Postgraduate Medical Journal (May 1981) **57**, 314–316

Polycythaemia vera presenting as an acute abdomen

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Summary

A patient who presented with severe abdominal pain was found to have an intrahepatic haematoma complicating previously undiagnosed polycythaemia vera (PV). Full recovery followed treatment with bed rest, control of hypertension, daily venesection and ^{32}P. The hazards of surgery in uncontrolled PV are discussed and re-emphasized.

Introduction

Serious postoperative complications occur commonly in undiagnosed or inadequately treated PV (Wassermann and Gilbert, 1963). It is therefore important to differentiate the intra-abdominal presentations of PV which require only medical management from those where venesection is necessary before urgent surgery (Cryer and Kissane, 1979). The case described here illustrates this point and to the authors' knowledge is the first report of PV presenting with severe abdominal pain due to intrahepatic haemorrhage.

Case report

A 65-year-old retired diplomat was admitted as an emergency in March 1979 following the sudden onset of severe tearing pain in his right lower chest and abdomen, while bending to put coal on his fire. He looked unwell and was noted to be cyanosed but not breathless. The pulse and BP were normal and all the peripheral arterial pulses were present. There was no clinical abnormality of the heart or lungs. There was fullness, marked tenderness and dullness to percussion in the right hypochondrium. The bowel sounds were present and both rectal examination and tests for faecal occult blood were negative. Urinalysis showed no abnormality. An X-ray of the abdomen showed a large soft tissue mass in the right hypochondrium. The serum amylase was normal. The diagnosis at this stage was considered to be extensive haemorrhage into the liver.

Further detailed questioning then revealed that he had been less well in the preceding year, noting chest pain and breathlessness on exertion, intermittent discoloration and pain in the toes of the right foot, and pruritus especially after a hot bath. Temporary loss of vision in the right eye lasting 15 min had occurred also on the night before admission. He was taking propranolol for hypertension which had been diagnosed 6 months previously but was on no other medication. These additional features raised the possibility of underlying PV. The results of further investigations supported this with Hb, 18·4 g/dl; PCV, 55%; WBC, 41×10^9/l; neutrophil leucocytosis and platelets, 520×10^9/l. Examination of sternal marrow showed a panmyelosis, excess of megakaryocytes and reduced stainable iron. Red cell volume was 40 ml/kg (normal range 25–35 ml/kg) and leucocyte alkaline phosphatase was 240 (control 60). The diagnosis of PV was thus confirmed.

Initially the patient was treated with bed rest and daily venesection, and as the abdominal tenderness settled, both liver and spleen became easily palpable. Four days after admission he developed bruising of both flanks consistent with blood tracking from the liver *via* the bare area to these sites. The presence of a haematoma was established later by ultrasound which showed a large transonic area posteriorly in the right lobe (Fig. 1). One month after receiving 5 mCi of ^{32}P his Hb was 14·4 g/dl, PCV 43%, WBC $12·0 \times 10^9$/l and platelets 280×10^9/l. One year later, a further ultrasound study showed only residual changes in keeping with resorption of his haematoma (Fig. 2). His BP and haematological indices had remained well controlled and he was looking forward to an active retirement.

Discussion

The paradoxical complications of haemorrhage and thrombosis in PV are well known. These occur with about equal frequency in 33–50% of patients, and were responsible for 50% of deaths before treatment with ^{32}P became available (Videbaek, 1950). Furthermore, Wassermann and Gilbert (1963) recorded a 37% mortality and an additional 46% morbidity postoperatively in uncontrolled PV due almost exclusively to these vascular complications.

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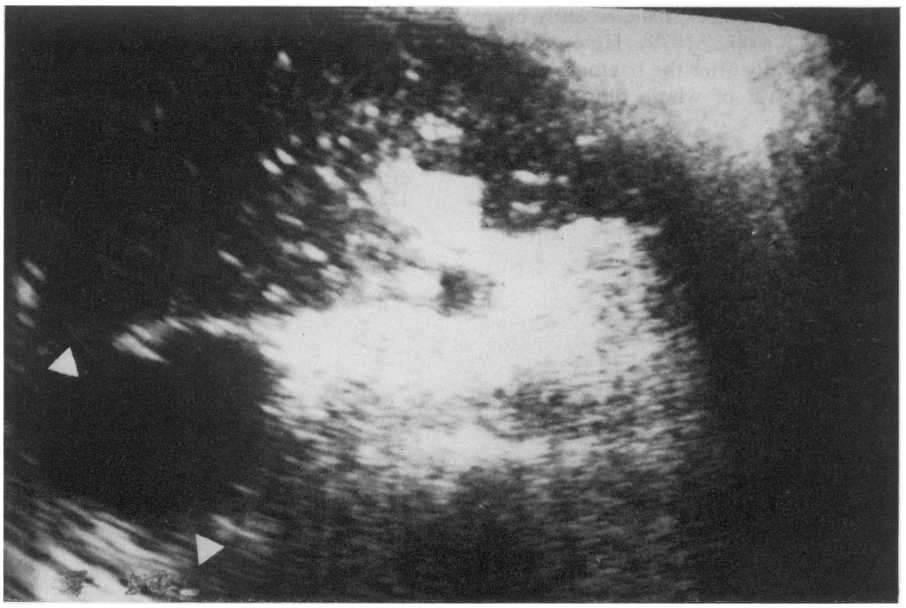

FIG. 1. Transverse scan of abdomen at the level of upper pole of kidney showing transonic area posteriorly in the right lobe of liver, one month after presentation. Arrows indicate site of haematoma.

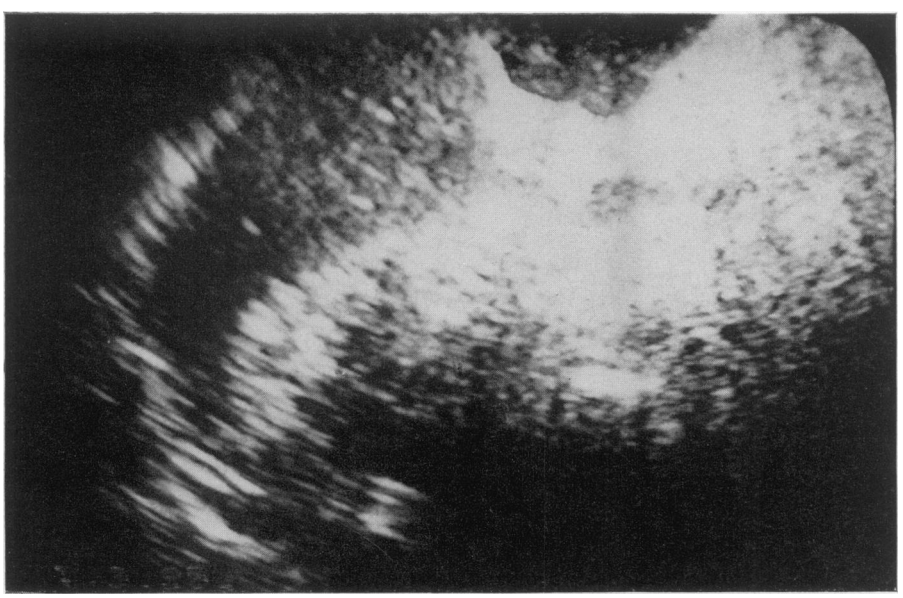

FIG. 2. Appearances one year later confirming resorption of haematoma.

Barabas (1980) has recently re-emphasized the exceptional hazards of surgery in this situation. Recognition of underlying PV is therefore critical in patients presenting with acute abdominal pain. Awareness of causes such as intrahepatic haematoma, retro-peritoneal haemorrhage and splenic infarction which do not require urgent laparotomy is equally important. Other causes of abdominal pain in PV,

including mesenteric thrombosis, have recently been reviewed (Cryer and Kissane, 1979). Here, surgery should be undertaken only after the haematocrit has been lowered by removal of whole blood and re-infusion of plasma.

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