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Primary angiosarcoma of the spleen: a diagnosis to consider

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We discuss a case of angiosarcoma of the spleen. Splenomegaly was discovered on a routine examination. At a later date it increased further in size and became painful. Peripheral smear and radio-isotope scannings suggested non-functioning spleen with haemorrhage within the tumour. Characteristic features of micro-angiopathic haemolysis, suggested in previous reports, was not found in our case.

Key words: Neoplasm; spleen.

Introduction

Angiosarcoma of the spleen is a rare primary malignant neoplasm. It is generally discovered at a late stage, having undergone metastatic spread, with a consequently poor prognosis. Although painful splenomegaly and micro angiopathic haemolytic anaemia are characteristic features, presentation is variable and may be in the form of spontaneous rupture. Thus the diagnosis is not usually made before laparotomy or post mortem examination. We report a patient in whom the condition was entirely unsuspected before operation.

Case report

During routine admission in February 1984 for transurethral prostatectomy a 78-year-old man was found to have a moderately enlarged spleen. His haemoglobin (Hb) was 11.0 g/dl, white cell count $8.0 \times 10^9/1$, and platelet count $493 \times 10^9/1$. On the peripheral blood smear there was anisocytosis, poikilocytosis and Howell Jolly bodies, but fragmented red cells were not seen. Bone marrow examination and chest X-ray were normal.

During the following months his spleen enlarged further and he became more anaemic, despite correction of iron and folate deficiency. Further investigation, including barium studies, revealed no alimentary source of blood loss. In view of his age, surgical intervention was not considered until February 1985, when he developed left upper quadrant pain and tenderness over the spleen, which was now found to be considerably more enlarged, extending down to the umbilicus. Blood examination showed a persisting hypochromic anaemia (Hb 7.9 g/dl).

A technecium colloid scan showed increased uptake compatible with blood loss in the spleen, but there was negligible uptake of technecium labelled heat damaged red cells, indicating that the spleen was not functioning. Ultrasound examination demonstrated solid inhomogeneous splenic enlargement. After correction of his anaemia splenectomy was performed in March 1985.

The spleen measured $23 \times 17 \times 10$ cm and was adherent to colon, omentum and lateral abdominal wall. The sites from which the adherent splenic tumour was dissected were marked with metal clips for the guidance of subsequent radiotherapy. Some small nodules on the surface of the liver were biopsied.

Pathological examination showed multiple white tumour nodules up to 10 cm in diameter within the spleen, the largest with haemorrhagic centres (Figure 1). On microscopy the pattern of a well-differentiated angiosarcoma was seen (Figure 2), with a histological spectrum ranging from cavernous spaces and vascular channels to almost solid sheets. No tumour was identified in the liver biopsies.

The patient made a good recovery from his operation. He underwent a course of abdominal radio-

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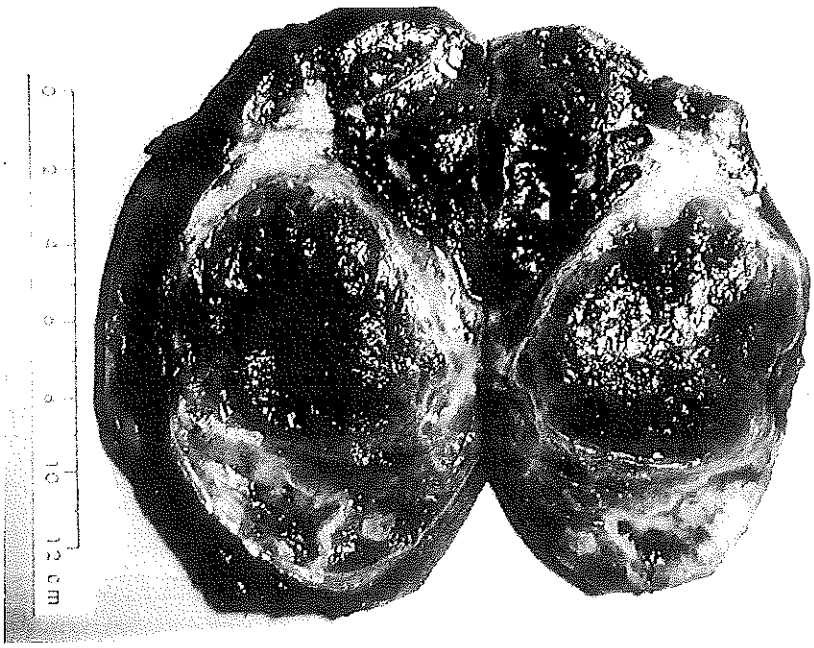

Fig. 1. Macroscopic section of spleen showing multiple tumour nodules of varying size. Haemorrhage has occurred into the larger nodules.

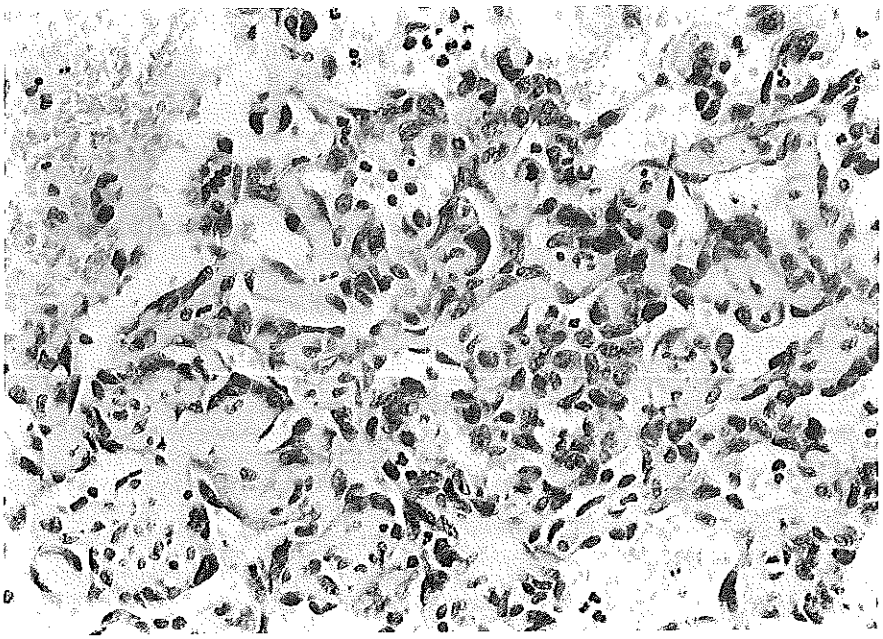

Fig. 2. Atypical endothelial cells lining blood-filled spaces. Three mitotic figures are seen. (Haematoxylin and eosin × 400.)

therapy, and remained well and clinically free of recurrence for 18 months. He then rapidly deteriorated, and was re-admitted with jaundice and gross hepatomegaly. Liver ultrasound examination showed the typical appearance of metastases, and he developed persistent gastrointestinal bleeding from which he died. Permission was not given for a post-mortem examination.

Comment

On the basis of their review of 50 reported cases of angiosarcoma of the spleen Autrey and Weitzer[1] suggested that this rare tumour be considered in patients with splenomegaly, an associated microangiopathic haemolytic anaemia, and no evidence of malignant lymphoma or leukaemia. Several recent reports[2–5] have indicated that radioisotope scanning, arteriography and computed tomography may provide further evidence for the diagnosis. Our patient did not fulfil the characteristic clinical criteria, as although he was anaemic he did not have features of microangiopathic haemolysis. It has been postulated that this results from traumatic disruption of erythrocytes passing through irregular vascular channels lined by malignant endothelial cells.[1] Although the radioisotope and ultrasound scans in this case showed some features of splenic angiosarcoma, we were not alerted to the diagnosis, mainly because of its rarity.

It is clear, however, that the tumour should be considered in the differential diagnosis of a non-functioning spleen, and increased awareness, supported by the results of imaging procedures, may allow earlier diagnosis. Although the outlook after discovery is extremely poor, with only 19% of patients surviving 6 months,[6] it seems likely that earlier operation may improve the prognosis.

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