Case reports

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Cardiac tamponade caused by metastasising haemangioendothelial sarcoma of the liver

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SUMMARY. A case of primary haemangiosarcoma of the liver with secondary deposits in the pericardium is described. The patient presented most unusually with cardiac tamponade. There was no association with vinyl chloride, thorium dioxide, or arsenic.

Primary haemangioendothelial sarcoma of liver is rare and usually presents with abdominal pain and weight loss (MacSween et al., 1973; Ludwig and Hoffman, 1975). Most patients die of hepatic failure or exsanguination from the hepatic tumour (Ludwig and Hoffman, 1975). Metastases are reported in less than half of the cases and are rare in the pericardium (Ludwig and Hoffman, 1975). We describe a patient with primary haemangioendothelial sarcoma of the liver who presented with cardiac tamponade from a haemopericardium resulting from secondary deposits in the right atrial wall.

Case history

A 58-year-old plumber developed bronchospasm in March 1976. A chest x-ray film in July 1976 showed an enlarged heart shadow. While awaiting a referral appointment, the patient suddenly developed severe epigastric pain, had a haematemesis, and collapsed.

On admission to hospital he was unconscious and shocked. On examination he was a pyrexial, jaundiced, and centrally cyanosed. He had a sinus tachycardia of 140 per minute, an unrecordable blood pressure, and a raised jugular venous pressure. The apex beat was not palpable but cardiac dullness to percussion was increased. Heart sounds were inaudible. The abdomen was soft with epigastric tenderness. A smooth, firm liver edge was palpable 6 cm below the costal margin. Central venous pressure was then recorded as +35 cm H₂O. A chest radiograph showed a large pericardial effusion and abdominal radiographs were unremarkable. A diagnosis of pericardial tamponade was made. After aspiration of 350 ml blood-stained fluid the patient became conscious, the heart rate fell to 80 per minute, blood pressure rose to 130/80 mmHg, and the central venous pressure fell to +8 cm. The patient was now able to give a history of long-standing indigestion, recent malaise, and weight loss. As a plumber he had been exposed to platinum and asbestos.

The pericardial fluid had a packed cell volume of 28 (blood PCV 32) and did not clot. Neoplastic cells were not found and the fluid was sterile on culture. The electrocardiograms varied showing sinus rhythm and occasionally atrioventricular dissociation or atrial fibrillation. There were no changes of myocardial infarction. Echocardiography disclosed no evidence of aortic dissection. On admission haemoglobin was 11·8 g/dl, ESR 3 mm/hour, SGOT 1960 U/l, HBD 2245 U/l, SGPT 1230 U/l, and alkaline phosphatase 236 U/l (normal <92 U/l). There was no bleeding diathesis. Alpha-fetoprotein was negative and alpha-1-antitrypsin normal.

Forty-eight hours after admission the patient developed peritonitis caused by a perforated peptic ulcer and died before an operation could be attempted.

Pathology

At necropsy, the immediate cause of death was a perforated peptic ulcer. The liver was enlarged (1174 g) and contained numerous purple nodules (up to 2 cm in diameter) which on section had haemorrhagic centres and ill-defined margins. The
pericardial cavity contained about 200 ml blood and the epicardium was partly covered by fibrin. There were 2 haemorrhagic secondary deposits (8 x 5 x 4 cm and 3 x 2 x 2 cm) on the visceral pericardium. Both were firmly adherent to the posterolateral aspect of the right atrium close to the venae cavae (Fig. 1). The endocardium was macroscopically intact. Both lungs showed numerous deposits 2 x 1 x 1 cm in size. Adherent fibrin covered the deposits on the lung surface.

Microscopically the liver nodules were vascular neoplasms consisting of slit-like spaces of variable size and shape lined by proliferating, plump endothelial cells (Fig. 2). These had a tendency to form clumps and new channels. At the periphery of the nodule the proliferating cells had infiltrated between the liver cell plates. A few individual liver cells were entirely surrounded by the neoplastic cells suggesting that the neoplasm was of primary liver origin. The rest of the liver showed centrilobular congestion.

Fig. 1 The liver shows multiple haemorrhagic nodules with ill-defined margins. The pericardial cavity contains 2 lobulated and haemorrhagic metastatic deposits, adherent to the epicardium (arrowed).

Fig. 2 Histology of liver nodules (x 300). (A) (H and E), showing blood containing spaces lined by large and hyperchromatic endothelial cells. (B) Reticulin stain.
Cardiac tamponade

The histological features of the pericardial and pulmonary metastases were similar to those of the hepatic neoplasm. The right atrial myocardium was densely infiltrated by malignant endothelial cells but the endocardium was intact. No other secondary deposits were found.

Discussion

Extrahepatic metastases from liver haemangioendothelial sarcoma have been reported in the spleen, lungs, portal lymph nodes, and abdominal viscera (Edmondson, 1958). Pericardial metastases appear to be rare; we have been able to find only one example, but without tamponade (Thomas and Popper, 1975).

Primary liver haemangioendothelial sarcoma usually presents with abdominal pains, malaise, and weight loss, while primary cardiac intramural haemangiosarcoma commonly presents with pericardial effusion and tamponade (Ludwig and Hoffman, 1975; Rossi et al., 1976; Patt et al., 1974). The primary tumours from both sites and their metastases are haemorrhagic (Ludwig and Hoffman, 1975; Rossi et al., 1976). Primary cardiac angiosarcoma is also mainly found on the right side of the heart (Rossi et al., 1976; Patt et al., 1974).

The histological findings in this case are in favour of a primary liver tumour because of the ill-defined margins of the liver nodules, the intermingling of the plump endothelial cells with hepatocytes, the presence of normal looking hepatocytes within the nodule, and the multifocal distribution of the nodules (Fig. 2). The presence of two deposits in the pericardial cavity in association with an intact endocardium makes it unlikely that the neoplasm was cardiac in origin. The pulmonary distribution and appearances are not compatible with a primary lesion in the lung.

The association of liver haemangioendothelial sarcoma with exposure to vinyl chloride, arsenic, and thorium dioxide is well documented, though the tumour has also been reported in patients with no obvious exposure to any of these agents (MacMahon et al., 1947; Roth, 1957; Creech and Johnson, 1974; Dalderup et al., 1976). Our patient, as far as is known, had never been exposed to vinyl chloride or its process of polymerisation, and he had never been given thorium dioxide. Postmortem analysis of his hair showed no significant content of arsenic (less than 1 p.p.m.). He had been exposed to asbestos though there was no evidence of asbestosis at necropsy. Though specific aetiological factors are absent in this patient, he may have been exposed to some agent of which we are as yet unaware.

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References


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