HEPATOBLASTOMA:

A Case Report

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An infant aged 8 months was recently admitted to St. Luke's Hospital with massive enlargement of the liver and abdominal distention.

M.C. was the second child of healthy unrelated parents. He was a full term normal, delivery following an uneventful pregnancy; the birth weight was 3.8 Kg. There was no diffuculty in the onest of respiration. His subsequent development was apparently normal. He was first seen at eight months of age with a three week history of persistent crying. On examination he was a well-covered, dyspnoeic child, obviously in distress with abdominal distention. The temperature was 101°F, the pulse 140 and the respiratory rate 40. The heart sounds were normal but there were diffuse medium rales in his chest. The liver was markedly enlarged, the lower border extending below the umbilicus; the lower edge was hard and irregular. There was no clinical evidence of ascites. The spleen was not palpable. The Hb was 11.0g. per c.mm.

with a normal differential. Examination of peripheral blood film showed normocytic, hypochromic erythrocytes. The fasting blood glucose was 74 mg/100ml and the glucagon tolerance test revealed a 20% rise in the fasting blood sugar at 30 and 60 minutes. The serum bilirubin was 0.6mg% with an indirect Van den Bergh's reaction; the S.G.P.T. was 2 I.U./litre and the prothrombin times 22 secs, with a control of 16 secs. The urine was normal. X-ray films of the chest showed scattered focal opacities throughout both lung fields; a plain X-ray of the abdomen and an intra-venous pyelogram were normal. The serum electrophoretic pattern showed almost complete agammoglobulnaemia with relative increase in α^1 and α_2 and a very marked increase in β-globulins; this pattern was considered to be non-specific.

His condition continued to deteriorate and he died five days after admission. The differential diagnosis included glycogen storage disease and tumour of the liver, either primary or secondary. Glycogen storage disease was excluded by the absence of hypoglycaemia, a normal response to glucagon and the absence of ketonuria. The plain X-ray of the abdomen and the intravenous pyelogram were helpful in excluding primary renal tumours and a neuroblastoma. In the light of the clinical findings of dyspnoea and rales over both lung fields, the X-ray chest was interpreted as a bronchopneumonia. The true nature of the hepatomegaly was determined at post-mortem examination.

Necropsy Findings

Gross examination. The body was that of a well-developed, well-nourished, white male infant weighing 10 Kg and showing slight central cyanosis and a markedly distended abdomen; there was eversion of the umbilicus and fullness of the epigastrium and both flanks. There was no palpable lymphadenopathy. The peritoneal cavity contained approximately 200 c.c. of straw-coloured clear fluid. The liver weighed 1260 gm (normal 254 gm); its lower border extended downwards to the level of the pelvic brim. There was a yellowish-white tumour mass within the right lobe of the liver and several smaller satellite nodules in the immediate vicinity. The entire external surface of the liver was peppered by small, haemorrhagic, often cystic lesions averaging 0.2 cm in diameter. The main tumour mass in the right lobe of the liver measured 10.0 cm in diameter. It had an irregular outline with tongues of tumour tissue invading with tongues of tumour tissue invading the surrounding, compressed parenchyma: the main mass was confluent with some of the satellite nodules. The cut surface of the main tumour mass showed areas of necrosis haemorrhage with pseudocyst formation and pools of clotted blood. The smaller tumour nodules varied in shape and size but otherwise had the same gross appearance. Inspite of their small size most of them consisted of cerotic and haemorrhage tumour tissue. The extra-hepatic biliary system was normal and there was no obstruction to the flow of bile into the duodenum.

The lungs together weighed 250 gm (normal 97 gm). Several elevated, well-circumscribed tumour nodules, averaging 1.0 cm in diameter, were seen on their external surfaces. On serial sectioning these tumour nodules were seen to occupy the immediate sub-pleural region: they consisted of haemorrhagic, necrotic tumour tissue identical in appearance to the smaller tumour nodules in the liver.

The lymphnodes in the region of the porta hepatis and the paratracheal and parabronchial lymphnodes were partly or completely replaced by tumour tissue. All the other organs were normal.

Microscopic examination. This showed predominantly an epithelial growth structurally resembling embryonic or foetal liver tissue. The tumour consisted of sheets and cords of closely packed cells, of uniform size, with hyperchromatic nuclei and a small amount of cytoplasm. The cytoplasm was more basophilic and less abundant than one sees in the normal parts of the liver. The nuclei showed coarse chromatin clumping with prominent nucleoli; the latter, however, were less acidophilic than those seen in mature liver cells. The tumour sheets and cords were arranged around large vascular channels partly lined by endothelium and partly by neoplastic

cells. Large haemorrhages within sheets of tumour cells and large areas of necrosis were seen. The cytoplasm of some of the tumour cells contained granules of bile pigment. Irregular, disorderly bile duct proliferation was a prominent feature. The stroma was of scanty or moderate amount, somewhat oedematous, but mature in form with collagen formation. The tumour was not sharply demarcated from the surrounding hepatic tissue; the latter was compressed and distorted and showed infiltration by neoplastic epithelial cells. Apart from these changes the adjacent hepatic tissue appeared normal. The metastases in the lung and in the paratracheal and porta hepatis lymphnodes showed the same histological picture. Haemorrhage and necrosis were particularly prominent in the lung lesions.

Discussion

This case presented with the typical clinical and histological picture of the hepatoblastoma. These tumours are usually highly malignant and metastasize rapidly to the liver, lungs and bones. They occur chiefly in young infants; Willis (1962) refers to seven cases from his collection whose ages ranged between 4 months and 7 years with six cases below the age of 3. Like all reported cases the main presenting feature in this case was an enlarging mass in the upper abdomen associated with an increase in the size of the abdominal girt. Pain was a factor in 45% of the cases reported by Shorter *et al* (1960) and this became particularly severe towards the terminal phase of the disease. The occurrence of pain in our case could not be determined with certainty but the infant presented with a three week history of persistent crying. There was no clinical evidence of ascites and at post-mortem only 200 c.c. of fluid were found in the peritoneal cavity. In Shorter's series ascites developed in 3 out of 11 patients and was a late finding in the course of the illness. The histological picture tallied with that of the typical hepatoblastoma and the liver parenchyma surrounding the tumour showed no indication of a possible aetiology.

References:

WILLIS R.A. (1962): The Pathology of the tumours of children. Oliver and Boyd Ltd.

SHORTER, R.G., BAGGENSTOSS, A.H., LOGAN, G.B. and HALLENBECK, G.A. (1960): Paediatrics, *25*: 191.