Original Articles.

A CASE OF MORBUS MACULOSUS WERLHOFII.
(PURPURA HÆMORRHAGICA. PSEUDO-
RHEUMATISM).¹

BY W. E. BOARDMAN, M. D.

ON April 17th I was requested to visit Mr. C., of this city. A stranger to me, he showed himself to be a man of considerable intelligence, of excellent physique, well formed, of medium height, good weight, and nervous temperament. Upon inquiry I learned that he had always enjoyed excellent health, had been accustomed to observe unusual regularity in all his habits of life, and had never been addicted to the use of stimulants or tobacco in any form. His occupation was such that he spent most of the daytime in the open air and required considerable active exercise. He was nicely housed, and in comfortable circumstances in all respects. So far as could be ascertained at the time there appeared to be no hereditary, congenital, or acquired disposition to disease in any form.

He had experienced his usual excellent health until a few days or a week prior to the date upon which I was consulted for the first time. During that interval he had complained of some languor and inability to attend properly to his customary routine duties. Without experiencing any chill or even any sensation of chilliness, observing no symptoms of fever, he soon noticed pain in his ankles, followed in a day or two by tenderness and swelling of these joints. Subsequently both knee joints became similarly affected. When he first came under my observation, he was in bed, pulse 100, temperature 100.6° F., respiration normal. General appearance somewhat pale and careworn, considered to be due, in a great measure, to loss of sleep and anxiety on account of the interruption of his business, in addition to the pain from which he had suffered. There had been sleeplessness, loss of appetite, and general deterioration of strength, but the ordinary functions appeared to have been well performed. The before-mentioned joints were painful on pressure, somewhat swollen, and slightly reddened. The surface of the body generally was warm, but not moist. The lower extremities, below the knees, were cooler to the touch than the rest of the body. Examination of the thoracic and abdominal organs showed nothing abnormal. No analysis of the urine was made at the time, but inquiry elicited nothing unusual concerning it. The diagnosis of rheumatism, probably subacute, was readily made. I recommended that the affected joints should be enveloped with cotton batting, and prescribed salicylic acid in the usual way. On the following day I learned that my directions had been carefully observed, that the ordinary effects of the acid had been experienced during the night, that the pains had been relieved considerably, and that he had had some sleep; pulse 98, temperature 98.6° F., respiration easy and regular, heart sounds normal. I advised a continuance of the acid for a day or two, five grains every six hours, and also of the cotton wool dressings, to be replaced subsequently by flannel, and also a resumption of ordinary diet.

April 21st, three days later, I was requested to visit the patient again, when I learned that, during the preceding night, there had occurred what he considered

to be a return of the symptoms which had existed at my first visit. I found the joints presenting about the same appearance as before, and the temperature had again increased to 100.4° F. The acid had been stopped the preceding morning. I advised its resumption, and gave general directions similar to those given at my first visit.

I heard nothing more from my patient until May 2d, eleven days later, when I was summoned to him in great haste to relieve a very profuse and obstinate epistaxis, which already had continued several hours, and had become more violent in spite of the vigorous use of ordinary household remedies. Upon my arrival, the blood was flowing freely from both nostrils and the mouth. The patient's face was blanched, the general surface, as seen at a hasty glance, was pale. Both lower extremities, below the knees, were enlarged, apparently œdematous. The wrists and hands were in a similar condition. These parts, especially the lower extremities, were thickly studded with purpuric spots, some of them the size of a bean, or a little larger. The pulse was rapid and weak, but its exact frequency was not determined. I resorted to compression of the facial artery and to the use of various styptics, and finally succeeded in controlling the hæmorrhage by cotton wool tampons soaked in a solution of subsulphate of iron. In the mean time I was informed that since my previous visit the condition of the patient had improved somewhat, that is, the pains had subsided and his strength had returned to some extent. He had been able to sleep tolerably well, especially in the sitting posture. The swelling of the joints, however, had continued without perceptible change (they had not noticed the enlargement, apparently œdematous, of the lower limbs and hands), and having misunderstood my directions, he had continued to take the salicylic acid during the interval for the purpose of reducing the swellings. As near as I could estimate, he must have taken in all about three hundred grains.

No immediate cause could be assigned by the patient or his wife to explain the hæmorrhage. Upon inquiry I was unable to learn that any of his family were "bleeders," or that he had been liable to attacks of hæmorrhage of any kind. He had never had a tooth extracted, and the only fact of any importance in this direction which I did ascertain was that he never made use of a stiff tooth-brush, because it was liable to make his gums bleed a little. Before leaving him I examined him quite carefully. The pulse was rapid and weak, but regular. The respiration was a little hurried and somewhat labored. Nothing abnormal could be detected in the thoracic or abdominal cavities. The joints presented about the same appearance as at my first visit, the lower limbs pitted on pressure, as also did the hands, and the purpuric spots were present, as they have before been described.

I ordered tincture digitalis and ergot with perfect rest and simple, unstimulating diet.

On the following morning I was again summoned in haste, on account of a recurrence of the epistaxis. Upon my arrival I found the blood issuing copiously from the mouth, obviously from the posterior nares, and oozing from the gums, which, however, on being cleansed, displayed the clear, glistening membrane peculiar to the tissues of the oral cavity, without any appearance of sponginess or inflammation. The blood was also making its way outside the styptic tampons, which were removed and fresh ones substituted; but

¹ Read before the Boston Society for Medical Improvement, November 28, 1881.

these did not suffice to arrest the hæmorrhage, and finally I was obliged to plug the nasal cavities, posteriorly and anteriorly. This procedure, together with styptic applications to the gums, checked the bleeding. During this visit I learned that in the night there had been a constant slight flow of bloody fluid into the mouth, that the patient had two evacuations of the bowels, consisting almost entirely of blood, some of it dark and some light red in color, and but little clotted, a feature which had been noticed with the preceding hæmorrhages.

At this time the pulse, of course, was still rapid and weak, but not fluttering. There was considerable dyspnœa, but the respiratory sounds were clear and distinct. The heart's action was rapid and weak, but no other abnormal conditions could be detected. In the evening of this day, as on the following day, the patient's condition was fair. The oozing continued to some extent from the posterior nares, and he had other bloody stools. The urine examined on this day had a specific gravity 1016, presented a trace of albumen, and, under the microscope, numerous blood corpuscles, but no casts. The abdomen, too, presented a uniform enlargement, mostly resonant, or, rather, I was unable to detect any fluid, but for this purpose I was unwilling to change the position of the patient through fear of again starting hæmorrhage.

On the following morning his condition was about the same, though he thought he had a little more strength. His mind continued clear, as it had been throughout the illness. He complained only of fatigue, weakness, and the discomfort occasioned by the plugs. I still thought it prudent to allow these to remain, this caution being emphasized by the slight increase in the amount, and intensity of color, of the oozing blood.

At the morning visit of the next day I feared it would be necessary, before long, to repeat the process of plugging the nares, but concluded to postpone it, if possible, till the afternoon. At noon he suddenly became worse, with faintness, great dyspnœa, ending in collapse and death, after a brief interval, before a physician could reach him.

Upon my arrival I learned that the oozing had not increased much since the morning visit, nor had any new hæmorrhage been observed from other localities. Upon examination of the body externally, I found nothing additional to what has already been related, except that the abdominal enlargement seemed to be considerably greater, and percussion yielded a general dullness, but no wave could be determined. There was no autopsy.

Primary diagnosis, subacute rheumatism; final diagnosis, morbus maculosus Werlhofii, purpura hæmorrhagica, sometimes termed pseudo-rheumatism.

Cause of death, asthenia from external and internal hæmorrhage.

This case surely possesses the element of rarity, and therefore has some interest. I have presented it somewhat in detail with the view of eliciting discussion as to the correctness of my diagnosis, which has been questioned by some of my professional friends, and that I may, perhaps, receive suggestions as to treatment which might be valuable to me under similar circumstances, should they recur.

A brief recital of the more salient points connected with the disease, more familiarly known under the name purpura hæmorrhagica, will serve to confirm the correctness of my view of this case.

According to the most recent authorities, the term morbus maculosus is applied to a rare form of hæmorrhagic disease, which is characterized by the fact of its acquired, transitory nature and its apparently spontaneous development, its ætiology being still buried in obscurity. It is distinguished from the more common disposition to hæmorrhage, which is frequently observed as an accompaniment or a sequel of several acute and chronic diseases, and also from those affections in which the hæmorrhagic diathesis is hereditary. In the former cases the hæmorrhagic tendency is symptomatic merely, while the affection under consideration is regarded as an *essential disease*, which is developed suddenly in a previously healthy individual who possesses no hereditary disposition thereto. Some cases present a mild febrile stage, while others are unaccompanied by any febrile symptoms. Again, in some cases the fever is initiatory, while in others it occurs sooner or later after extravasation of blood takes place. Oftentimes the disease is ushered in by certain local disturbances, either with or without fever, which sometimes persist during the continuance of the hæmorrhages. The more common of these are pain and swelling, one or both, of the joints, usually of the lower extremities. These have been regarded by some as rheumatic in character, and hence the terms purpura rheumatica and peliosis rheumatica. The most recent authorities appear to agree that the articular affections in these cases are *consequent upon* the hæmorrhagic disease, a view which is not susceptible at present of absolute proof, though the probabilities in the matter undoubtedly confirm it.

The disease occurs at all ages in both sexes, and under every variety of constitutional condition, preferably, however, at or shortly after puberty and in females. Apart from the articular pain, to which reference has already been made, the hæmorrhages occur without pain or warning of any kind. The extravasations may take place over the entire surface of the body, and the viscera and cavities also may be the seat of hæmorrhage, though usually it is confined to the mucous cavities. The spots of discoloration vary in size and shape. Vesicles, too, sometimes are observed, but these are quite exceptional. The color of the spots is indicated sufficiently well by the common name of the disease.

Many cases end in recovery, the patient frequently manifesting no serious disturbance of the general health, while others present alarming symptoms of great pallor, extreme prostration, dropsical swellings, and fainting fits, and many terminate fatally after short or longer intervals.

Post-mortem examinations reveal practically nothing as to the cause of the malady, but merely its effects, nor have repeated examinations of the blood during life afforded any satisfactory conclusions. Recent authorities, however, appear to be in accord with the view that the affection has its origin in some primary disease, either of the blood or the vascular walls, or of both together.

This short summary of the chief points of interest which characterize the disease under consideration, I think, will enable us at once to eliminate, in a differential diagnosis, most of the affections which might be suggested by some as the essential disease. We may, therefore, exclude, without any discussion, diseases of the thoracic and abdominal viscera, leukæmia, progressive pernicious anæmia, typhus and typhoid fever, cere-

bro-spinal meningitis; embolism, for the history of the case furnishes nothing to indicate the existence of any of these affections.

In some instances it is not an easy matter to distinguish between hæmophilia and morbus maculosus. Our present case, however, does not present the hereditary element which is an essential of the former disease, nor did the patient furnish any evidence of an hereditary hæmorrhagic tendency. Nothing in his record would suggest this unless, possibly, the fact that his gums were apt to bleed slightly when he made use of a stiff tooth-brush, a not uncommon occurrence with many of us.

Under some exceptional circumstances it is difficult to distinguish between *scurvy* and morbus maculosus. Our patient, however, was in good circumstances, hygienic and otherwise; had been accustomed to the ordinary variety of diet; had been subject to no privation. No similar case occurred in the family or neighborhood. The disease evidently was "spontaneous" in its origin. There were no distinct prodromic symptoms, except a feeling of languor and inability to attend to his ordinary duties for a few days. Previous to that time he was in perfect health. The gums, from which the blood was seen to exude, presented a perfectly healthy appearance. As far as we were able to judge, none of the local symptoms afforded any evidence of an inflammatory process which is very common in scurvy.

The so-called rheumatic symptoms presented by our patient, that is, the swollen and painful joints, accompanied by a slight rise of the pulse and temperature, claim, perhaps, further consideration in making the diagnosis. It has already been stated that recent authorities are inclined to agree that mild, apparently rheumatic, symptoms occurring in association with phenomena which, without them, would indicate the existence of the purpuric disease, may, with good reason, be regarded as *symptomatic* merely. Reference to the recorded experience with acute rheumatism cannot but confirm this view. It has been shown that with the association of the symptoms the rheumatic elements never obtain any decided prominence; that they are not attended by notable perspiration and never are followed by the grave complications peculiar to the articular disease. In some undoubted cases of rheumatism extravasations of blood do occur, but it is known that this accident is due to embolic processes which have their origin in *endocarditis ulcerosa*, a condition of things which the clinical history of our patient could not suggest.

Admitting, however, with many, that it is impossible, with our present knowledge, to decide absolutely whether one or the other class of symptoms is symptomatic, it will be evident from the whole character of this paper that I rely upon the theory of probabilities in these cases in making my diagnosis, and so regard the hæmorrhagic affection as the essential disease.

It occurred to me, and has been suggested by others, that the condition presented by my patient may have been due to poisoning by salicylic acid, he having taken in all about three hundred grains. This supposition, however, may be dismissed with a few words. I know of no case on record where this agent is supposed to have given rise to symptoms resembling those presented by my patient. Excessive and long-continued doses, and smaller amounts in certain individuals, of this acid have been known to occasion a condition of extreme prostration, in some cases sudden and rapid collapse, together with cerebral symptoms varying from

those resembling cinchonism to acute delirium. Sometimes, too, it produces a miliary eruption which often becomes pustular in character. Urticaria, also, occasionally occurs as a complication. In no case, however, has the eruption become hæmorrhagic in character. Hæmorrhagic *erosions* of the stomach and intestines have been observed, but in these cases this accident has been considered to be due, probably, to adulteration with carbolic acid.

GROWTH OF BONE IN KNEE-JOINT DISEASE FROM MEASUREMENTS OF NINETY-TWO CASES.

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THE question which has been agitated during the past few years regarding the influence of disease of the joints upon the bones entering into their formation has resulted in the publication of very few cases illustrative of these changes. Jutland¹ has reported cases of elongation of the femur associated with disease of the hip-joint. Wagstaffe² publishes twenty-five cases, of which the femur was found to be lengthened in fourteen, and shortened in one case only. Helferich³ has given measurements of one hundred and forty-one subjects of caries of the long bones, but without special reference to disease of the knee-joint. In eighteen of these there existed an elongation of the femur in three cases only, while in fifteen some shortening was noticed.

The accompanying statistics were obtained from measurements of consecutive cases in children which came under my observation at the daily clinic during the past two years. The greatest degree of accuracy attainable was secured, and no reference is made to those cases in which there were well-defined evidences of disease, but in which no accurate notes were taken.

Of the cases under consideration sixty-eight were of articular osteitis, and gave the following results:—

In 2 cases	1 inch lengthening.
In 10 cases	$\frac{3}{4}$ inch lengthening.
In 19 cases	$\frac{1}{4}$ inch lengthening.
In 18 cases	$\frac{3}{8}$ inch lengthening.
In 8 cases	$\frac{1}{4}$ inch lengthening.

In six there was an average shortening of three eighths of an inch, while in five no change in length was appreciable. Owing to the difficulty of estimating so slight a variation as a quarter of an inch I would not claim for this measurement any great degree of reliability. In addition to this evidence of bony proliferation there was noticed in most of these cases a marked lateral expansion at the condylar extremity of the femur.

That variety of articular disease in which the morbid process was situated primarily within the synovial membrane, and may be included under the general term of synovitis, numbered twenty-four. In these there appeared to be little alteration as regards osseous development; in fact, there seemed to exist in some cases an arrest in growth rather than an intrinsic enlargement, as indicated by this table:—

In 5 cases	$\frac{1}{8}$ inch elongation.
In 3 cases	$\frac{3}{8}$ inch shortening.
In 2 cases	$\frac{1}{4}$ inch shortening.
In 14 cases	unaltered.

¹ British Medical Journal, February 5, 1881.

² St. Thomas's Hospital Reports, 1880.

³ Deutsch. Zeitsch. f. Chir., Bd. x., 1879.