Leiomyosarcoma of the Inferior Vena Cava Propagating into the Right Atrium

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Primary tumours of the venous system are not common. Thomas and Fine (1960), reviewing the published material, collected 29 cases (including 2 of their own) of smooth muscle tumours arising from the media of the veins. Of these 29 tumours, 17 were malignant. The inferior vena cava was the most commonly involved vein (9 of 29 cases). Of the tumours involving the inferior vena cava, 8 were leiomyosarcomas and only 1 was an endotheioma. Hoffbrand and Lloyd-Thomas (1964) reported 14 cases of leiomyosarcoma of the inferior vena cava and added a case of their own. We are not aware of any case reports in which the diagnosis of leiomyosarcoma of the inferior vena cava has been made before an operation.

The purpose of this paper is to report a case of leiomyosarcoma, diagnosed in life by venocavography.

Case Report

The patient was a 35-year-old housewife, the mother of 5 healthy children. Her past history was non-contributory. Eleven months before her admission, while in the fourth month of her last pregnancy, she complained of pain in the right upper abdomen. On admission, in another hospital, physical examination revealed tenderness in the right hypochondrium. In addition the ESR was raised. She continued to suffer from abdominal pain until after the delivery. Subsequently the pains stopped for four months.

Two months before admission to our hospital, pain of increased severity reappeared. The pain occurred periodically but was not colicky in nature. She complained of nausea, loss of weight, and had a low-grade fever.

On admission the patient was found to be well nourished but appeared to be ill. Physical examination revealed a large, tender liver. The laboratory examinations were negative, except for the bromsulphalein test, which was positive. Radiological examination of the gall-bladder, intravenous pyelogram, and gastro-intestinal tests were all normal. Varices were suspected on barium swallow.

During her stay in hospital her condition deteriorated progressively. She had a high temperature and ascites appeared. To remove fluid repeated abdominal punctures were performed and diuretic treatment was begun. The abdominal fluid was sterile and contained 4·2 g protein/100 ml. No malignant cells were demonstrated on cytological examination of the fluid. Her symptonmatology, the normal liver function tests in the presence of an enlarged and tender liver, and suspected oesophageal varices suggested venous obstruction and the Budd-Chiari syndrome was suspected. Inferior venocavography was performed.

Contrast material was injected into the femoral vein and an occlusion of the inferior vena cava, distal to the renal veins, was demonstrated. The contrast material drained through collaterals and the aygos vein into the superior vena cava and through it into the right atrium. A filling defect was suspected in the latter chamber (Fig. 1). For a more conclusive demonstration intravenous angiocardiography was performed through the left cubital vein. This investigation clearly demonstrated a large polypoid mass which occupied the distal two-thirds of the right atrium, but did not involve the tricuspid valve (Fig. 2).

The history of her illness, the clinical examination, and the radiological findings, led to a diagnosis of leiomyosarcoma of the inferior vena cava, with extension into the right atrium.

The patient's poor condition precluded surgical intervention. A few weeks later uncontrollable oedema of the lower half of her body appeared; the patient lost consciousness and died in coma.

Necropsy revealed a leiomyosarcoma of the inferior vena cava. The tumour began at the level of the left renal vein, extended to the hepatic veins, and entered and filled the right atrium. The liver was very congested and showed fatty changes and fibrosis. Most of the hepatic veins were occluded by thrombi, largely composed of tumour cells. The tumour process also involved the retroperitoneal lymph glands in the region
Fig. 1.—Inferior venocavography. The inferior vena cava is occluded below the renal veins. The contrast material drains through the azygos system into the superior vena cava. In the right atrium a filling defect is outlined.

of the inferior vena cava. Microscopically the tumour was proved to be a leiomyosarcoma (Fig. 3, 4, and 5).

Discussion
To the best of our knowledge this is the first case of a primary tumour of the inferior vena cava diagnosed by venocavography. Except for the cases diagnosed accidentally during operation, it is probably the first case of leiomyosarcoma of the vena cava recognized during life.

The case is also interesting from the diagnostic point of view, in view of the history of the illness, which began with pain in the right upper abdomen; the onset of pains coincided with the beginning of pregnancy and disappeared after delivery. At this stage of the illness, the liver was not enlarged. A few months after delivery her complaints reappeared, but at this stage the liver was enlarged. This observation suggested that the primary disease process arose from the abdominal area, causing occlusion of the inferior vena cava, extending cranially, and involving the hepatic veins.

It was assumed that partial occlusion of the inferior vena cava by the tumour itself existed during pregnancy, and that the occlusion was aggravated by the pressure of the pregnant uterus, which also added to the volume of the venous return proximal to the obstruction. These factors were relieved by the delivery which thus resulted in symptomatic improvement in the patient's condition. The subsequent deterioration was probably due to occlusion of the hepatic veins.

The first diagnosis considered was thrombosis of the inferior vena cava, extending into the right atrium. It is known that thrombosis of the inferior vena cava extends in the direction of the blood flow. It is also known that a thrombus has an irregular surface when present in one of the heart cavities. In the case outlined above, extension of the process in the direction of the blood flow was assumed, but the presence of a large polypoid mass with a regular surface in the right atrium eliminated the possibility of thrombosis. In the differential diagnosis, myxoma of the right atrium had to be taken into consideration. Myxoma of the right atrium usually arises from the atrial septum in the region of the fossa ovalis and frequently has polypoid contours. Myxoma almost always grows in the direction of the adjacent valve ring, causing stenosis of the valve.
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The latter features were not present in our case. Another possibility was that of an abdominal tumour, which invaded the inferior vena cava and extended through it. This possibility could not be eliminated with certainty.

Intracavitary primary tumours of the heart, apart...
from myxoma, are rare and therefore no angiocardio graphic signs for diagnosis exist. On the other hand, metastatic tumours of the heart are more common. These are usually mural and not intracavitary.

Leiomyosarcoma is a slow growing tumour which infrequently metastasizes. It usually spreads by continuity. The clinical features caused by these tumours are due to obstruction of the blood flow and are dependent on the level at which they occur. The occurrence at the upper third of the inferior vena cava produces all the features of the Budd-Chiari syndrome. When the middle third of the cava is occluded by the tumour, the clinical picture of renal vein thrombosis is produced. Tumours of the lower third cause oedema of the legs. Sometimes, depending on the collateral circulation, the signs and symptoms of venous occlusion are minimal, the patient suffering only from abdominal or back pain.

In our case the diagnosis was made too late, but if it had been made at an earlier stage, an operation (Cope and Hunt, 1954) could have been attempted.

Venocavography, and if necessary intravenous cardioangiography, provide the only reliable method for diagnosing tumours of the inferior vena cava.

References