Hepatoblastoma with Glomerulocystic Disease – a Mere Coincidence or an Association?

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Dear Sir,

Over the past few years, glomerulocystic disease of the kidney (GCK) has become a specific entity, distinct from the other types of cystic diseases. Various associations between GCK and extrarenal lesions including cysts in other organs, patent ductus arteriosus, congenital heart disease, hydrocephalus, enlarged thyroid and retinitis pigmentosa have been reported in the literature [1]. With particular reference to liver lesions in GCK, the known associations are hepatic cysts, congenital hepatic fibrosis and hepatocellular adenoma [2, 3]. We report here a coexistence of hepatoblastoma and GCK in a 3-month-old child and discuss the possibility of an addition to the already known associations between liver lesions and GCK.

A 3-month-old male child born at full term by normal vaginal delivery presented with a mass in the right hypochondrium extending to the umbilical region. There was no history of fever, jaundice, hematuria or gastrointestinal manifestations. Examination revealed firm and nontender hepatosplenomegaly and an abdominal mass 7×11 cm with irregular surface and well-defined margins. The ultrasound and CT scan suggested a mass arising from the liver. The child's progress worsened with a hospital-acquired respiratory infection and died.

At autopsy, there was a 6 × 5 × 4 cm well-circumscribed tumor in the left lobe of the liver (fig. 1) with extensive hemorrhage and necrosis. The kidneys were normal in size with subcapsular pin head sized cysts which on cut surface were seen confined to the cortex. The lungs showed focal bronchopneumonia and the brain showed subarachnoid hemorrhage. No other abnormality was noted.

Histological examination of the hepatic tumor confirmed hepatoblastoma with a predominant epithelial component of embryonal and fetal hepatocytes. The kidneys showed a diffuse cystic dilation of the Bowman's space (fig. 2) with a normal glomerular tuft in most

Fig. 1. Liver showing a well-circumscribed tumor in the left lobe.

Fig. 2. Kidney showing marked dilatation of the Bowman's space with a normal glomerular tuft, a characteristic appearance of GCK. HE. × 52.

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spaces. The cysts showed a single layer of cuboidal cells with stratification and papillae formation. The tubules, interstitium and blood vessels were normal.

Cysts in the kidneys can be found in a multitude of conditions in infancy, but the present case fits with the specific entity of GCK, as the only change was dilatation of the Bowman's space.

Although this condition is said to be often discovered incidentally at autopsy [4], many cases present with renal masses and progressive renal failure [5]. The reported associations between GCK and liver lesions include only benign conditions like cysts and adenomas [3]. This patient had a hepatoplasma associated with GCK. Though one may be tempted to dismiss GCK in this patient as an incidental finding at autopsy, the presence of malignancy and the short life span not allowing the kidney lesion to manifest clinically should be considered in not undermining its significance. In this context, the accepted associations between GCK and the reported benign lesions of liver can be well explained as these patients are much older and initially manifest with kidney mass, the liver lesion being as incidental finding at autopsy. Thus, the possibility that hepatoblastoma may be an addition to the established associations between liver lesions and GCK should be strongly considered. By highlighting this hitherto unreported association, the way to discover newer associations between GCK and extrarenal abnormalities may be made.

References

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