some believing in cerebrospinal fluid diversionary procedures. The treatment of choice seems to be shunting of the lateral ventricles. In the majority of cases there is free communication between the dilated 4th ventricle and the rest of the ventricular system. In the case of normal lateral and 3rd ventricles, the dilated 4th ventricle alone can be shunted. Combined shunts are suggested when a lack of communication is suspected or demonstrated, e.g. when there is an associated aqueductal stenosis.[6,7]

The best surgical results obtained to date are those of Carmel *et al.*,[6] who reported a mortality of 27%. Most other series, as evidenced by our 4 cases, have a bleak outcome.[2,4,5]

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Metastatic Hepatocellular Carcinoma of the Heart

A Case Report

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SUMMARY

A metastatic tumour growing in the heart more than 2 years after total removal of the primary hepatic growth is a rare event. Jaundice as a result of bile production by the metastasis is most unusual. The primary tumour did not recur in the regenerated liver. Features which may prove useful in making a clinical diagnosis are briefly considered. Death resulted from a thrombo-embolus from the right atrium lodging in the opening of the tricuspid valve.

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Metastatic cardiac tumours are not often diagnosed clinically and are infrequently seen at necropsy. In this article we report a case of metastatic hepatocellular carcinoma growing in the right ventricle of the heart. The primary liver tumour had been excised $2\frac{1}{2}$ years previously, and no recurrence was evident at postmortem examination. The cardiac metastasis and the pulmonary emboli from this source produced a large amount of bile, resulting in icterus. We are not aware of a similar case in the literature.

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CASE REPORT

A 59-year-old Black man was admitted to hospital on 9 February 1977 with complaints of progressive dyspnoea on exertion for about 3 months, swelling of the lower legs, and progressive, painful distension of the abdomen. In 1974 a partial right-sided hepatectomy had been performed for a well-circumscribed liver cell carcinoma. A cholecystectomy had been done simultaneously.

On examination the patient was found to be fairly well nourished. He was slightly jaundiced, and had signs of right-sided heart failure. The left lobe of the liver was palpable and no right lobe was evident. The old operation scar was unremarkable.

Laboratory investigations showed a haemoglobin concentration of 14,9 g/100 ml; a red blood cell count of 4,53 million/μl; a white cell count of 4 400/μl with a differential leucocyte count of neutrophils 51%, lymphocytes 37%, monocytes 9%, eosinophils 2% and basophils 1%. The erythrocyte sedimentation rate was 8 mm/1st h (Westergren); total protein 70 g/l, albumin 31 g/l and

globulin 39 g/l. The total bilirubin was 23 μmol/l. The α-fetoprotein level was not raised. Aspartate aminotransferase was 85 U/l, alanine aminotransferase 57 U/l, lactic dehydrogenase 618 U/l and alkaline phosphatase 110 U/l. Urea was 6,2 mmol/l, blood glucose 6,2 mmol/l and electrolytes were normal.

Radiography of the chest revealed a large heart, but no congestion of the lungs; there was collapse of the right middle lobe and elevation of the right side of the diaphragm with basal collapse. There were gas shadows in the liver area and large bowel. A liver scan revealed only a left lobe with no abnormalities. A heart scan showed an enlarged heart with what could have been a pericardial effusion. A lung scan revealed reversed and increased flow in the upper regions but no other abnormalities. The ECG showed a regular sinus rhythm with no abnormalities. The patient did not respond to therapy and died on 4 March 1977.

Autopsy Findings

A moderate amount of straw-coloured ascitic fluid was found. The mild jaundice noted clinically was no longer apparent. Pleural adhesions were present.

The heart was enlarged and weighed 925 g. The main tumour involved the right ventricle only. However, sparse, round, white epicardial tumour deposits, seen best with a hand lens, were noted over the right ventricle only. The cavity of the right ventricle was virtually obliterated by a diffuse, smooth nodular mass (Fig. 1) which was a bright green colour, indicative of the bile formed by the tumour. The lesion extended from immediately below the tricuspid and pulmonary valves to the apex. Although this lesion was markedly exophytic in its growth pattern, it had extensively infiltrated the ventricle wall and septum. The tumour bulged behind the posterior cusp of the tricuspid valve and involved the papillary muscles and adjacent distal attachment of the chordae tendineae. The cusps of both valves were free of tumour.

The right atrium was dilated and hypertrophied and extensive antemortem mural thrombus was present in the atrium and its appendage. From this a large irregular rounded embolus had arisen which was firmly lodged in the orifice of the tricuspid valve. The inferior vena cava contained no tumour or thrombus. The pericardium, apart from a 'milk spot' and the sparse minute epicardial tumour deposits, was unremarkable.

The left ventricle had a small chamber and the wall was 1,3 cm thick. There was no evidence of tumour involvement at this site. The lungs were congested, oedematous and no tumour deposits could be seen macroscopically.

The liver weighed 1 200 g, presumably as a result of regeneration and was situated mainly to the left. Numerous adhesions to the colon were present in the right bed of the liver at the site of the previous operation. Chronic passive congestion was noted. Numerous slices were made through the entire organ, but no tumour recurrence could be found. The tissue was sampled randomly for microscopical examination.

The kidneys and spleen were congested. The endocrine organs were unremarkable. The vertebrae contained no visible tumour.

Microscopical examination of the cardiac metastasis revealed well-differentiated liver cell carcinoma producing unusually large amounts of bile (Fig. 2). Mainly a trabecular and a lesser tubular component were present. Sinusoidal channels were lined at fairly frequent intervals with normal-looking Kupffer cells. A few small areas of necrosis were observed. The tumour infiltrated on a broad, blunt front into the outer third of the myocardium (Fig. 3). Beyond this, in the epicardial fat, were occasional small tumour deposits. The pulmonary arterial tree contained infrequent small tumour emboli that were histologically identical to the tumour in the heart. Some of the emboli were beginning to show an expansile growth pattern; mild congestion and oedema were present. The liver, apart from congestion, siderosis and mild fatty change, showed no trace of tumour recurrence or of cirrhosis. Splenic cords and sinusoids were congested and small

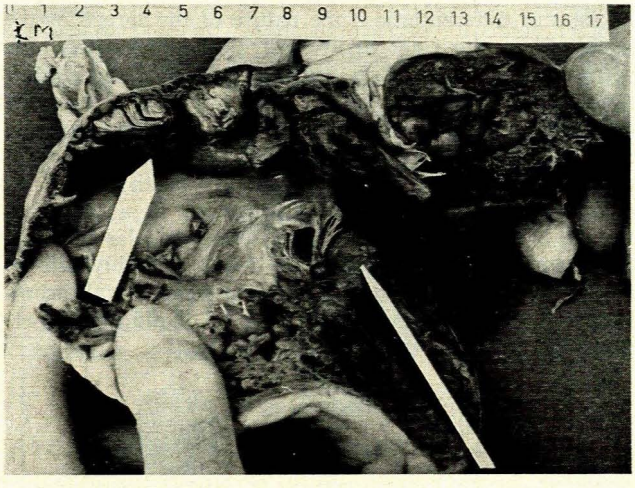

Fig. 1. Macroscopical appearance of the heart. The long arrow points to the tumour in the right ventricle. Above this is the tricuspid valve and a thrombo-embolus pinned in original position in the valve orifice. The short arrow points to a mural thrombus in the right atrium.

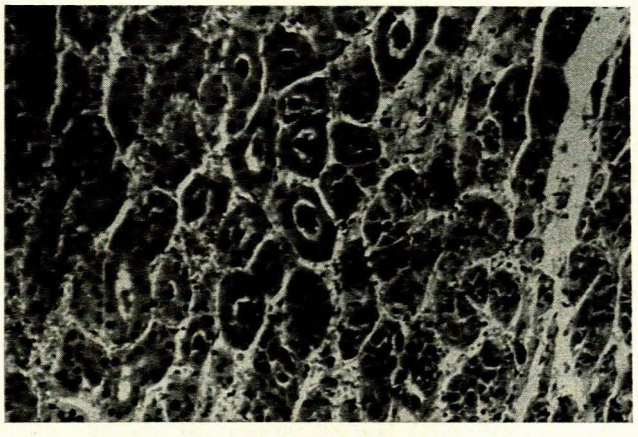

Fig. 2. Bile in the tubular component of the tumour.

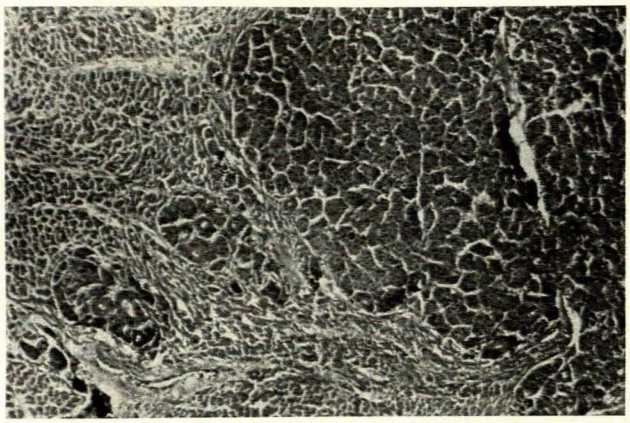

Fig. 3. Blunt tumour infiltration of heart muscle. The tumour can be seen in the right half of the field.

amounts of iron were noted. Renal congestion and moderate atheroma of the renal arteries and abdominal aorta were present. The other organs were unremarkable.

The final diagnosis of the cause of death was acute-on-chronic right-sided heart failure as a result of a thrombo-embolus lodging in the tricuspid valve opening. This had originated from a mural thrombus in the right atrium. Growth of a metastatic hepatocellular carcinoma in the right ventricle wall and septum had previously caused impedance of blood flow through the right ventricle.

DISCUSSION

The symptomatology of cardiac metastases has been stated as variable, depending as much or more on location than on size. Extensive involvement of organs may occur with few or no symptoms; and generally there is an absence of symptoms referable to the heart.[1]

In our case, apart from cardiomegaly and right-sided heart failure, the patient showed no other remarkable cardiac features, or ECG changes. Rarely, an ECG may show changes of infarction, or evidence of previous heart disease may be recorded. Clinical features suggesting tumour spread to the heart[1] are heart block, symptoms referable to the site of the tumour other than heart block, symptoms without apparent cause in a patient with a known malignant disease, accumulation of haemorrhagic pericardial and pleural fluid and suggestive radiographic observations. There are no characteristic fluoroscopic or electrocardiographic signs in patients with tumour nodules in the myocardium. Angiocardiography may prove valuable in delineating the presence or absence of intracardiac tumour masses.[1]

Growth of metastatic hepatocellular carcinoma of the heart is a rare event. Primary hepatocellular carcinoma usually exhibits extrahepatic spread in 55% of cases.[2] Lung, regional nodes, adrenal glands and bone are the usual sites to which spread may occur. The mortality rate exceeds 95% 6 months after a diagnosis has been made,[2,3] and within 1 year of onset of symptoms.[3]

The commonest sources of cardiac metastases are lung and breast carcinomas, malignant melanomas, lymphomas and leukaemias, and miscellaneous malignancies including hepatocellular carcinoma. The incidence of hepatocellular carcinoma varies in accordance with geographical distribution, almost certainly due to environmental influences.[4] Most areas of Africa and parts of south-east Asia show a high incidence of the disease, especially in males. Thus, Lothe and Somers[5] examined 6 644 necropsy specimens with 377 malignancies, and 26 of these showed macroscopical evidence of metastases to the heart and pericardium. In 2 of the 26 cases, the primary source was liver cell carcinoma. Metastatic carcinoma to the heart is usually accompanied by foci elsewhere, usually in the lungs. It is usually a late manifestation of spread.

Intravascular tumour growth is an important feature of hepatocellular carcinoma.[6] Retrograde lymphatic spread and direct spread are highly unlikely in a case like ours. We feel that spread of this tumour was probably haematogenous with direct implantation on the endocardium or as a result of extension from local myocardial metastases after coronary artery embolization. The extension was clearly not from a pericardial direction.

It is a likely assumption that in our patient cardiac seedings were present at the time of or before his operation in 1974. However, perhaps because of the excellent differentiation of the metastasis, slow growth occurred. Subsequently, microscopical emboli from the cardiac tumour appeared to have spread into the pulmonary arterial tree.

Although neoplastic implantation on the endocardium is uncommon, tumour emboli reaching the heart chambers may implant themselves on the endocardium directly, with 'subsequent development in the direction of least resistance, i.e. out into the cavity and between the muscular trabeculae'.[7] This description could well fit the present case.

The following features were unusual in our patient: the long postoperative survival of over 2 years, despite the tumour differentiation; the nature of the spread of the metastasis in the right ventricular wall and septum; the microscopical lung emboli breaking off from the cardiac tumour; the large amount of bile produced in a very well-differentiated metastasis and the resulting icterus; no evidence of primary tumour recurrence in the remaining regenerated liver; and sudden death caused by a large portion of right atrial thrombus as a thromboembolic ball lodging in the right atrioventricular opening. The increased weight of the heart was probably due to the weight of the tumour and the right atrial mural thrombosis and the embolus, rather than to significant hypertrophy of the heart muscle.

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