EDITORIAL

Acute Vascular Purpura
An Immuno-Vascular Disorder

EVANS,1 in writing of idiopathic thrombocytopenic purpura (ITP), cited several interesting relationships between that disease and acquired hemolytic anemia and suggested that ITP might well be due to the presence of an immunologic abnormality. For the auto-immune type of acquired hemolytic anemia, he suggested the term “immuno-hemolytic anemia” and for ITP “immuno-thrombocytopenia.” Since the appearance of Evans’ provocative article in January 1951 there has been abundant confirmation of his suggestions, and the analogies between auto-immune hemolytic anemia and ITP have become even more impelling.

An immunologic abnormality seems particularly likely in the acute form of ITP.2,3 Here, a patient previously well, suddenly develops a generalized and very severe purpura, often with mucous membrane bleeding. The platelet count is extremely low and the bone marrow megakaryocytes show many degenerative changes. The history often reveals either that an infectious disease (“grippe” sore-throat, chicken pox, etc.) has antedated the purpura by about seven to fourteen days or less, or that a drug, such as quinidine, previously given intermittently, has recently been taken. One cannot escape the conclusion that an infectious or drug antigenic factor has set up an auto-immune or allergic reaction which, after a variable “build-up” period, explodes into purpura. The acute cases seem to present not only thrombocytopenia, but small blood vessel injury as well; in fact, the latter may be responsible for much of the purpura, since bleeding and purpura usually subside prior to an increase in blood platelets.

In many respects, acute vascular purpura may be likened to acute thrombocytopenia purpura. Variously called nonthrombocytopenic purpura, Henoch’s purpura (with gastrointestinal manifestations) Schönlein’s purpura (with joint manifestations), Henoch-Schönlein purpura, and anaphylactoid purpura, its course may be analyzed as follows:

1. There is an infection, usually of the streptococcal variety, often accompanied by a severe sore throat. The infection subsides either spontaneously or with the aid of antibiotics.
2. There is a lag or build-up period of seven to fourteen days.
3. There is a sudden “explosion” during which, with infinite variations, the following take place:
   a. Severe, very deeply purple “purpura”, seen for the most part about the joints but also present to lesser extent over the trunk.
   b. Joint pains and swellings.
   c. Severe abdominal pain often with bleeding from the bowel.
   d. Hematuria.
4. A state of defervescence during which all of the various manifestations, except as a rule the renal one, show striking improvement. The renal lesion may become chronic. Some of the purpuric lesions of the extremities may become ulcerated.
According to this schema and other data, it seems likely that acute vascular or Henoch-Schönlein purpura is an allergic or immuno-allergic disturbance which is characterized not simply by purpura but by a generalized small blood vessel disturbance or angiitis affecting the skin, joints, intestinal tract, renal glomeruli, and probably other sites as well. The term “anaphylactoid”, as used by such observers as J. H. Pratt many years ago, appears to have considerable merit for this condition; or to carry out Evans’ usage, one might describe this syndrome as an immuno-vascular disease.

That hemagglutinating and hemolytic antibodies exist has become abundantly clear. In the last few years, investigation in the field of platelet antibodies has made rapid progress. Although acute vascular purpura is in some measure more clearly immunologic than the majority of cases of thrombocytopenic purpura, actual studies of blood vessel antibodies have been relatively few. They are summarized in a rather prophetic article published in this journal in 1950 by Clark and Jacobs. These authors first pointed to a number of important and almost completely neglected Japanese investigations which they obtained in English translation through the help of the U.S. Army authorities. They cited Katsura who, among others, produced vascular endothelial antisera by injecting the endothelial lining of guinea pig aorta into rabbits. When the antisera were then injected in guinea pigs, a generalized hemorrhagic purpura resulted, involving particularly the skin, lungs, gastrointestinal tract, intestines, and diaphragm. The platelet count did not become depressed. Clark and Jacobs continued this type of investigation hoping to test out the efficacy of certain anti-hemorrhagic materials if the method was successful. They used suspensions of dog aorta and produced rabbit antisera. When these were injected in dogs a generalized hemorrhagic purpura resulted. The blood vessel lesions in this experimentally produced immunovascular purpura resembled closely those of Henoch-Schönlein purpura.

Thus, from these various sources and including such clinical observations as the favorable results of therapy with ACTH and Cortisone it becomes increasingly clear that acute vascular purpura is probably an immunologic disturbance affecting small blood vessels throughout the entire body. In this respect, the disorder may be more or less closely related to another generalized vascular disturbance, namely, periarteritis or polyarteritis nodosa in which larger blood vessels are involved. There is increasing evidence that the latter condition is also an immunologic or allergic state in which relatively large blood vessels throughout the body are affected thus resulting in numerous clinical manifestations.

Lecutier has recently suggested that periarteritis nodosa and acute vascular purpura “constitute the same condition, and that whether the arteries or capillaries are involved depends on variations in the conditions of pathogenesis operating in any particular case.”

To carry the analogy a bit further, one could also include as possible immuno-allergic disturbances such diseases as thrombotic thrombocytopenic purpura and disseminated lupus. Indeed lupus may show simultaneously a number of auto-immune immunologic processes: thrombocytopenic purpura, auto-immune hemolytic anemia, and even a coagulation disorder based on an im-
When one considers that various clinical syndromes may present themselves in an incomplete fashion (*forma frustes*) it becomes possible that some cases of acute nephritis and of acute arthritis may represent simply one feature of a more fundamental disturbance. This, like an iceberg, may be submerged, leaving only a small portion clinically visible.

As a system of disease, immunohematology, which we have often cited as beginning in 1910 with Chauffard, who first proposed the term, may in the long run assume almost as much importance as the generalized adaptation syndrome of Selye. In fact, there are grounds for thinking that these two systems are even now openly competing ideologies for explaining apparently diverse conditions.

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**REFERENCES**

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