MALIGNANT HEPATIC TUMOUR PRESENTING AT BIRTH REPORT OF A CASE*

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Primary malignant tumours of the liver parenchyma (malignant hepatoma, hepatoblastoma) are a well recognised but rare condition. Although it is likely that most of these tumours develop during intra-uterine life very few of them are present at birth. Noeggerath[1] probably presented the first case in which a malignant hepatic tumour was present at birth in 1854; in his case the tumour caused obstruction during delivery.

Anderson,[2] reviewing 175 cases of malignant tumours in infancy and childhood, found twelve cases of primary carcinoma of the liver, but only three of these were of the hepatic cell type of carcinoma. The youngest case in her group was three years old. Tomsykoski and Stevens[3] reported a case aged nine months, and Gross[4] recorded eleven cases from the Boston Children's Hospital; five of these were under one year of age; the youngest was two days old.

Steiner[5] collected all available cases from the world literature and after careful scrutiny found 75 cases of proven primary carcinoma of the liver in children and added two further cases of his own. Only two of these 77 cases were under one month old. Bigelow and Wright[6] reviewed the literature since Steiner's paper and added a case of their own.

The presenting symptoms appear to have been abdominal swelling, loss of weight, palpation of the tumour by a parent but only rarely clinical hepatic insufficiency. The only feasible treatment of the condition is excisional surgery but the condition itself carries a very grave prognosis and the operative mortality is extremely high.

In our case the tumour was felt immediately after birth and there can thus be no doubt that the tumour had developed in utero. We feel therefore that the following case is worth reporting:

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A. K. (59-8381), a female infant, was admitted to the Hacettepe Children's Hospital on 27.6.59 within a few hours of delivery. The doctor delivering the baby at the maternity hospital had noticed the abdominal swelling immediately after birth. Pregnancy and delivery had been normal and to the best of our knowledge the mother had had no irradiation (diagnostic or therapeutic) during this pregnancy. This was the second child of healthy parents, although the first baby had died.

On examination this was a full-term female infant weighing 2840 grams who appeared healthy and normal except for the abdominal findings.

There was a mass, the size of an orange, in the left upper quadrant extending up to the costal margin and to the midline. It appeared to be fluctuant and fairly superficial; the left kidney could be palpated behind this mass and was separate from it. The mass itself appeared to be only slightly mobile but was not attached to the anterior abdominal wall. In view of the the cystic appearance of the mass the differential diagnosis of a mesenteric cyst, hepatic cyst or

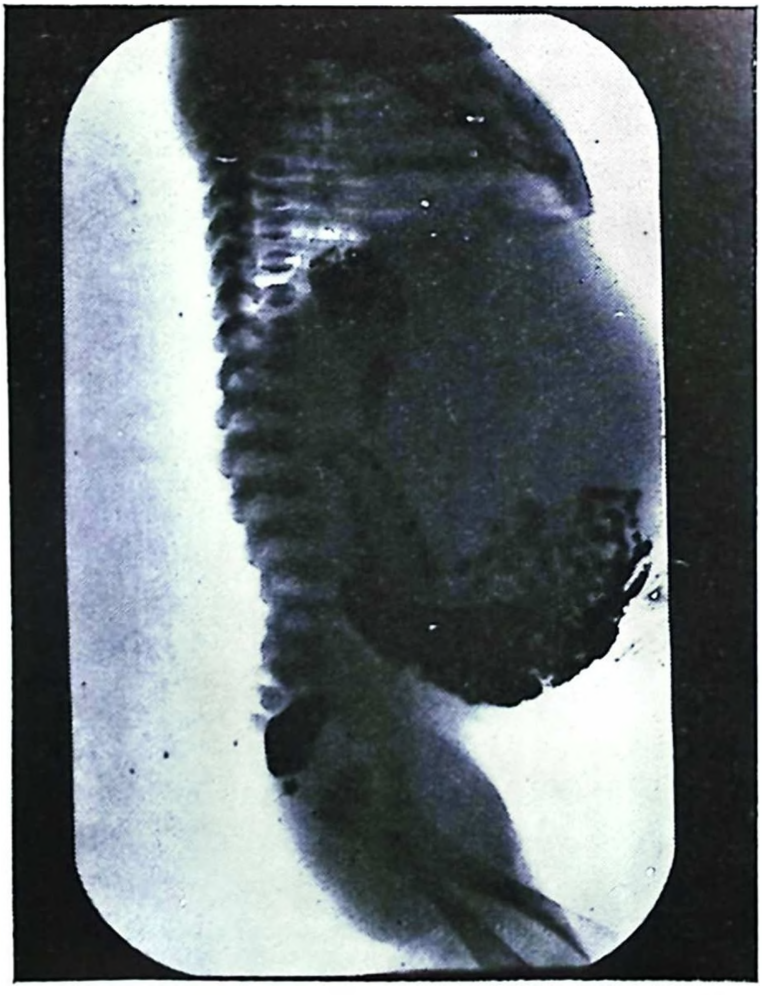

Figure 1 : Lipiodol swallow to show displacement of stomach.

possibly splenic cyst was considered. A radiograph of the abdomen showed no calcification in the area of the mass and a lipiodol swallow showed that the stomach was displaced posteriorly by the mass and smoothly indented on its lesser curve. (Fig. 1.)

After observing the child for two days preparations were made for laparotomy but unfortunately at this stage the parents withdrew their consent and removed the baby from the hospital against medical advice.

However, three days later the baby was brought back, but her general condition was much worse. Although she had not been vomiting, there was marked dehydration and the tumour had definitely increased in size. The baby, after admission, was regurgitating all feeds, and it was felt the mass was causing obstruction. Dehydration and electrolyte disturbances were therefore corrected by intravenous infusion through an indwelling polyethylene tube, and laparotomy was carried out the next day.

Operation: (H. B. E.) 4.7.59. After infiltrating the abdominal wall with 1% Procaine the abdomen was opened through a left upper rectus splitting incision, directly over the mass. There was no free fluid in the peritoneal cavity, but there was a large round tumour in the left lobe of the liver extending to the junction of the left and right lobes. The greater omentum was firmly adherent to the mass and had to be divided between ligatures. A tentative attempt to enucleate the tumour, which still appeared definitely fluctuant, was quite unsuccesful as no plane of cleavage could be found. Diagnostic aspiration resulted in a few drops of blood only. The diagnosis of malignant hepatoma was now made with some certainty and it was felt that the baby's only chance would be excision of this mass.

After division of the left triangular ligament the left lobe of the liver with the tumour could be delivered out of the abdomen and a soft intestinal clamp was placed across healthy liver tissue to the right of the tumour and the left lobe of the liver with the tumour was cut off. The raw liver edge was oversewn with two continuous catgut sutures and haemostasis appeared to be satisfactory. However, just as the abdomen was about to be closed, the baby became cyanosed and the heart ceased to beat. Artificial ventilation through an endotracheal tube and transdiapragmatic cardiac massage were unsuccessful. About 150 cc blood had been given during the operation.

At autopsy the following day (K. M.), bilateral atelectasis and bilateral pneumothorax were found. There appeared to be no hole

in the diaphragm nor was there a macroscopic rupture of the bronchi or lungs. The right lobe of the liver and the kidneys were hyperaemic. No metastases were found.

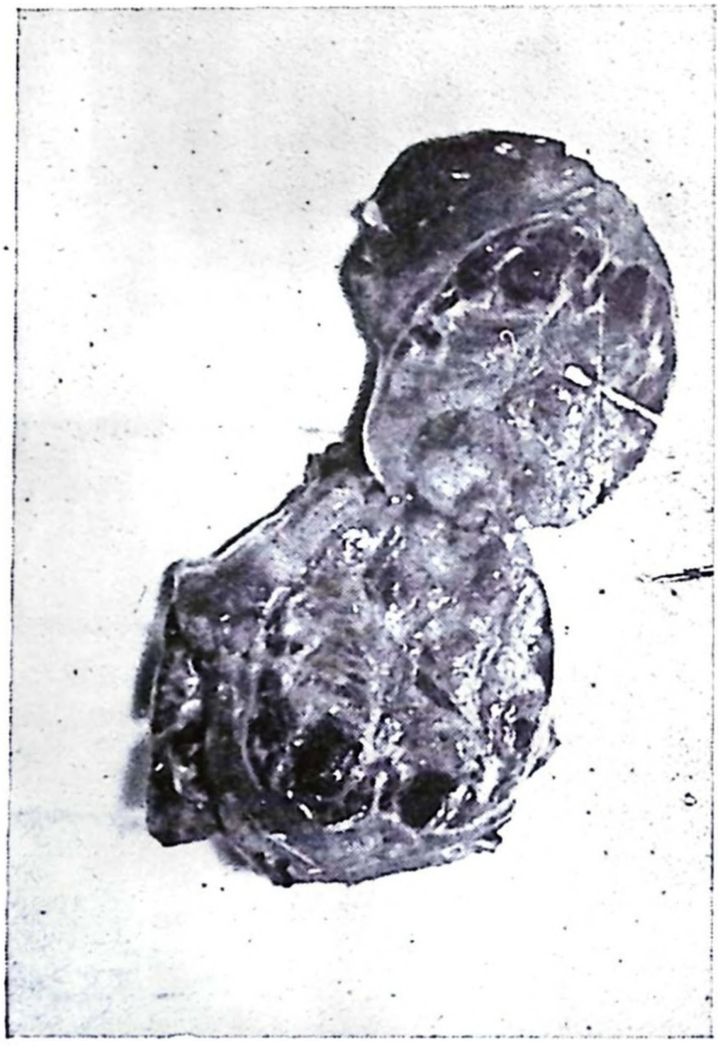

Figure 2 : Excised tumor.

The specimen was reported on by Prof. Muharrem Köksal as follows : "The specimen of resected tissue, said to have replaced the left lobe of the liver, weighed 200 grams, was approximately oval in shape, and was 7 - 8 cm. in diameter. The right border of the mass contained a row of sutures (divided greater omentum); along the left anterior surface there was a strip of tissue about 6 cm. in length, having the appearance of normal liver tissue (Fig. 2.). The tumoural mass proper was irregularly rounded, soft and well encapsulated; the capsule however seemed to include the normal appearing liver tissue mentioned above. The external surface of the tumour was greyish to yellowish-pink. The cut surface of the tumour showed several necrotic and haemorrhagic areas.

"Microscopic examination of sections which included the residual normal tissue and the tumour proper revealed no sharp distinction

nor capsule between these two but rather a gradual transition from one to the other. The liver tissue revealed intense haemorrhage as well as necrotic changes in the tissues. There was also some fatty degeneration of liver cells. The transition areas showed that tissue had been invaded by tumoural elements and in these areas of active invasion necrotic changes were predominant. Within the tumoural portions of these sections as well as others from different areas of the tumour, the histologic appearance of the tissues suggested embryonic connective tissue elements in some areas, while other areas showed myxomatous changes and still others had the appearance of angiomatous tissue. Finally, there were areas closely resembling liver tissue in cellular structure though not in architectural arrangement.

"These findings strongly suggest a diagnosis of hepatoblastoma, an embryonic liver neoplasm."

Discussion

There can be no doubt that the tumour in this baby developed in utero. The rapid increase in size between the baby's two admissions was probably due to haemorrhage into necrotic tumour tissue. Partial hepatectomy appeared to be the only possible treatment but was unfortunately unsuccessful.

The cause of the bilateral pneumotharax is difficult to explain; it could have been due to perforation of the diaphragm during mobilisation of the liver (in which case this was certainly the cause of death) or it could have been due to excessive endotracheal ventilation (in which case the pneumothorax would have been a post-mortem development). Unfortunately autopsy did not reveal the actual cause of the pneumothorax.

The diversity of the cell types in the tumour is characteristic of malignant tumours in infancy.

Summary

A case of malignant hepatoma in a new-born baby is described; attempted partial hepatectomy was unsuccesful.

We should like to thank Prof. Muharrem Köksal for the pathology report.

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