Rare Tumors of the Abdomen in Children

CME Credits

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Medical History

This is a two years and eleven months old girl without abnormal growth and development. Preventive vaccines were administered as scheduled. The family history was not remarkable.

Her mother identified a palpable lump at her upper right quadrant abdomen. Patient had a normal appetite without symptoms such as nausea, vomiting, constipation or abdominal pain. Physical examination on admission confirmed the solid mass with dullness percussion at upper right quadrant abdomen.

Blood chemistries three days later showed leukocytes 8890 per uL, C-reactive protein 1.7 mg/dL, aspartate aminotransferase 29 IU/L, Alanine aminotransferase 11 IU/L, alkaline phosphatase 149 IU/L, Gamma-glutamyl transferase 14 IU/L, total bilirubin 0.3 mg/dL, lactate dehydrogenase 399 IU/L, blood urea nitrogen 8 mg/dL, creatinine 0.26 mg/dL, calcium 9.9 mg/dL, phosphate 3.8 mg/dL and alpha-fetoprotein 821 ng/mL.

Abdomen x-ray [Figure 1] revealed an irradiation impenetrable mass in upper right abdominal cavity. Sonography [Figure 2] showed a heterogeneous tumor 12 centimeters in size in upper right abdominal cavity. Abdominal computed tomography [Figure 3a and 3b] confirmed the huge calcified tumor with heterogeneous density between the liver and the right kidney. The differential diagnosis includes hepatoblastoma, neuroblastoma, pheochromocytoma, and tumor of the adrenal cortex. Postoperative pathological findings confirmed the adrenocortical carcinoma. Patient received subsequent chemotherapy and radiation therapy.

Discussion

Liver tumors are classified as benign and malignant. The malignant tumors are mainly from the liver cells. The main pediatric liver tumors include hepatoblastoma and hepatocellular carcinoma. Hepatoblastoma is the most common primary liver tumor in children. [1] The statistics from the Taiwan Children's Cancer Foundation show that the incidence rate of childhood cancer is about 1.8%, mostly in infants and young children less than six years of age. The average age of onset is 1 year 8 months old. The incidence of male and female accounts for about 1–2:1.

Hepatocellular carcinoma were uncommon in children. The clinical symptoms and pathological features of pediatric hepatocellular carcinoma are similar to those of the adult. The statistics from the Taiwan Children's Cancer Foundation showed that the incidence rate in childhood cancers is about 1.5%, mostly in children over 6 years old. The incidence of male and female accounts for about 3-4:1.

Liver cancers are common in Asia and Africa but are relatively rare in the Europe and the United States. The relatively higher rate of liver cancers in Taiwan is correlated with higher prevalence of viral hepatitis B. Hepatitis B vaccination program has significantly reduce the incidence of hepatocellular carcinoma in Taiwan.

The statistics from the Taiwan Ministry of Health and Welfare show about 13,000 people die each year from chronic liver disease, cirrhosis and liver cancer. Chronic liver disease and cirrhosis are the ninth leading causes of death; and liver cancer is the second leading causes of cancer death in Taiwan.[2] The main causes of chronic liver disease, cirrhosis and liver cancer in Taiwanese are viral hepatitis B (70%) and C (20%). For hepatitis B surface antigen bearing carriers with active hepatitis; about 15 to 20% of them develop cirrhosis with higher incidence of liver cancer. More than 70% of patients with viral hepatitis C develop chronic hepatitis; about 20%

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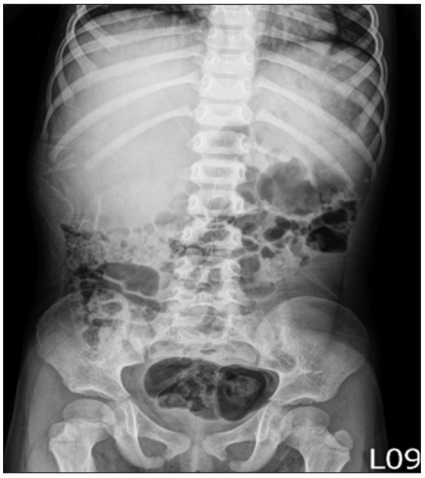

Figure 1: Abdominal X-ray revealing a visible mass of irradiation impenetrable in upper right abdominal cavity

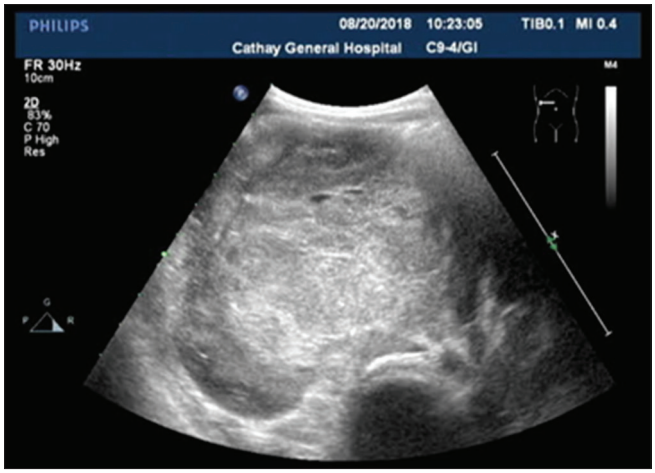

Figure 2: Abdominal ultrasound showing a huge heterogeneous tumor in the upper right abdominal cavity

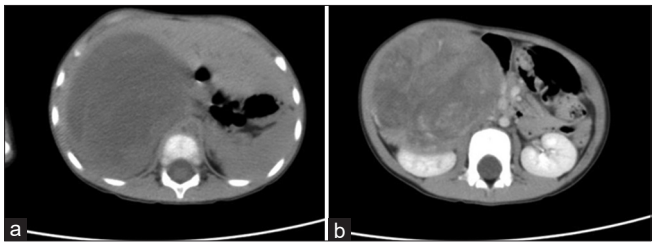

Figure 3: Non-enhancement (3a) and enhancement (3b) images of computed tomography of the abdomen showing a huge calcified tumor with heterogeneous density between the liver and the right kidney

of them progress to cirrhosis and about 3 to 5% develop liver cancer per year.

Second only to leukemia, brain tumors, and lymphomas, neuroblastoma is the fourth common solid malignant tumor among childhood cancers. Neuroblastoma originates from embryonic tumors in the neural crest cells. It caused by the poor differentiation of pluripotent sympathetic neuron mother cells and sympathetic neuroblasts. Neuroblastoma may occur in any part of the sympathetic nervous system.[3] However, Neuroblastoma most often occurs in the posterior abdominal cavity, especially in the adrenal glands. The statistics from the Taiwan Children's Cancer Foundation show about 30–40 new cases in Taiwan every year.

Pheochromocytoma and paraneuroma are catecholamine secreting tumors originating from adrenal medulla and sympathetic ganglia, respectively. Both of the two tumors have similar clinical manifestations.[4] The main treatment for pheochromocytoma is surgical resection. Pheochromocytoma may secret a large amount of catecholamine secretion during surgery, which may result in a hypertensive crisis. The goal of preoperative management is to control hypertension and to avoid hypertensive crisis.

The prevalence of the adrenocortical carcinoma is very rare. There were a total of 2,014 cases of adrenocortical carcinoma between 1973 and 2014 with an age-adjusted incidence of 1.02 per million populations. The median age at diagnosis was 55 years with the majority of them being females and whites.[5] The proportion of cases by different genders, races and age at diagnosis had not changed significantly over time. These malignancies were mostly the only primary malignancy, unilateral and of high grades at diagnosis.

The prognosis of adrenocortical carcinoma is poor. Complete surgical resection is essential for a curative approach and significantly determines overall prognosis. Tumor resection is sophisticated and complicated by the vulnerability of the tumor and its invasive growth. Chemotherapy and Mitotane are additional therapeutic approaches that are combined with surgery in an interdisciplinary strategy.[6] The studies from Italy and Germany show that adrenocortical carcinoma has a high risk of recurrence after radical resection. The multivariate analyses show that mitotane use after radical resection may improve the recurrence-free survival and prognosis.[7]

Declaration of patient consent

The author certifies that he has obtained all appropriate patient consent forms. In the form the patient's parents have given their consent for their child's images and other clinical information to be reported in the journal. The parents understand that their child's name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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