X-ray Examination of Hands.—Osteoporosis could be seen adjacent to several joints, and several joint erosions, considered by the radiologists to be compatible with gout or rheumatoid arthritis.

Screening of Chest.—This confirmed isolated dextrocardia with left aortic descent. The stomach was normally situated. The lungs were very translucent but otherwise normal. On the films the pattern of the smaller lung vessels was thought consistent with enlarged bronchial arteries.

Electrocardiogram.—There was sinus rhythm at rest, with grade I block (P-R 0.3 sec.), but, on exercise, nodal rhythm with some ventricular escape. The record was difficult to interpret: there was a tall R wave in aVR, a deep S wave in aVF, aVL, and from V₃R to V₆, reaching a maximum of 38 mm. in V₂. In V₃R the R wave was 12 mm.

Cardiac Catheterization (Dr. R. M. Marquis).—The catheter traversed the right superior vena cava and a capacious atrial chamber. It could be passed into two ventricles but not into pulmonary artery or aorta. Its behaviour suggested a common atrioventricular canal. The atrial pressure was high—20 mm. Hg systolic, 5 mm. Hg diastolic—and the ventricular pressures were each about 125/10 mm. Hg. Gross oxygen undersaturation was confirmed by the following results: brachial artery 68.4%, superior vena cava 49.3%, atrium 58.3% (average), and ventricle 49.3% (average).

Selective Angiocardiography.—A large ascending aorta and grossly enlarged pulmonary arterial branches opacified simultaneously. It was impossible to decide whether the pulmonary arteries came off the aorta.

From the accumulating evidence it was thought that there was severe pulmonary arterial hypertension, and that the basic abnormality was possibly a truncus arteriosus or modified Eisenmenger complex. The congenital defects were so complicated as to rule out any possibility of surgical relief.

COMMENT

It seems unlikely that a gouty diathesis was present in this case and merely exacerbated by the prolonged anoxic polycythaemia. Again, although the patient had been having chlorothiazide, this was only in recent months and flagrant gout had been present long before this. Attention has recently been drawn to the hyperuricaemic effects of the thiazide diuretics and how they may make latent gout overt (*Brit. med. J.*, 1960). Though none of the patient's relatives have been studied, there was certainly no history to suggest familial gout, and the gout had become apparent at the age of 31, thereafter persisting with no particular seasonal fluctuation.

While the exact nature of the congenital cardiac defects could not be determined it was clear that a veno-arterial shunt had been present from birth. In turn, the polycythaemia could be reconciled with the patient's anoxia, and the haematological findings were not in accord with polycythaemia vera. If, as seemed likely, the daily red-cell breakdown was at least twice normal, this would be associated with an enormous turnover of erythroid precursors, and it was thought that this adequately explained the splenomegaly, hyperuricaemia, and gout. Talbott (1959) estimates that a blood dyscrasia is present in 10% of all patients who have gout. Whitby and Britton (1957) state that gout occurs too often with polycythaemia to be explained by chance.

The essential difference between familial and secondary gout lies in the mechanism responsible for the increased concentration of uric acid in the body fluids, and increased excretion. Familial gout results ultimately from an inborn metabolic error regarding amino-acid pathways. In secondary gout there is no such metabolic error; rather there is increased formation or destruction

of red and white corpuscles present in excess, resulting in increased nucleic-acid metabolism in the haemopoietic tissues, with elaboration of the intermediate purine metabolites. The eventual outcome is increased uric acid available for excretion. Uric-acid levels may be similar in both types of gout. Nor does there appear to be anything characteristic about the distribution of affected joints.

In the case described here it was concluded that the patient had gout solely because he had such a high degree of compensatory polycythaemia, and in the circumstances outlined the only treatable aspect was the hyperuricaemia. Mason (1959) regards probenecid as the drug of choice in the interval therapy of gout, and the patient was discharged taking this drug, 0.5 g. thrice daily.

That clinical gouty arthritis is not seen more commonly with cyanotic congenital heart disease may be because patients with this form of heart disease and the necessary degree of polycythaemia do not usually survive long enough for this complication to develop.

The patient was under the care of Dr. M. R. Geake, Preston, and later of Dr. M. B. Matthews, Edinburgh, whose permission to publish the above case report is gratefully acknowledged.

EWEN A. CAMERON, M.B., M.R.C.P.Ed.,
Senior Medical Registrar, Royal Victoria Infirmary,
Newcastle upon Tyne; formerly locum Senior Registrar,
Cardiac Department, Western General Hospital,
Edinburgh.

REFERENCES

- Brit. med. J.*, 1960, 2, 45.
Hickling, R. A. (1953). *Lancet*, 1, 57.
Mason, R. M. (1959). *Brit. med. J.*, 1, 1522.
Sydenham, Thomas (1683). *Works*, translated from Latin edition of Dr. Greenhill by R. G. Latham, Vol. 2, p. 123. Sydenham Society, London, 1850.
Talbott, J. H. (1959). *Medicine (Baltimore)*, 38, 173.
Whitby, L. E. H., and Britton, C. J. C. (1957). *Disorders of the Blood*, 8th ed., p. 501. Churchill, London.

Spontaneous Rupture of the Spleen in Acute Myeloid Leukaemia

It is well known that myeloid leukaemia may present with symptoms due to splenic enlargement, such as increasing size of the abdomen, or left-sided abdominal pain caused simply by the weight of the enlarged organ. Such pain may, however, be due to perisplenitis over an area of infarction. The patient at this time not infrequently appears to be in good health. It is very rare for this condition to present as abdominal pain from splenic rupture, although such presentation has often been reported in other diseases, such as malaria, infective mononucleosis, tularaemia, typhoid, etc.; and even the apparently normal spleen has been known to undergo spontaneous rupture. Only a handful of cases of spontaneous rupture of a leukaemic spleen have been recorded. Wolfson *et al.* (1954) found that in most of them the rupture occurred in acute leukaemia, and they report such a case in which the splenic rupture was the presenting feature. This case would appear to be one of acute lymphatic leukaemia. Cooray (1952) reports a case of spontaneous rupture of the spleen in chronic lymphatic leukaemia occurring in a Sinhalese man, and spontaneous rupture has been reported in chronic myeloid leukaemia (Neal, 1940; Greenfield and Lund, 1944; and Sarin and Sarin, 1957). As, at the time of writing, no case of spontaneous rupture of a leukaemic spleen had been reported to have occurred in the United

Kingdom in post-war years, we think it worth while to record a case of acute myeloid leukaemia presenting with spontaneous splenic rupture.

CASE REPORT

A man aged 58, an Irish rigger, was well and in apparent good health on June 6, 1959, and had not missed a day's work in the past two years. On June 7, after returning from church, he started to feel cold, shivery, and "off colour." After a brief rest he felt better. That night he retired at his usual hour and slept well, but awakened next morning feeling somewhat unwell. He remained away from work on June 8 and rested most of the day, and at 3.45 p.m. was awakened with a sudden severe pain in his left shoulder. The pain gradually subsided and he was able to get up and shave before his evening meal. He retired to bed early, and at 3 a.m. on June 9 was awakened with another severe bout of left shoulder pain. Over the next two days this gradually radiated down the left side of the back of his chest and the left side of his abdomen. He was able to tolerate the pain while he was at rest, severe pain remaining a matter of a few hours. During the evening of June 11 the pain became much more severe; it was now more abdominal in type, was continuous, and kept him awake all night. After examination by his own doctor he was admitted to hospital on June 12. He denied absolutely any trauma of recent months, and his wife likewise could not recall any bumps, knocks, or such injuries.

Examination on admission showed him to be in some distress from upper abdominal pain. The pulse rate was 112 a minute, soft, and of fair volume; B.P. 120/80, and respirations 24 a minute. The heart was clinically normal, but the lungs showed signs of chronic bronchitis and emphysema. The temperature was raised to 99.4° F. (37.4° C.). No lymph nodes were palpable, and the retinæ were normal. The tongue was coated but moist.

Examination of the abdomen showed it to be visibly distended, with marked tenderness and rigidity, especially in the left hypochondrium and epigastrium. Shifting dullness was thought to be present, and liver dullness was reduced, but this was difficult to interpret, as the overlying lung was emphysematous. No mass could be palpated, and the bowel sounds were faint and infrequent. Rectal examination revealed a ballooned rectum with some tenderness anteriorly.

A provisional diagnosis of perforation of a peptic ulcer, probably into the lesser sac, was made. A straight radiograph of the abdomen failed to show any gas under the diaphragm. Blood was ordered, and the pathology technician, while checking the Rhesus factor, astutely noticed an excess of white blood cells. He carried out a count, and this was found to be in excess of 200,000/c.mm. The differential count was: myeloblasts 88%, myelocytes 7%, polymorphs 3%, lymphocytes 2%. The patient was thus shown to be suffering from acute myeloid leukaemia, and the possibility of his condition being one of a rupture of a leukaemic spleen was then considered. With these two diagnoses in mind laparotomy was immediately performed, two hours after admission. Blood transfusion had been begun prior to the operation.

A right paramedian incision was made alongside the umbilicus, and on opening the peritoneal cavity a small amount of dark blood escaped. Exploration upwards was rewarded by the escape of a larger quantity of dark blood. The spleen was four to five times the normal size and was surrounded with blood clot and adhesions. The paramedian incision was extended upwards, and further access was gained by a left transrectus extension. A loculated collection of blood clot measuring about two pints (1.14 litres) was found between the diaphragm and the spleen, and further loculations were present anterior to the lesser omentum. The spleen was extremely friable, and the loculations as well as perisplenic adhesions hindered delivery. The splenic vessels were secured and splenectomy was performed.

The patient was placed on routine post-operative care, with an antibiotic cover, and his condition remained

reasonably good until the third post-operative day, when he complained of increasing left upper abdominal pain and of pain in the left shoulder. His condition thereafter deteriorated progressively in spite of resuscitative measures, and he died on the fifth day after operation.

Haematological studies were not complete, as it was thought unjustifiable to subject the patient to frequent blood examinations at the time. On the first post-operative day the white-cell count was 200,000/c.mm., myeloblasts were still in the vast majority, and the haemoglobin was 12.7 g./100 ml. On the third post-operative day the white cells numbered 292,000/c.mm., and the platelet count 189,000/c.mm. His blood group was B Rh-negative.

At necropsy the peritoneal cavity contained a quantity of sero-purulent fluid, mainly confined to the left upper abdomen. The liver appeared to be the seat of leukaemic infiltration, as did the recto-sigmoid junction of the large bowel, and pieces of these were taken for section.

Pathology.—The spleen was enlarged, measuring 8½ by 5 by 3½ in. (21.5 by 12.5 by 9 cm.), weight 910 g., and had a rather nodular surface. On cut section it was diffusely pale. Microscopically, sections showed diffuse infiltration of the Malpighian bodies and pulp with large round cells, many deposits of brown pigment, and several small sub-capsular infarcts. The liver showed diffuse infiltration of the sinusoids with large nonnuclear cells, focal infiltration of the central veins with similar cells and some polymorphonuclear leucocytes and lymphocytes. There was hypertrophy of the muscle coats of the colon, with heavy infiltration of polymorphonuclear leucocytes and lymphocytes in the serous coat.

COMMENT

The complete absence of a history of trauma from the patient or his wife supports the view that this is a case of so-called spontaneous rupture of the spleen. We feel, however, that, owing to the frequency of minimal unremembered bodily "traumata," it is impossible to rule out "trauma" as a factor in the aetiology in any case of so-called spontaneous splenic rupture.

The picture of the typical cell types and their numbers in the peripheral blood, the presence of splenic enlargement and especially infarction without enlargement of the lymph nodes, the histological features particularly of the spleen and liver, together with the very acute history, establish this as a case of acute myeloid leukaemia.

We are indebted to Mr. M. R. Ernst, surgeon in charge of this case, for allowing us to report it. We are also grateful to Dr. S. Marshall, pathologist, Rush Green Hospital, for the histology reports, and to the pathology staff generally for their help with the haematological studies.

M. J. FLOOD, M.B., F.R.C.S., F.R.C.S.Ed., F.R.A.C.S.,
Late Surgical Registrar, Rush Green Hospital, Romford, Essex.

R. A. CARPENTER, M.A., M.B., B.Chir., F.R.C.S.,
Surgical Registrar, Rush Green Hospital, Romford, Essex.

REFERENCES

- Cooray, G. H. (1952). *Brit. med. J.*, **1**, 693.
Greenfield, M. M., and Lund, H. (1944). *Ohio St. Med. J.*, **40**, 950.
Neal, J. M. (1940). *Med. Bull. Veterans' Adm. (Wash.)*, **17**, 96.
Sarin, L. R., and Sarin, J. C. (1957). *J. Indian Med. Ass.*, **29**, 286.
Wolfson, I. N., Croce, E. J., and Fite, F. K. (1954). *New Engl. J. Med.*, **251**, 735.

The revised edition of *The Medical Laboratory Technician* gives an outline of the techniques and methods involved in hospital laboratories and other medical laboratories, including the blood transfusion services, and indicates the training and personal qualities required. (H.M.S.O., Choice of Careers Series, No. 57, price 6d. net.)