IDIOPATHIC PURPURA WITH UNUSUAL FEATURES

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Many authorities refuse to regard purpura as a disease entity and consider it merely as a manifestation of some other disease. Its extreme rarity is evidenced by the fact that at the Massachusetts General Hospital there were only sixty-four cases in 155,884 medical and surgical admissions; at Johns Hopkins Hospital there were forty-one cases in 18,594 medical cases, and at the Hamburg General Hospital there were seventy-three cases in 100,000 admissions; B. Ramwell 1 saw sixteen cases among 5,256 ward patients. In his table of 258 cases of all kinds of purpura fifty-four were primary and showed arthritis.

In looking over the literature of purpura, it is difficult to find any mention of familial tendencies in this disease. In fact, familial tendencies have been used as a point against purpura in a differential diagnosis from hemophilia, so rarely are they found. There are, however, a few instances reported to which we must add that of the two brothers in this paper. Cousin 2 describes a family in which the disease occurred in three different branches. A case is described by Dohrn in which purpura was transmitted from the mother to her new-born child. Occurrence of the disease in three sisters is reported by Forster. Wagner reported a case of chronic purpura in which paternal uncle of the patient died of acute purpura hemorrhagica. Bauer also describes familial tendencies to a purpuric diathesis occurring in several members at a certain age. The occurrence of these familial cases of purpura rather speaks for the disease as a definite entity. It would seem that there is in certain families an inherent weakness of the germ plasm which manifests itself either in a deficiency of blood platelet formation or in a primary vascular degeneration, or in both these factors (Duke, 3 Wright and Kennicutt, 4 Deny and Ledingham, 5 and Hess, 6).

REPORT OF CASES

Case 1.—Nelson C.; aged 11; schoolboy; born in Canada.

Family History.—The family history is entirely negative, there being no history of any similar ailment on either parental side, with the exception of the patient's brother whose case is reported later. The father and mother enjoy excellent health. As far back as they can trace, there has never been a bleeder in the family or anyone with hemorrhagic tendencies.

Previous History.—This boy was born normally at full term. He developed normally up to the sixth year after having passed through attacks of measles and whooping cough at the age of 1 year. He had mumps and scarlet fever, complicated by otitis media, at the age of 6. During the whooping cough he would often have epistaxis with paroxysms of coughing but this always stopped promptly and there was never noticed any delay in the clotting of his blood.

The child has been brought up in good surroundings and has had at all times a healthful mixed diet with abundance of fresh vegetables and fruits.

Present Illness.—The present illness began about four years ago with a sudden attack of pain in his elbows. They became swollen, stiff and extremely tender but not reddened. A similar condition later attacked his knees and from then up to the present he has never been entirely free from trouble in some joint, although the knee joints have been the main sufferers. There has been little, if any, fever with the attacks. The swellings appear quite suddenly and appear to distend the tissues about the joints until they are tense and so tender that the merest touch causes excruciating pain. At first they subsided rapidly and seemed to leave the joint no worse for the attack, but of late the knees have become stiffened and somewhat flexed. Simultaneously, the mother noticed that the child "bruised easily" and showed occasional small hemorrhages into the skin. Also on several occasions there would be a sudden swelling, discoloration, hardness and tenderness of the calf muscles. The boy has never had any involvement of the toes, hips, shoulders, sternoclavicular or temporomaxillary joints. There is no history of hemorrhage from the gums, nose, ears, kidneys, bowels or stomach. His blood clots normally after injury.

Fig. 1.—Elbow joint (Case 1) with area of rarefaction in olecranon process; enlargement of epiphyses of humerus and radius, and roughening of medial articular surface.
and he seems normal in every other way, except that he has always been pale. With the exception of the pains in the joints he has no complaints whatever.

Physical Examination.—This reveals a normal sized boy, pale and with a puffiness of the eyelids. The conjunctiva and mucous membrane of the mouth are free of petechiae. The fundi are both clear. There are shotty posterior cervical and inguinal glands. The lungs are clear. The heart is negative, except for a loud systolic murmur over the pulmonic area. The liver, spleen and kidneys are not palpable. The abdomen is negative. The interest centers in the joint condition, especially that of the knee joints. There is great muscular atrophy of both legs emphasizing the enlargement of the knees. The legs are held flexed. The knee joints are both distended, and there is a thickening of the peri-articular tissues. Riding of the patella is demonstrable. The skin over the knees and on the right side, extending down the back of the leg, is tense and of a bluish-yellow hue due to old blood extravasation. The right knee is extremely tender. There is limitation of motion. No crepitation can be elicited. The right gastrocnemius muscle is tense, tender and hard resembling the so-called scorbutic scleroderma. The skin shows a few small purpuric spots with several larger subcutaneous hemorrhages in the various stages of absorption and discoloration. These are mainly in the forearm, elbow and thigh.

Laboratory Examination.—Blood examination shows: hemoglobin, 45 per cent.; red blood cells, 4,000,000; leukocytes, 5,600. Differential count: normal.

Fig. 2.—Knee joints in Case 1; anteroposterior and lateral views.

The red blood cells show anisocytosis, poikilocytosis, stippling and polychromatophilia. The blood platelet count is 98,039 per c.mm. The coagulation time is normal. The bleeding time is prolonged to ten minutes instead of being from one to three minutes.

The urine is completely negative. The von Pirquet tuberculin test and the Wassermann test are both negative.

Case 2.—Clarence C., aged 15; schoolboy; born in this country. A brother of the patient reported in Case 1. His complaint was “sore elbows.”

Previous History.—He was born normally at full term and developed normally. Had attacks of measles, whooping cough, scarlet fever and mumps. Had an attack of jaundice at the age of 5. He had no other acute illness of any kind, and his general health has been good. It was noticed that he “bled easily and freely” on the slightest provocation but no delay in the clotting of his blood was ever present. For instance, on changing altitude, he would suffer with epistaxis, although the changes were not great enough to affect the ordinary person.

Present Illness.—His present illness dates from the age of 5 when he had his first attack of joint trouble. This centered in both ankles, which suddenly became swollen and tender. This condition cleared up rapidly only to reappear after a short interval and then the knees were similarly affected. The
patient was unable to walk. He suffered with these attacks up to the present, although lately they have become less frequent and less severe. For the past three years the attacks have been confined to the elbows and he is able to walk without any difficulty whatever. On one occasion, a diagnosis of purulent arthritis was made and a knee joint was aspirated. Bloody serum was obtained. The boy bruised easily and often had large hemorrhages under the skin. He never had an attack of extensive purpura nor was there ever spontaneous oozing from the gums. Six weeks ago he had an attack of severe colicky pain in the right side in the costo-vertebral region radiating downward and forward over the bladder. To use his words, he "felt something pop inside" and

then he "felt a hot flush through his stomach." Shortly after he noticed that his urine was bloody and stringy. As soon as he began to pass the blood the pains subsided somewhat. This attack lasted for three days. The urine became clear and the pain disappeared. A similar attack appeared four days ago in the left side. The boy's mother noticed no blood in the urine in this attack which was less severe than the previous one. A microscopic examination, how-

Fig. 3.—Knee joint of Case 1, lateral view, showing definite area of osteoporosis in epiphysis of femur with undue enlargement; thickening of soft structures of the knee joint, commonly seen in repeated blood effusions. Condylar surfaces are rougher than is the case in normal persons.
ever, showed the presence of red blood cells. In neither attack was there any purpura, nor did the boy show any melena. There was no nausea or vomiting nor was there any exacerbation of his joint symptoms. There have been no other symptoms of any importance.

**Physical Examination.**—On physical examination, we find a bright, normal sized, well-nourished boy. He does not appear to be anemic. The head is entirely negative. The eyes show no petechiae. The fundi are clear of any hemorrhages. Mucous membranes are normal. The gums are healthy and the teeth are excellent. There is slight general glandular enlargement. The tonsils are negative. The heart and lungs are clear. The abdomen shows no abnormality. There is marked left costovertebral tenderness present. The liver, kidneys, gallbladder and spleen are not palpable. The skin shows a few large brown and blue bruises, one over the right eye and one near the left elbow. There are a few smaller purpuric spots in the skin of the left forearm. There are no hemorrhages in the muscles.

As in the first case, the interest centers in the joint condition. The left elbow is the seat of an acute process. The joint is very much enlarged and discolored but not especially tender. The bony landmarks are almost obliterated by the effusion into and the distention of the joint capsule. The olecranon process shows as a depression between two large fluctuating bulging sacs. There is limitation of motion in this joint. Both knees are enlarged due to a peri-articular swelling and thickening but they are not tender, there is no limitation of motion nor is there any sign of an effusion at present. There are scars over both knee joints at the site of the previous aspiration. All the other joints are normal.
Blood Examination.—Hemoglobin, 85 per cent.; red blood cells, 4,900,000; white blood cells 6,600. Differential count, normal. The red blood cells were normal. The blood platelet count was 204,000 per c.mm. The coagulation time was normal. The bleeding time was ten minutes. The urine showed a faint trace of albumin with many red blood cells. The remains of the last attack of colic.

The roentgenograms of the joint (Figs. 1, 2, 3, 4 and 5) show, in general, an enlargement of the epiphyses of the bones with well defined areas of rarefaction, small roughening of the articular surfaces, as in osteo-arthritis; and in the knee joint, a definite thickening in the soft structures of the joint such as one sees in bloody joint effusions.

Clinical Course.—The two patients were seen after an interval of seventeen months' army service. The younger brother had had at least one attack of joint trouble a month during this period. He had also had one severe hematemesis in which he lost almost one quart of blood. This occurred six months...
after therapeutic intramuscular injections of whole blood. On my return, I
found him with a hemorrhage under the tongue, one under the angle of the
left jaw, many purpuric spots over the sternum, shins and ankles, and a fresh
hemarthrosis of the wrist. His general physical condition was the same, except
that the urine showed albumin, casts and red blood cells.

The older boy seemed much improved. Fourteen months after I saw him
he suffered a fracture of the right olecranon. He did not know just how or
when it occurred. The bone united readily but two months later he developed
a fresh hemorrhage in the same joint. He had also had epistaxis but no bleed¬
ing into the skin, bowel, stomach or kidney.

A second feature of these two cases is the occurrence of repeated
hemarthrosis in each instance. Some even go so far as to doubt its
occurrence in purpura, stating that if the arthritic manifestations are
at all marked the case should be classed as a simple purpura with
arthritis. According to McCrae 7 no case has been found in recent
literature. Wagner 8 searched the literature up to 1886 without finding
a single case of purpura in which a large hemorrhage had occurred

Fig. 6.—Elbow joint of Case 2.

into a serous sac. Hoffman found no mention of a single definite
instance of hemorrhage into a joint in morbus maculosus. There
could be no doubt of their occurrence in the above cases. The sudden
painful distention of the joints, the external discoloration and sub¬
cutaneous ecchymosis of the surrounding tissues and the result of
ill-advised aspiration all go to prove the presence of hemorrhage in
the joints. The rapidity of recovery of normal appearance and
function were also characteristic in the early years of the disease.

A third rare feature is the association of hemarthrosis with the
visceral symptoms described under the name of Henoch's purpura or
purpura abdominalis. In typical cases of Henoch's purpura joint pains
are associated with cutaneous and intestinal hemorrhages and acute
nephritis, often of the hemorrhagic type. The arthritis, however, does
not go on to the hemorrhagic stage. Each of these two patients, at

some time in the course of his disease, has shown one or both of these two visceral conditions in addition to joint hemorrhages. Nor is hemorrhage into a joint mentioned as one of the symptoms of peliosis rheumatica by Schönlein in his original description of this condition. This description, although vague, has been taken by most clinicians to include only those cases in which there are fever, malaise, purpura and erythema and joint pains. Barker, however, in his classification of the purpuras, states that often there is hemarthrosis in these cases of Schönlein's disease. The absence of fever, malaise, erythema and pains in joints other than those distended by blood, definitely excludes these patients from the peliosis rheumatica group.

Another feature of note is the fracture of the olecranon sustained by the older brother. From the description of the patient this was in all likelihood a spontaneous or pathologic fracture in the neighborhood of a joint that had been the seat of a hemorrhage. If one bears in mind the pathology of this disease, especially with regard to the

rarefaction it effects in the bone substance, it is quite conceivable how this might happen. It might be well, therefore, to consider purpura as a possible etiology for pathologic fracture.

From a perusal of the literature and a consideration of these two cases it would seem that no strict line can be drawn between the various types of purpura described in our modern classification. Morawitz considers all these diseases of the so-called idiopathic purpura group as being different manifestations of one and the same process. Hoffmann, Litten and other German writers have the same view of the primary purpuras, classing them all as "morbus maculosus Werlhofii" of varying degrees of severity. In this group are: (1) purpura simplex, with hemorrhages in the skin and subcutaneous tissues; (2) purpura hemorrhagica, with hemorrhages of the skin and mucous membranes; (3) peliosis rheumatica with cutaneous hemorrhage erythema and joint disease; (4) purpura abdominalis, with cutaneous and intestinal hemorrhages and joint pains, and (5) purpura fulminans, with cutaneous hemorrhages which run a rapid and fatal course.

The cases here reported partook of features that might allow them to be put in any of the above type, except the last.

The pathology of this condition, when the hemorrhage has been repeated often, is quite interesting. Our knowledge of it has been gained by roentgen-ray study of the living subjects, by operations and by a few necropsies in cases that have succumbed to intercurrent conditions. The hemorrhages occur most often in the knee joint, as trauma is most active here, although other joints may be affected simultaneously.

Following a slight blow or often spontaneously, there develop signs of acute joint affection with joint effusion. The condition may stop here, the effusion be absorbed and the joint heal without change. This, according to the description of König,10 is the primary stage of the disease. If the effusion persists—secondary stage—it goes on to an inflammation with a fibrinous effusion as in the ordinary tuberculous "white swelling." This condition it simulates almost perfectly, but the roentgen-ray will show that the bone is not involved and that the process is confined to the soft parts of the joint (Love,11 Carless12). In this stage, synovial villae are present as well as defects in the joint cartilages, the changes resembling closely those seen in arthritis deformans or osteo-arthritis. The roentgen ray shows changes in the epiphyseal lines which are zigzag and have a worm-eaten appearance. The condylar outlines are not sharp. The capsule shadow is darkened. There

is bone atrophy with rarefaction of the condyles. Mankiewicz \textsuperscript{13} questions whether this is a true osteoporosis or a retrogressive metamorphosis in the architecture of the bony trabeculae. If hemorrhage occurs in a joint often enough, König describes a third stage in which there are contractures, bone and joint changes, thickening of all the periarticular structures and even ankylosis. Ponfick also has described hemorrhages into the bone-marrow. Bowlby \textsuperscript{14} thinks these cases resemble osteo-arthritis far more closely than is generally acknowledged. He cannot explain the nodular outgrowths of cartilage, new bone formation, fibrillation of the cartilage matrix and the formation of adhesions all as a result of blood extravasation. The attacks, he states, are different than those exhibited by a healthy patient who is suffering from a traumatic hemarthrosis. He thinks that many bleeders have a special tendency to attacks of acute arthritic inflammation which very closely resemble those of the more severe forms of osteo-arthritis. Barker \textsuperscript{15} states that the condition also resembles luetic arthritis.

**SUMMARY**

Two cases of iodiopathic purpura are described with the following unusual findings:

1. They showed a familial tendency.
2. Repeated joint hemorrhages occurred in both patients.
3. There was an association of joint hemorrhages with the symptoms of Henoch’s purpura.
4. Spontaneous fracture occurred in one case.

\textsuperscript{13} Mankiewicz: Berl. klin. Wchnschr. \textbf{76}:2174, 1913.
\textsuperscript{14} Bowlby: St. Bartholomew Hosp. Rep., London \textbf{26}:77, 82, 1890.
\textsuperscript{15} Barker: Monographic Medicine \textbf{4}: p. 95.