Identifying a new type of liver tumor

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Investigators at Children's Hospital Los Angeles have succeeded in better defining a rare pediatric malignant liver disease—a necessary step in achieving an optimum treatment.

The research into hepatocellular malignant neoplasm-NOS (HEMNOS) is the largest case study of its kind to date. HEMNOS is a recently described entity that has features of the most common pediatric liver cancer—hepatoblastoma (HB), and hepatocellular carcinoma (HCC)—a liver cancer more commonly seen in adults.

While HEMNOS typically has been treated much like HCC, the CHLA investigators found that treating HEMNOS like a subtype of hepatoblastoma yielded more positive treatment outcomes in children.

The researchers analyzed 11 patients (ages four to 15) with liver cancer, originally seen from 2000-2016, all of whom received HB-targeted chemotherapy and complete surgical resection. HEMNOS had been thought to have a very poor prognosis, an impression created by the single previous case series on this topic. However, all of the CHLA patients survived and achieved remission, despite being considered high risk.

"The high survival rate of our patients further supports our argument that HEMNOS is best considered a subtype of hepatoblastoma," said Leo Mascarenhas, MD, MS, deputy director of the Children's Center for Cancer and Blood Diseases and senior author on the study. "Without such a clear definition of the tumor, patients may be treated

inappropriately."

The investigators performed a series of analyses to better define this unique group of tumors. Their pathology review of pre-chemotherapy specimens showed that six tumors had overlapping tissue features of HB and HCC, four had predominant HB histology, along with focal HCC-like histology and one had HB histology. "No matter what their original diagnosis, each patient benefited when HEMNOS was treated like hepatoblastoma," said Shengmei Zhou, MD, a pediatric pathologist at CHLA and lead author on the study.

Investigators also looked for the expression of the telomerase gene (TERT), a repetitive DNA-protein complex crucial for the survival of cancer cells. They identified TERT expression in nine of the 11 cases.

In their next step, the researchers plan to study the genomic difference of the HEMNOS tumor compared to classic hepatoblastoma and hepatocellular carcinoma to gain new insights into the pathophysiology of this cancer.

The study was published online in the journal *Histopathology*, in June 2017, in advance of print publication.

More information: Shengmei Zhou et al, Hepatocellular Malignant Neoplasm-NOS: A Clinicopathologic Study of 11 Cases from a Single Institution, *Histopathology* (2017). DOI: 10.1111/his.13297

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