thrombus in the inferior vena cava were detected. A decision was made to perform liver transplantation. Due to the lack of a donor, however, mass excision, diaphragm resection, inferior vena cava resection and vein grafting were performed instead. Six cycles of iphosphamide, cisplatin and etoposide chemotherapy were administered to the patient after surgery. The patient, who is within the fifth year of her first diagnosis, continues to be followedup in the post-operative period. Informed consent was taken from the patient about this publication.

Among hepatoblastoma cases, 95% are aged below 5 years (2). It presents very rarely during adolescence and

early adulthood (1). Therapeutic regimens are based on chemotherapy and total excision of the tumor (3). While anthracycline-containing regimens have been used preferentially as chemotherapeutic agents, most recent studies have highlighted that cisplatin-based regimens without anthracycline were as efficient as those including anthracycline (4). In patients with resectable tumors, 5-year survival is approximately 80%

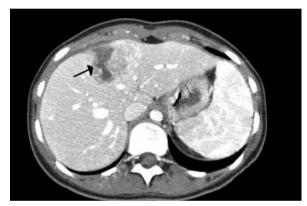


Figure 1. An axial CT section, in the segment 4A-8 junction, shows a massive lesion with peripheral pseudocapsular staining at a late phase with dimensions of 4.5x5.5 cm, peripheral localization, an hypervascular periphery in the arterial phase and an apparently necrotic central cut.

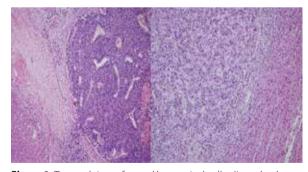


Figure 2. Tumoral tissue formed by atypical cells aligned as layers or trabeculae, with narrow, clear or granular cytoplasms, similar to fetal hepatocytes, in hepatic tissue. Hepatoblastoma (Fetal epithelial pattern). X100, H&E.

Address for Correspondence: Tülay Akman, Department of Medical Oncology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey E-mail: tulaytuzel@gmail.com

© Copyright 2015 by The Turkish Society of Gastroenterology • Available online at www.turkjgastroenterol.org • DOI: 10.5152/tjg.2015.6544

(5). Recent advances in neoadjuvant chemotherapy have prolonged the survival rates for unresectable tumors (6). While more than 60% of unresectable tumors become resectable, 20% of cases remain unresectable despite chemotherapeutic intervention. In these patients, liver transplantation is the treatment of choice (7).

Although hepatoblastoma is rare and life expectancy is short in adult patients, survival rates can be increased with effective surgical procedures and chemotherapy.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author contributions: Concept - T.A., M.O.; Design - T.A., M.O., I.U., T.U.; Supervision - T.A., T.Y., A.Y.; Resource - T.A., I.O., A.Y.; Materials - T.A., O.S., Data Collection&/or Processing - T.A., T.Y., A.Y.; Analysis&/or Interpretation - T.A., M.O., I.O.; Literature Search - T.A., M.O.; Writing - T.A., M.O., Critical Reviews - T.A., A.Y., I.O.

Conflict of Interest: No conflict of interest was declared by the authors. **Financial Disclosure:** The authors declared that this study has received no financial support.

Tülay Akman¹, Mehmet Asi Oktan², İlkay Tuğba Unek¹, Tarkan Unek³, İlhan Öztop¹, Tuğba Yavuzsen¹, Ahmet Uğur Yılmaz¹, Özgül Sağol⁴

¹Department of Medical Oncology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

²Department of Internal Diseases, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

³Department of General Surgery, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

⁴Department of Pathology, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

REFERENCES

- 1. Altmann HW. Epithelial and mixed hepatoblastoma in the adult. Histological observations and general considerations. Path Res Pract 1992; 188: 16-26. [CrossRef]
- Ortega JA, Krailo MD, Haas JE, et al. Effective treatment of unresectable or metastatic hepatoblastoma with cisplatinand continuous infusion doxorubicin chemotherapy:a report from the Children's Cancer Study Group. J Clin Oncol 1991; 9: 2167-76.
- 3. Stringer MD. The role of liver transplantation in the management of paediatric liver tumours. Ann R Coll Surg Engl 2007; 89: 12–21. [CrossRef]
- Perilongo G, Maibach R, Shafford E, et al. Cisplatin plus doxorubicinfor standard-risk hepatoblastoma. N Engl J Med 2009; 361: 1662-70. [CrossRef]
- Otte JB, de Ville de Goyet J, Reding R. Liver transplantation for hepatoblastoma: indications and contraindications in the modern era. Pediatr Transplant 2005; 9: 557-65. [CrossRef]
- Langevin AM, Pierro A, Liu P, Filler RM, Greenberg ML. Adriamycin and cis-platinum administered by continuous infusion preoperatively in hepatoblastoma unresectable presentation. Med Pediatr Oncoll 1990; 18: 181-4. [CrossRef]
- Brown J, Perilong G, Shafford E, et al. Pretreatment prognostic factors for children with hepatoblastoma—results from the International Society of Pediatric Oncology (SIOP) study SIOPEL 1. Eur J Cancer 2000; 36: 1418-25. [CrossRef]