

# BLOOD

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### THE SCHOENLEIN-HENOCH SYNDROME OF VASCULAR PURPURA\*

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#### DEFINITION

**T**HE Schoenlein-Henoch syndrome is a condition in which nontraumatic hemorrhage, with or without edema, occurs in the skin, or subcutaneous tissue, or joints, or viscera, or in any combination of them, and in which the blood platelets are normally abundant.

The name Schoenlein-Henoch purpura is well established, yet the eponym is ill chosen (as so many eponyms seem to be). Willan<sup>10</sup> described one type of the syndrome in a London publication in 1808, while a description of Schoenlein's disease was recorded not by Schoenlein himself but by one of his pupils in 1837.<sup>11</sup> According to this description, the patients were often rheumatic sufferers, and they presented purpura, pains, and swellings, particularly in the knee and knuckle, but occasionally in the hand and shoulder joint. Henoch's work appeared in 1868<sup>7</sup> and he referred to 7 cases in a paper in 1874.<sup>8</sup> Apert<sup>1</sup> (1897) gave an excellent clinical description under the name of exanthematic purpura. Osler,<sup>9</sup> in 1914, in his paper, "The Visceral Lesions of Purpura and Allied Conditions," did much to clarify ideas on the condition, but his view that the anaphylactic key would unlock the mystery of these cases found too ready an acceptance.

Numerous case reports have appeared, but most observers have described but few cases each and only a hazy idea of the natural history has emerged. The description which follows is based on 44 personally observed cases seen in eight years, against a background of 1200 cases of purpura (mostly benign). An account of the first 500 of these cases has been given by Davis<sup>3</sup> (1943).

#### SEX, AGE, DURATION

The condition was about three times as common in women as in men, but, in children, boys and girls were about equally affected. Patients aged 4 to 71 were affected, but there were peaks of incidence of first attacks from 4 to 10 years old, and from 16 to 40. Half of the patients had had more than one attack, and half were seen in their first attack. It is probable that about one third had had but one attack. A few had had numerous attacks. Each attack usually lasted for a few weeks. Attacks had recurred over periods varying from a few months up to 50 years,

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This One



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but in most patients they ceased within five years from the onset. Of the 44 cases, 5 patients were each members of different families with hereditary familial purpura simplex. No familial instances were encountered.

#### ETIOLOGICAL FACTORS

In 10 patients attacks were preceded by acute tonsillitis. In one patient, each of her three attacks was heralded by an acute tonsillitis. One patient suffered from



FIG. 1. Girl, aged 8. (a) Edema of eyelids and face. (b) Petechiae of buttocks. Melena, hematuria; joint swellings.

pulmonary tuberculosis and had two attacks. In another patient true acute cholecystitis preceded the attack. There was a history of acute rheumatic fever earlier in life in 4 patients, but in at least 2 of these patients the so-called acute rheumatism was almost certainly an earlier attack of Willan-Schoenlein-Henoch disease. Only 6 of the patients were affected with fibrositis. In 8 patients an antistreptolysin titer was done at the height of the disease; in 6 the titer was normal, and in 2 raised to 800 and 1250 units. (Several of the patients with acute tonsillitis had normal titers.)

## TEMPERATURE

Only 11 patients were febrile during attacks. Three were febrile for one month, another had low grade fever for one year, a fifth had bouts of low grade fever lasting about a week every few months for over five years, and a sixth had persistent fever of up to 100-101° for months at a time for the eight years she was under my observation. Blood cultures were negative in all.



FIG. 2. Girl, aged 7. Petechiae back of neck and shoulder; blood in cerebrospinal fluid; melena; joint swellings.

## PURPURA, RASHES, EDEMA

Of the 44 cases, 38 showed spontaneous ecchymoses or petechiae or both. Most showed skin purpura only during attacks but a few had ecchymoses between attacks. The ecchymoses, which occurred mostly on the limbs, varied from  $\frac{1}{2}$ -4 inches in diameter (fig. 4) and were painless. Petechiae were seen, particularly over the skin of the buttocks and scapulae (figs. 1 and 2), but the number of petechiae varied enormously, and the widespread rash with dense petechiae (fig. 5) was rare. Petechiae were also seen in the mucous membrane inside the mouth and occasionally on capillary microscopy of the nail bed.<sup>4</sup> In two patients the purpura

was in the form of a purpura nodosum, the erythema of erythema nodosum being replaced by purpura (fig. 3). Melena was seen in 5 patients. Epistaxis, hematemesis, and menorrhagia each occurred in 4 patients, hematuria in 3, mouth bleeding in one; blood was found in the cerebrospinal fluid of one patient with meningeal irritation.<sup>5</sup>

Urticaria accompanied the attacks in 3 patients and 4 others had a papular or papulovesicular rash. Localized edema occurred in the following order of fre-



FIG. 3. Woman, aged 22. Ecchymoses of legs; "purpura nodosum"; hematuria; febrile one year; joint swellings.

quency: backs of hands, around the eyes, face (fig. 1a), neck, lips, legs, arms, and penis.

#### JOINTS, VISCERA

Joints particularly involved were those of the fingers, wrists, knees, elbows, and ankles. Any one or any combination of these joints could be affected. Joints were painful and nearly always swollen. In some patients the edema was peri-articular and para-articular rather than in the joint itself. Between attacks all affected joints recovered full function and did not ankylose, but in a few patients pain persisted. Radiological changes did not develop.

Gastrointestinal pain or hemorrhage was pronounced in 12 patients, while 3

patients had hematuria, a fourth casts and albuminuria without hematuria, and one patient gallbladder disease. Menorrhagia was present in 4 patients and uterine pains in a fifth. The spleen was not palpable.

#### GENERAL

The time relationship of the features of an attack was very variable. Petechiae, edema, joint pains, and visceral features often appeared within hours of one another, but skin petechiae at times preceded or followed other signs by an interval



FIG. 4. Woman, aged 40. Ecchymoses; febrile eight years; joint swellings.

of several days. Thus a puzzling visceral pain or joint swelling could be diagnosed only when a purpuric rash developed days later, or the nature of a purpuric rash might remain obscure until swollen fingers, orbital edema, and melena occur. In some cases, skin manifestations were absent entirely. By way of contrast, angioneurotic edema might be the sole feature of an attack. A classic illustrative case from this series has been described by Green.<sup>5</sup>

#### BLOOD FINDINGS

Blood counts, platelet counts, bleeding and coagulation times, clot retraction, and prothrombin time were normal but occasionally a polymorphonuclear leuco-

cytosis was seen. Blood sedimentation rate was usually normal, but occasionally high, likewise the antistreptolysin titer. When these were high there was also leucocytosis. The capillary resistance test was positive in only some 25 per cent of cases.

#### PROGNOSIS

The outcome for a particular attack and the ultimate outcome were good. Recovery was invariable, good health was enjoyed between attacks, and in most



FIG. 5. Man, aged 30. Massive petechiae legs, hematuria, joint swellings.

patients attacks rarely recurred after 5 years. The 4 patients with renal involvement had no residual lesions and made full recoveries despite the gloomy prognosis so often given to Schoenlein-Henoch purpura with renal complications. The child with the subarachnoid hemorrhage recovered completely.<sup>5</sup> But some patients required treatment for hematuria or hematemesis or melena, which they naturally found alarming. Persistent fever in several patients was worthy of note, particularly in the case presenting pyrexia lasting for eight years. This experience demonstrates that Schoenlein-Henoch purpura must be added to the causes of persistent low grade fever.

## DIFFERENTIAL DIAGNOSIS

It is clear that abundant opportunities for confusion with other diseases exist. Among the labels attached to cases I encountered were acute rheumatic fever, septicemia, subacute infective endocarditis, rheumatoid arthritis, infective arthritis, nephritis, bleeding peptic ulcer, and trichinosis. The differential diagnosis from palindromic rheumatism<sup>6</sup> must also be considered. The cases of angioneural arthrosis described by Solis-Cohen<sup>13</sup> (1914) may be identical with Schoenlein-Henoch purpura.

## TREATMENT

Treatment was essentially symptomatic. Vitamin C was of no value; the preparations of vitamin P used were not beneficial; and vitamin K was not indicated. The patient with persistent attacks of fever for one year recovered after tonsillectomy. The patient with the eight years' fever was not helped by protein shock, auto-hemotherapy, blood transfusions, or sulphonamides. After the exhibition of penicillin an attack was cut short and she enjoyed three months of exceptional well-being, fever-free. But a second course of penicillin given for the relapse was ineffective. Spontaneous recovery had occurred after protracted attacks in the pre-penicillin era.

## DISCUSSION

Most authors regard purpura simplex and Schoenlein-Henoch purpura as different facets of the same condition. By purpura simplex, I understand a condition in which ecchymoses with or without petechiae occur from time to time in the skin without any known trauma and without ascertainable cause. Now there is no doubt that one group of Schoenlein-Henoch cases is closely linked with purpura simplex—witness the occurrence of a case of Schoenlein-Henoch purpura in five different families of hereditary purpura simplex, while some cases of purpura simplex become almost indistinguishable from Schoenlein-Henoch purpura. The blood findings in both conditions are identical. There are distinct differences, however. Purpura simplex is overwhelmingly more frequent in women, is very often familial and is not common in children, but Schoenlein-Henoch purpura is not uncommon in males, is not familial and is quite common in children. One third of the patients with Schoenlein-Henoch purpura have only one attack, but single attacks of purpura simplex are much rarer. The fibrositic diathesis is much commoner in purpura simplex. My conclusion is that Schoenlein-Henoch purpura is a syndrome and not a disease and is a nonspecific reaction to different factors. The causes of purpura simplex and Schoenlein-Henoch purpura overlap but are not identical. One great cause of both conditions is recent infection by hemolytic streptococci. (It is possible that in some instances Schoenlein-Henoch purpura represents aborted or modified rheumatic fever, sparing the heart. Osler<sup>9</sup> stated that rheumatic poison is believed to be responsible for a large group of these cases.) In most of my cases the cause was not found. In none of my cases of Schoenlein-Henoch purpura were food or drugs or known allergens found to be the cause,

but I would agree that allergic factors cause this condition occasionally. It is to be noted that no support for the anaphylactoid etiology was given by Bartley and Bell<sup>2</sup> (1936), Poncher<sup>10</sup> (1935), and Siedlmayer<sup>12</sup> (1939). Osler<sup>9</sup> was optimistic in his belief in the anaphylactic key, but he was right to insist that urticaria, angioneurotic edema, and vasomotor instability were all aspects of the same condition.

#### SUMMARY

The Schoenlein-Henoch syndrome is described on the basis of 44 personally observed cases. It is defined as a condition in which nontraumatic hemorrhage with or without edema may occur in the skin, or subcutaneous tissue, or joints, or viscera, or in any combination of them, and in which the blood platelets are normally abundant. The causes of the syndrome are varied, but streptococcal infection is important, and "anaphylactoid" causes rare. The prognosis is good, and the 4 cases with renal involvement also did well. Schoenlein-Henoch purpura and purpura simplex overlap, but in contrast with the latter, Schoenlein-Henoch purpura is not familial, and is common in males and children.

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