SPLENOMEGALIC POLYCYTHAEMIA WITH HIGH BLOOD PRESSURE.

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It has been maintained by one of us (F. P. W.) that, contrary to what one would formerly have expected, in true cases of splenomegalic polycythaemia (Vaquez-Osler disease), or erythraemia, the blood pressure is generally not, or not much, above the average, in spite of the excessive so-called "blood viscosity"—in fact, that the presence of excessive blood pressure would suggest that any case in question is either not a true one of erythraemia or that it is complicated with contracted kidney or arteriolo-sclerosis (especially of the medulla oblongata, as a cause of arterial hyperpiesia).^{*} The following case bears on this and some other questions.

The patient, a single woman aged 53 years, returned from Switzerland in the early summer of 1927, complaining of violent indigestion, neuralgia in the eyes, cardiac palpitation, and slight irregular loss of blood per vaginam. A Swiss oculist had examined the eyes and pronounced them normal. An abdominal swelling, evidently an enlarged spleen, was discovered. It was rather hard, and reached down to below the umbilical level. A radiogram showed the greater curvature of the stomach indented, and the viscus displaced to the right by a mass corresponding in position to the spleen. The first blood counts showed no excess of erythrocytes, but a count on October 4th, 1927, gave: haemoglobin 104 per cent., erythrocytes 7,800,000 per c.mm., white cells 21,000 per c.mm. (of which the polymorpho-nuclear neutrophils were 74 per cent. and the lymphocytes were 18 per cent.). There could be no doubt that the case was one of true splenomegalic polycythaemia (the Vaquez-Osler disease, or erythraemia). The patient was thin and her face was pale-bronzed in hue, later rather high-coloured and telangiectatic in appearance. The fauces were deep red, and the whole picture was fairly typical of erythraemia. A most important feature, however, was the presence of high blood pressure, which had apparently been first noted during her visit in Switzerland. The brachial systolic blood pressure was on several occasions as high as 280 mm. of mercury.

It should likewise be mentioned that her liver could not be felt and there was no evidence of any abdominal neoplasm. None of the superficial lymphatic glands were enlarged. The heart was not obviously hypertrophied. The urine showed nothing abnormal, excepting a trace of albumin. The blood urea (October 7th, 1927) was 0.04 per cent. The blood serum gave a negative Wassermann reaction. The faeces were examined for occult blood, with nega-

tive result. The patient was edentulous. The menopause had occurred about September, 1926, but since then there had been occasional irregular very slight losses. Like some of her relatives, she had been subject to occasional migraine-like attacks. The nervous reflexes were normal. There were no retinal haemorrhages or optic neuritis.

Treatment by a restful life and a non-stimulating diet, with occasional blood-letting, led to temporary apparent improvement. In March, 1928, however, mental confusion developed, together with some loss of power in the right hand and occasional diplopia. From the beginning of April until her death the patient was troubled by recurrent pain and distension in the abdomen, sometimes with a few days' pyrexia. A small tender mass was felt low in the left iliac fossa, and partial obstruction, probably due to diverticulitis in the pelvic colon, was diagnosed (F. J. B.). The rectum was always empty and ballooned. Improvement in the mental and parietic conditions took place for a time. Early in June, 1928, the little toe of the right foot became gangrenous, and she died on June 17th, after having been more or less unconscious for two days. The last brachial systolic blood pressure taken (May 30th) had been 260 mm. of mercury. The maximum registered in her case was 280 mm. The last blood count had been on May 23rd, when the erythrocytes numbered 8,900,000 per c.mm. (the highest erythrocyte count noted in this case), and the white cells were 26,000 per c.mm.; the haemoglobin was 110 per cent.

For an account of the necropsy and the microscopic examination we are indebted to Dr. W. E. Carnegie Dickson, who made a most careful *post-mortem* examination, about thirty-six hours after death. We have confined ourselves to what appear to be the essential points.

There was gangrenous discoloration of the right small toe. The subcutaneous and extra-peritoneal fat was reduced to a minimum, and had a congested appearance, the blood vessels, especially the larger veins, being very prominent and full of rather thick, viscid, and somewhat dark blood.

The heart weighed 250 grams (9 ounces), and though the whole organ was rather small the left ventricle was relatively hypertrophied. There was considerable coronary sclerosis, very little atheroma of the arch and descending thoracic aorta.

The lungs were somewhat emphysematous, and showed hypostatic congestion.

The liver was comparatively small, weighing

990 grams (35 ounces), and was not cirrhotic. Microscopically there was no leukaemic infiltration. The reticulo-endothelial cells appeared rather diminished than increased in numbers, and very inconspicuous.

The gall-bladder contained a little viscid orange-coloured bile, and 32 smooth, rounded, or slightly faceted cholesterol gall-stones, varying in size from small peas to thick beans.

The spleen was considerably enlarged, weighing 480 grams (17 ounces), and measuring 6½ by 3½ by 1½ inches. It was free from areas of infarction, and microscopically (see Fig. 2) showed general fibrosis of the reticulum, the Malpighian bodies (lymphatic nodes) having practically completely disappeared. It was congested with blood, and there were small areas of diffuse haemorrhage. The endothelial cells of the sinuses, and also of the reticulum, were comparatively small in size, and there was very little evidence of phagocytosis and destruction of erythrocytes.

The left kidney was small and macroscopically and microscopically fibrosed; it weighed about 40 grams (under 2 ounces). There were three small calculi in the pelvis, the walls of which were

FIG. 1.—Microscopic appearance of the bone marrow from the centre of the shaft of the right femur. ($\times 400$)

^{*} Cf. F. Parkes Weber, *British Medical Journal*, 1925, i, p. 1108, and 1927, ii, p. 98; *Lancet*, 1925, ii, p. 1138.

thickened. The *right kidney* weighed 127 grams ($4\frac{1}{2}$ ounces), and microscopically the larger arteries (interlobular, etc.) showed considerable thickening of both middle and inner coats, whilst the glomeruli showed hyaline swelling of their capillaries. There were decided degenerative and necrotic changes in the epithelium of the convoluted tubules, which were, however, doubtless "terminal" in nature, or rather, of *paulo-ante-mortem* and partly *post-mortem* origin.

There were *old fibrous adhesions of the great omentum*, and chronic adhesions of the left broad ligament and Fallopian tube to the omentum and left wall of the pelvis. The sigmoid flexure and the rectum were much elongated and convoluted. There was excessive sacculation in the pelvic colon. The caecum was filled with pultaceous faeces, and the transverse colon, sigmoid flexure, and upper part of the rectum contained marble-like scybalous faecal masses, and the mucous membrane showed catarrhal changes.

The *uterus* was small and atrophied, and on its anterior surface an old, partially calcified, small fibromyoma projected. The *ovaries* showed marked fibrotic atrophy.

Bone Marrow.—The bone marrow of the shaft of one femur which was examined was red, as also was that of the cancellous bone and of the ribs. Microscopic examination of the bone marrow showed great excess and activity of erythroblastic tissue, and also, to a lesser extent, excess of leucoblastic tissue. There were small areas of diffuse haemorrhage. A striking feature of all the bone marrow preparations was the unusually large number of giant cells of the megakaryocyte class (see Fig. 1). An equally striking feature was the relative absence of evidence of phagocytosis and processes of blood destruction by the endothelial cells.

There was nothing of importance to note in regard to the pancreas, thymus, thyroid, pituitary gland, and pineal body. The calvarium was of medium thickness, but very compact and relatively heavy. There were arterio-sclerotic changes in the cerebral arteries, and terminal local thrombosis in some of the vessels, with consequent softening of the right supramarginal and angular gyri. There were, as above mentioned, arterio-sclerotic changes in many of the medium-sized visceral arteries, especially coronary, renal, and cerebral, and the gangrene of the left small toe was probably of arterio-sclerotic origin.

REMARKS.

The high blood pressure during life in this case of erythraemia was explained by the renal and vascular changes demonstrated at the *post-mortem* examination. It is clear that the blood destruction in the spleen and bone marrow, as evidenced by the phagocytosis of erythrocytes, etc., was not relatively excessive. But even if blood destruction in most cases of erythraemia were relatively deficient, the abundant and invariable evidence of greatly excessive erythrocyte formation shows that deficient erythrocyte destruction (as has been long recognized) is not the main cause of the polycythaemia rubra. Nevertheless, as Dr. Carnegie Dickson points out to us, the relatively deficient blood destruction in the present case might well be contrasted with the evidence of excessive destruction of erythrocytes in the bone marrow and spleen (reticulo-

endothelial system) of, let us say, typical cases of pernicious (Addisonian) anaemia. In regard to the great excess of megakaryocytes in the bone marrow, it is rather unfortunate that no exact count of the thrombocytes (blood platelets) in the circulating blood of the present case was made. In ordinarily stained blood films the thrombocytes did not appear to be in excess.

We are indebted for the illustrations to the kindness of Dr. Carnegie Dickson. Fig. 1 is to illustrate the microscopic appearance of the bone marrow from the centre of the shaft of the right femur, where in ordinary persons there is only fatty marrow. It shows excessive erythroblastic activity, the cells being mostly erythroblasts. To a lesser extent there is evidence of leucoblastic activity, but this was more obvious in the rib marrow. An especial feature is the large number of megakaryocytes, which were scattered throughout the sections of the marrow in both femur and rib. Fig. 2 is from the spleen; it shows fibrosis of the reticulum, with more or less disappearance of the lymphocytes of the Malpighian corpuscles and pulp.

FIG. 2.—From the spleen; showing fibrosis of the reticulum, with more or less disappearance of the lymphocytes of the Malpighian corpuscles and pulp. ($\times 400$.)

The engorgement with blood was better shown in other sections. In both the bone marrow and the spleen there is no evidence of any excessive destruction of erythrocytes.

PARAPHIMOSIS OF THE CLITORIS.

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On September 18th, 1928, I was asked by Dr. Hepplewhite of Wallsend-on-Tyne to see in consultation with him a female child, aged 9 years and 4 months, who had a painful swelling at the vulva. Five days previously when out for a walk with an aunt it was noticed that the child was limping, but she denied that anything was the matter. On the day before I saw her she complained for the first time of pain, and only then was she examined, and a painful swelling in the region of the clitoris was noticed, but nothing was done until the next day, when Dr. Hepplewhite was called in.

I was informed that the child was a devotee of a "fairy cycle," and the parents thought that the child had probably injured herself by the saddle when riding this. Dr. Hepplewhite made a diagnosis of paraphimosis of the clitoris, a diagnosis which proved to be correct.