

Chronic purpura of two years' duration connected with malignant endocarditis : wrongly supposed to be connected with splenic anaemia / by F. Parkes Weber.

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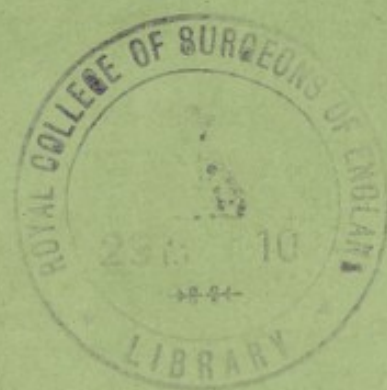
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CHRONIC PURPURA OF TWO YEARS' DURATION CONNECTED WITH MALIGNANT ENDOCARDITIS, WRONGLY SUPPOSED TO BE CONNECTED WITH SPLENIC ANÆMIA.

By F. PARKES WEBER, M.D., F.R.C.P.,
Physician to the German Hospital, London.

THE patient was a sailor, aged 28 years, who had visited South Africa several times, and had often been in the Mediterranean and the surrounding countries. He had had a short attack of jaundice without colic about five years ago. About a year later he contracted malaria when in the Straits Settlements, and he had had two slight recurrences since then. He stated that he had had gonorrhœa twice (when 20 and when 21 years of age), but never syphilis. In the harbours he used to drink much spirits and beer. He married at 24 years, and had one child, who was living and healthy, aged 2 years. His parents were said to be healthy, and there was nothing special in his family history.

He thought that the present illness had commenced about two years ago, when purpuric spots appeared on both legs. From that time he had gradually become weaker and paler, and had noticed an increasing swelling (evidently the spleen) in the left side of his abdomen. There had been occasional oozing of blood from the gums and slight epistaxis. He had been treated at several London hospitals, where apparently the diagnosis of splenic anæmia was made.

I am much indebted to Dr. H. G. Turney and Dr. G. G. Butler for notes of the case when he was at St. Thomas's Hospital, August to October, 1908. His spleen was then much enlarged, its lower limit reaching a level of half-way between the umbilicus and pubes, the splenic dulness extending upwards to the seventh rib in the axillary line. The liver was definitely enlarged. The apex-beat of the heart was felt in the fourth intercostal space, $\frac{1}{2}$ in. external to the left nipple

line. The cardiac dulness reached from the apex-beat to the mid-sternal line and upwards to the third space. There was a fairly well-marked apical systolic murmur conducted into the axilla, and there was likewise a systolic murmur at the base best marked over the "pulmonary area." Treatment by arsenic was continued throughout his stay at St. Thomas's Hospital. Blood-examinations gave the following results :

	August 14th, 1908.	September 23rd, 1908.	October 8th, 1908.
Red cells (per c.mm. of blood)	5,625,000	3,600,000	3,268,000
Hæmoglobin	50 per cent.	50 per cent.	60 per cent.
Colour index	0·4	0·7	0·9
White cells (per c.mm. of blood)	2840	1740	1360
<i>Differential count of white cells.</i>			
Neutrophile polymorphonuclears	27 per cent.	28 per cent.	26 per cent.
Small lymphocytes	58 per cent.	57·5 per cent.	41·5 per cent.
Large lymphocytes	8·5 per cent.	9·5 per cent.	27 per cent.
Large hyalines	5·5 per cent.	4·5 per cent.	5 per cent.

In the German Hospital (admitted on July 1st, 1909) the patient was extremely pale, weak, and wasted. Both legs below the knees and the lower inner aspects of the thighs were covered with spots of brown pigment. This remarkable brown pigmentation was evidently the result of repeated spots of hæmorrhage into the skin, for amongst the pigment-spots were some fresh red petechiæ. On the arms and trunk there were likewise a few petechiæ. The ankles were œdematous. There was a tendency to bleed from the gums. The heart was somewhat enlarged, the impulse was forcible, and the apex-beat could be located in the fifth intercostal space just outside the nipple line ; there was a systolic apical murmur. On auscultation over the lungs a few dry bronchitic sounds were heard. The spleen was greatly enlarged and hard ; it reached to the umbilicus on the right, and downwards into the inguinal region. The liver was likewise enlarged and hard, its lower edge being felt at the umbilical level. Ophthalmoscopic examination showed a few retinal hæmorrhages (Dr. R. Gruber), and there were some conjunctival petechiæ. The urine contained a little albumen, some granular and epithelial tube-casts, and afterwards a little blood and some blood-casts. There was no evidence of active malaria.

Blood-examinations :

	July 1st and 2nd, 1909.	July 19th, 1909.
Red cells (per c.mm. of blood)	1,700,000	1,700,000
Hæmoglobin	30 per cent.	35 per cent.
Colour index	0.9	1.03
White cells (per c.mm. of blood)	1900	1200
<i>Differential count of white cells.</i>		
Neutrophile polymorphonuclears	63.5 per cent.	58.0 per cent.
Lymphocytes	29.5 per cent.	26.0 per cent.
Intermediates	4.0 per cent.	8.0 per cent.
Large hyalines	2.5 per cent.	7.0 per cent.
Eosinophiles	0 per cent.	1.0 per cent.
Mast-cells	0.5 per cent.	0 per cent.

For these differential counts I am indebted to the kindness of Dr. A. E. Boycott, who added that no nucleated red cells were found. Amongst the erythrocytes were oval forms. Very little polychromatophilia. The coagulation-time of the blood, as estimated on July 21st by Dr. Engelsmann with Sir A. E. Wright's coagulometer, was as much as about thirty minutes.

I accepted "splenic anæmia" as the clinical diagnosis, and the patient was treated with an arsenic preparation. He likewise received orange juice, but on July 19th I prescribed calcium lactate on account of the hæmorrhagic tendency. The pulse was usually 100 to 110 per minute. The systolic brachial blood-pressure (July 26th) was 105 mm. Hg. During the first ten days in the hospital his temperature was often raised (up to about 101° F.), but after July 10th there was seldom any fever. On the day before the patient's death, which occurred on July 29th, there was, however, again slight fever. Towards the end there was considerable œdema, with bronchitic signs and œdema.

NECROPSY (ABOUT HALF-A-DAY AFTER DEATH).

The *brain* (weight, 51 oz.) showed nothing special, except the small cystic remnant of an old hæmorrhage above the front part of the internal capsule on the left side.

The *heart* was large, weighing 21 oz., and the left ventricle was hypertrophied. The cardiac musculature was very pale. There was old thickening of the mitral valve, and adherent to one part of the valve was a soft, organised thrombus of about the size of a hazel-nut.

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In the left auricle, and apparently growing from the inter-auricular septum, was a large polypoid excrescence, which measured about 70 mm. in length and about 35 mm. in breadth. It was attached by a short, circular pedicle, about 7 mm. in diameter, to a spot on the inter-auricular septum,* about half-way between the thrombus on the mitral valve and the (closed) foramen ovale. The free end of this polypoid excrescence, which was evidently an old organised thrombus, was opaque, white and hard, its surface feeling as if it were partially calcified. No other valvular or endocardial disease was observed.

The lungs showed some recent bright-red pleural petechiæ at their bases. There was a moderate amount of serous effusion in the left pleura.

There was likewise some ascites.

The spleen was very large, and, together with a small splenulum, weighed 110 oz. There were several subcapsular (non-suppurative) anæmic infarcts, some of which were connected by peri-splenic adhesions to the surrounding parts. On section the substance of the spleen (and also that of the splenulum) had a milky-red appearance and was rather friable.

The liver was large and hard, weighing 156 oz. The capsule was roughened at parts, and there were some peri-hepatitic adhesions. There were many bright-red recent petechiæ in the capsule. On section the substance of the liver suggested fatty infiltration and chronic passive congestion, together with some increase of fibrous tissue. The gall-bladder and bile-ducts showed nothing special.

The kidneys (weight together 17 oz.) were large and rather pale; the cortical substance was not diminished and the capsules stripped readily. Petechiæ were noted in both renal pelves. In the right kidney there was a non-suppurative embolic infarction.

There were petechiæ in the mucous membrane of the stomach, ileum, and cæcum, confluent in part of the ileum and cæcum. Some of the lymphatic glands near the hilum of the spleen were enlarged. The splenic and portal veins were very large, but otherwise nothing

* It is interesting to note how frequently in cases of chronic malignant endocarditis the polypoid or warty excrescences are attached, not to the valves, but directly to the auricular or ventricular walls. Similar excrescences are sometimes found attached to the walls of the aorta or pulmonary artery beyond the valves (*vide* Fürth and F. Parkes Weber, "A Case of Malignant Pulmonary Arteritis after Gonorrhœa," *Edin. Med. Journ.*, July, 1905, p. 33).

abnormal was found in the abdominal veins or arteries. The bone-marrow of the shaft of the left humerus was examined and found to be quite red.

MICROSCOPIC EXAMINATION.

For this I am indebted to Dr. J. C. G. Ledingham, who examined sections of the spleen, liver, kidney, and a piece of the bone-marrow from the middle portion of the shaft of the left humerus. He kindly gave me the following report:

Spleen.—The capsule is considerably thickened, and there is some fibrosis of the splenic reticulum immediately subjacent. Otherwise there is little evidence of increase in the trabecular system except in the adventitial sheaths of the follicular arteries (? so-called "fibro-adénie" of Banti). A few Malphigian nodes remain, and in these the lymphoid cells are quite healthy. No karyorrhectic foci present. In the spleen-pulp, which shows much congestion, are large numbers of plasma-cells, lying mainly in rows or in small groups in the neighbourhood of larger trabeculae and the adventitial sheaths of the follicular arteries. These plasma-cells are evidently taking part in the new fibroblastic formation pervading the pulp and proceeding from the margins of the larger trabeculae. There are a few small foci of normoblasts. The polynuclear cells in the pulp are much increased, but there are no purulent foci. Megakaryocytes are very numerous. No micro-organisms seen.

Liver.—The most marked features are the cellular infiltrates in the portal spaces. There is no coarse fibrosis of the organ, but around each portal vein in the portal space there is thickening of the adventitia, with prolongations extending outwards and enclosing in its meshes young fibroblasts and large numbers of plasma-cells. In some parts this plasma-cell infiltrate extends slightly into the adjoining lobule (this condition is essentially analogous to that in the spleen-pulp). There is some "cyanosis" of the organ, with fatty infiltration. The inter-acinous capillaries contain fairly numerous polynuclear cells and occasionally megakaryocytes.

Kidney.—There is no coarse cirrhotic process, but in some parts marked fibroblastic proliferation is present, causing atrophy of groups of tubules. Plasma-cells are also present, namely, in the neighbourhood of the adventitial sheaths. No micro-organisms seen.

Bone-marrow.—This tissue is extremely cellular and the fat-elements have disappeared. The cells are mostly plasma-cells, with small and large lymphocyte forms. Many mitoses are seen in the plasma-cells. Giant-cells are very numerous.

All four organs (spleen, liver, kidney, and bone-marrow) present as their most conspicuous feature a plasma-cell development, with the elaboration of new fibroblastic tissue.

REMARKS.

In the first place it is interesting to know that the chronic purpura of two years' duration, which produced the remarkable brown pigmentation of the patient's legs, was due to a form of chronic malignant endocarditis. This form of endocarditis has been well described recently by W. Osler* and T. J. Horder† in the *Quarterly Journal of Medicine*. The pathogenic micro-organism which caused the disease in the present instance was not discovered.

The marked leucopenia and chronic splenomegaly seemed to point to a diagnosis of splenic anæmia or Banti's disease, but the post-mortem examination showed that the fatal illness (and the cause of the purpura and cutaneous pigmentation) was really the cardiac disease. Leucopenia is indeed no longer regarded as pathognomonic of splenic anæmia (or Banti's disease), for it is not rarely present in various conditions associated with chronic enlargement of the spleen; for instance, in some cases of chronic splenomegaly from malaria. In the present case the leucopenia appears to have preceded the actual anæmia (oligocythæmia), as far as one can judge by the blood-count taken on August 14th, 1908.

I am not sure that splenic anæmia (of adults) should be regarded as a distinct disease. Certainly I have not seen any case myself which has been convincing in this respect. Perhaps "splenic anæmia" is nothing more than a group of symptoms which may be set up by various pathogenic agencies. Not only are there cases of congenital or acquired chronic acholuric jaundice (in which the jaundice is very slight‡) with splenomegaly and anæmia, which might be wrongly

* *Quart. Journ. Med.*, Oxford, 1909, vol. ii, p. 219.

† *Ibid.*, vol. ii, p. 289.

‡ Occasionally (at least for a time) no obvious jaundice is observed in some of these cases.

regarded as examples of "splenic anæmia"; but in many cases of supposed splenic anæmia and Banti's disease a necropsy has given evidence of considerable vascular or endocardial disease, very probably of infectious origin. Thus, in one of the earlier fully described cases in England, that published by Dr. S. West* (at the post-mortem examination on which I happened to be present), there was evidence of considerable old disease of the aortic valves of the heart, and the patient had a septic pyrexia at the end. In several cases of "splenic anæmia" thrombotic obstruction of the splenic and portal veins (cases of Dock and Warthin, 1904, Oettinger and Fiessinger, 1907, F. Dévé, 1908, etc.) or disease of the splenic artery has been found.

* *Med.-Chir. Trans.*, London, 1896, vol. lxxix, p. 323.

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