A CASE OF POLYARTERITIS ACUTA NODOSA.¹

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(PLATES XXIV.-XXV.)

THE name "polyarteritis acuta nodosa," is one suggested by Ferrari for a group of cases, in which there are, at an early stage, small localised nodules on the walls of the small arteries and arterioles, with later widespread degeneration and aneurysmal dilatation of these arteries. The condition is an interesting one, not only on account of its rarity, but also because of its unknown etiology, and because of the difficulty in interpreting the nature of the lesions found.

The following is a short history of the case which has come under our observation:—

W. B., a cutler, æt. 19 years. There was nothing of note in his family history. He was a teetotaler, smoked very little, had not had any venereal disease, and up to the commencement of this illness had enjoyed good health. He first complained of pain in the head, back, and limbs after a thorough soaking by rain last November. He continued at work for a week, but was then compelled to take to bed. At first he was considered to be suffering from influenza, but later he became worse, vomited several times, complained of pains in the chest and abdomen, and was slightly delirious at night. Enteric fever was suspected, and although the "Widal" reaction was negative, the patient was sent to the Sheffield Fever Hospital on 18th December 1911.

On admission the patient was found to be very thin. The skin was pale and slightly yellow. The tongue was thickly furred with a white, moist, easily detached fur, which extended on to the soft palate and tonsils. The pupils were equal and active. The glands were not enlarged. There were a few sibilant sounds on the left side of the chest, but no dulness. The heart sounds were quite clear. The abdominal wall was rigid, and the patient complained of pains when pressure was made on any part of the abdominal wall. The abdominal reflex was abolished. There were a few hæmorrhagic spots on the anterior surface of the legs. The temperature was rarely over 100° F. The respirations were about 24 per minute, and the pulse-rate varied from 82 to 118. The condition gradually improved. The temperature came down to normal, and the patient was feeling better, although occasionally he had attacks of abdominal pain, vomiting, and diarrhoea. Early one morning, after

¹ Received August 3, 1912.

a comfortable night, he awoke with acute pain in his abdomen. He rapidly grew very ill, became collapsed and died in a few hours.

The autopsy was performed by one of the resident officers. Unfortunately full notes are not available, and only the heart, liver, spleen and kidneys were sent for examination.

NAKED-EYE CHARACTERS.—The *heart* was not enlarged. The pericardium showed no thickening, and there were no adhesions. The branches of the coronary arteries were very distinct, standing out markedly all over the surface of the heart, with here and there small dark-coloured dilatations (Plate XXIV. Fig. 1). The vessels could be felt like knotted cords. On cutting in to them, their walls were seen to be much thickened, with here and there small aneurysmal dilatations, filled with dark thrombi. The heart muscle was firm, and scattered throughout it were light patches and streaks. There was no endocarditis. The valves were competent and the cavities showed no change of any note. The aorta showed a few minute streaks of fatty change, otherwise it appeared normal. The openings of the coronary arteries were healthy, and the arteries showed no thickening for about the first inch of their length.

The *liver* was not enlarged. There was some slight congestion and some cloudy swelling. There was a great increase of the fibrous tissue in a number of the portal spaces. In some the arteries were greatly thickened, while a number showed aneurysmal dilatation and were filled with dark thrombi.

The *spleen* was enlarged and markedly congested. The arteries did not show such definite thickening as those of the other organs, but careful examination indicated that the smaller arteries were thickened, and had multiple minute nodules in their walls.

The *right kidney* showed a remarkable, large, firm, bright-red hæmorrhage under the capsule, which compressed the kidney substance (Plate XXIV. Fig. 2). The arteries showed naked-eye changes, similar to those in the heart and liver—thickening and aneurysmal dilatation. Scattered through the kidney were numerous pale infarcts of varying size, each corresponding to the area supplied by a thrombosed artery.

The *left kidney* was not enlarged. Here also the arteries in the intermediate zone were affected, showing numerous aneurysms, the largest being almost half an inch in diameter. There were also numerous pale infarcts.

MICROSCOPICAL EXAMINATION.—The most marked changes were in the walls of the small arteries, of the size of the branches of the coronary artery of the heart. The larger arteries were not affected. The appearances differed greatly in different vessels. In this case the earliest changes were seen in the small arteries of the spleen (Plate XXIV. Figs. 3 and 4). The coats of the vessels were best differentiated by Weigert's elastic fibre stain and picro-fuchsin. In the section of a nodule on a small artery, it is seen that the muscle coat at that place has entirely disappeared. The internal elastic lamina is ruptured, and can frequently be seen curling round the broken-off end of the middle coat. The nodule consists of young granulation tissue which extends into the adventitia, and in some sections it can be traced to the lumen of the vessel, from which young blood vessels appear to be invading the inflamed area (Plate XXV. Fig. 5). In a few of the most acute areas, fibrin deposit can be made out and leucocyte infiltration of the adventitia, and the perivascular lymphatics are seen to be filled with leucocytes.

In the older lesions there is invariably great thickening of the intima, some of the smaller arterioles being almost completely blocked. In some arteries the internal elastic lamina is breaking up and the middle coat has entirely disappeared. Hyaline degeneration is very commonly seen, but there is no fatty and no amyloid degeneration and no calcification.

The aneurysms are formed in one of two ways.—(1) Most commonly after the degeneration of the coats of the artery the vessel dilates with the forma-

tion of a fusiform aneurysm at the weakened part (Plate XXV. Fig. 7); (2) at a weakened part of the vessel wall the aneurysmal sac protrudes and dissects its way in the outer coat (Plate XXV. Figs. 8 and 9).

Microscopical examination further confirmed the nature of the secondary changes in the organs. At places in the heart there was extreme chronic myocarditis. The muscle fibres showed, at places, slight fatty degeneration. In the aorta there were patches of early fatty change in the intima. Infarcts were seen in the kidney and marked congestion in the spleen.

A large number of sections were stained for organisms by various methods, but after careful examination none could be found. Sections stained for the *Spirochaeta pallida* also gave a negative result. A pure culture of streptococci, however, was obtained from the renal hæmorrhage, but we have no definite evidence connecting this organism as a causal agent. Unfortunately no cultural examination of the blood was made during life. The history of the case was that of an infective disease, and the finding of a streptococcus in the extravasated blood round the kidney, very soon after death, is certainly suggestive.

CONCLUSIONS.

The literature has been well reviewed by Carnegie Dickson (1907¹) and by Longcope (1908²). Most observers are agreed that this disease is due to some, as yet unknown, toxic substance, probably of organismal origin, and also that it is an acute local inflammation attacking the vessel walls. No definite source of infection has so far been proved. Syphilis has been present in the history of a number of cases, but not in all, and some observers consider that it could be absolutely excluded in their cases. In our case syphilis was denied, and it is very improbable that the patient's statement was incorrect.

The disease is an acute one, as evidenced by the histories and by the nature of the lesions. Our patient died eight weeks after the earliest symptom, and even then none of the nodules present were at so early a stage as is described by some writers. The nodules appear to be healing in our case. From a consideration of the earliest nodules it is impossible to say in which coat the first affection took place, though the first destructive changes appear in the middle coat. It is possible that the primary inflammation is in the outer coat, carried by the vasa vasorum or lymphatics, but the middle coat is attacked at an early stage. The thickening of the intima is a later process and is probably to some extent protective.

In some cases early changes in the adventitia seem to be the most marked feature, and Carnegie Dickson, holding that there are two distinct diseases, has classified the recorded cases into two groups—(1) true periarteritis, the majority of which are syphilitic in nature; (2) polyarteritis acuta nodosa, into which class this case would fall.

In conclusion, we have to thank Dr. Egerton Williams of the City Fever Hospital, Sheffield, for allowing us to make use of the clinical notes of this case.

REFERENCES.

1. DICKSON, CARNEGIE . . . *Journ. Path. and Bacteriol.*, Cambridge, 1907, vol. xii. p. 31.
2. LONGCOPE *Bulletin of the Ayer Clinical Laboratory of the Pennsylvania Hospital*, No. 5, December 1908.

DESCRIPTION OF PLATES XXIV.-XXV.

PLATE XXIV.

- FIG. 1.—Outer wall of right ventricle, showing the thickened branches of the coronary artery with aneurysmal dilatations.
- FIG. 2.—The right kidney shows a large hæmorrhage. Several of the arteries in the intermediate zone show aneurysmal dilatations, and there are also several infarcts.
- FIG. 3.—Transverse section of a small artery in the spleen. It shows, at one point, disappearance of the middle coat, rupture of the internal elastic lamina, the ends of which curl outwards, thickened intima and nodule of granulation tissue. Haem. Weigert's elastic fibre stain, van Gieson. ($\times 50$)
- FIG. 4.—Longitudinal section of artery in the spleen shows degeneration of media and rupture of internal elastic lamina. Haem. Weigert, van Gieson. ($\times 50$)

PLATE XXV.

- FIG. 5.—Transverse section of coronary artery of the heart. A portion of the media, which has given way, is seen at the lower right-hand corner. The newly formed vessels are seen passing in from the lumen of the artery. Haem. Eosin. ($\times 300$)
- FIG. 6.—Longitudinal section of the coronary artery of the heart. Intima greatly thickened. Internal elastic lamina and media degenerating and breaking up. Haem. Weigert, van Gieson. ($\times 50$)
- FIG. 7.—Artery of kidney, dilated and filled with blood clot. Haem. Eosin. ($\times 50$)
- FIG. 8.—Coronary artery of heart. Transverse section showing commencement of small saccular aneurysm. The walls of the artery are completely disorganised at this part. Haem. Weigert, van Gieson. ($\times 50$)
- FIG. 9.—Coronary artery of heart. Transverse section, a short distance away from the last, showing the aneurysm dissecting its way in the outer coat. Haem. Weigert, van Gieson. ($\times 50$)

FIG. 1.

FIG. 2.

FIG. 3.

FIG. 4.

FIG. 5.

FIG. 6.

FIG. 7.

FIG. 8.

FIG. 9.