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Sarcoidosis and Idiopathic Thrombocytopenic Purpura

First Report of Coexistence

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A patient is described in whom sarcoidosis of the mediastinal lymph nodes was found during the course of acute idiopathic thrombocytopenic purpura. The association of these two diseases should be differentiated from the more commonly seen thrombocytopenia of hypersplenic origin, occurring during the disseminated stage of sarcoidosis, in which granulomatous involvement of an enlarged spleen is found. An elevated serum IgM immunoglobulin level was present. These observations possibly strengthen the concept that an immunological disorder plays a significant role in sarcoidosis.

The etiology of sarcoidosis, a granulomatous disease of world-wide distribution, remains shrouded in mystery. Its relationship to the collagen diseases and the significance of its immunologic abnormalities have eluded understanding. Recently we observed an unusual presentation of the disease, focusing attention on an autoimmune feature, which, we believe, deserves further consideration.

Case Report: C.M., a 29-year-old Negro male baker was admitted to the hospital on March 25, 1968, with complaints of severe epistaxis and bleeding from the oral mucous membranes. A week earlier he had visited his private physician because of a sore throat, coryza and slight fever. He was treated empirically with aspirin, cough syrup and penicillin G by injection. His symptoms promptly regressed. He felt well until the day of his admission, when painless, atraumatic bleeding from his mouth and nose brought him to the hospital. There was no cough, dyspnea, arthralgia, or abdominal cramps.

His past medical history contained nothing of significance, and his family history was unremarkable.

Positive findings on physical examination were limited to the skin and mucous membranes. Moderate bleeding from both anterior and posterior nares was noted, and blood was also oozing from the gum margins. The oral and conjunctival mucosa was covered with many purpuric lesions. Purpuric and ecchymotic lesions covered the skin of his extremities and trunk. The remainder of the examination revealed no other abnormalities. In particular, there was no peripheral lymphadenopathy, hepatomegaly, or splenomegaly. Cardiac and pulmonary findings were normal, as were those on fundoscopy and neurologic examination.

Laboratory investigations revealed the following: Hgb. 13 gm per 100 ml; WBC 9,600 per cu mm, with 67% neutrophils; 1% eosinophils; 24% lymphocytes; and 8% monocytes. The peripheral blood smear showed mildly hypochromic and polychromatophilic erythrocytes. A marked depletion of platelets was noted. The platelet count was 9,600 per cu mm. The bone marrow aspirate yielded a normocellular specimen with a myeloiderythroid ratio of 3.9:1.0 and orderly maturation. However, megakaryocytic elements were present in slightly increased numbers and showed the following differential analysis: megakaryoblasts, 4%; promegakaryocytes, 20%; and megakaryocytes, 76%. Fifty-two per cent of the megakaryocytes were interpreted as undergoing degeneration, and no platelet "budding" was observed.

Normal values were found for the following serum chemistries: creatinine, amylase, calcium, phosphorus, alkaline phosphatase, electrolytes, bilirubin, glutamic oxal-
Figure 1
Posterior-anterior x-ray view of chest in patient, C.M., showing right hilar and bilateral paratracheal lymphadenopathy. Fine nodularity present in parenchyma of both lungs cannot be appreciated in this photographic reproduction.
Sarcoidosis and ITP

Acetic transaminase and lactic dehydrogenase. The heterophile agglutination, Coombs' test, and L. E. cell preparation were negative.

The serum complement was 108.6 CH₅₀ units per ml (normal: 150-220). The serum protein electrophoresis was normal, except for the gamma globulin level of 1.64 gm per 100 ml. Quantitative immunoglobulins by gel diffusion on two occasions gave the following results: IgA, 104 and 119 (normal 30-135 mg %); IgM, 228 and 200 (30-120 mg %); and IgG, 877 and 1216 (600-1400 mg %).

The chest film revealed a lobulated mass in the right perihilar region with widening of the right and left paratracheal regions (Fig 1). There were fine nodular changes in both lung fields. Laminography of the lung roots and paratracheal areas further established the presence of mediastinal lymphadenopathy. The coccidioidin, histoplasmin, blastomycin and mumps skin tests were negative, but the intermediate strength tuberculin test was positive. Repeated examination of the sputum was negative for pathogenic bacteria or fungi.

Clinical Course: During the first week 2500 ml of whole blood were required to replace the blood loss from epistaxis. Treatment with prednisone was started soon after initial laboratory values were available. The platelet response in relation to prednisone dosage is depicted in Fig 2. Clinical improvement was gratifying, but serial chest films failed to show any change in the size of the lymph node masses. A right scalene lymph node biopsy, performed when the platelet count had reached a safe level, revealed noncaseating granulomas, consistent with sarcoidosis (Fig 3a, b). Subsequently, a biopsy of mediastinal lymph node material was obtained through a mediastinoscope, and this specimen displayed the same pathologic process (Fig 4a, b). Cultures of the nodes for tuberculosis were negative.

The acute thrombocytopenic purpura has remained in complete remission for the ensuing nine months despite the withdrawal of prednisone, and the patient is clinically well. However, the mediastinal lymph nodes remain unchanged in size.

Discussion

In 1938, Jersild¹ first described the case of a man with uveoparotid fever and lung changes, who also had thrombocytopenia (16,000 to 57,000 platelets per cu mm) and purpura. From 1939 to 1952 several similar reports have appeared.²-⁴ In all these cases splenomegaly was present, but splenectomy was not done. Nordland and coauthors⁵ were the first to describe the therapeutic effectiveness of splenectomy in controlling the thrombocytopenia associated with sarcoidosis. This and subsequent confirmatory papers⁶-¹¹ all described clinical splenomegaly and the presence of sarcoid granulomas in the spleens removed at surgery or examined at autopsy. Although some of the published data are incomplete, it is fair to assume that virtually all of the thrombocytopenia previously reported is the expression of a secondary hypersplenism due to splenic sarcoidosis.

In contradistinction, our case represents the coexistence of early sarcoidosis, limited to supradiaphragmatic lymph nodes, and autoimmune thrombocytopenic purpura (ITP). The fulminating acute onset of purpura, the severe degree of platelet decrease, the absence of splenomegaly, the megakaryocyte differential and morphology, and the response to corticosteroids all point to this conclusion.
Figure 3a

Scalene lymph node biopsy, × 140 showing multiple, noncaseating granulomata replacing normal nodal architecture.
Figure 3b
View of scalene lymph biopsy, $\times$ 480.
Figure 4a
Mediastinal lymph node biopsy, × 140 duplicating the microscopic picture seen in the scalene node. Special stains for micro-organisms were negative on both biopsy specimens, as were cultures for fungi and acid fast organisms.
Figure 4b
Mediastinal lymph node biopsy, × 480.
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The sarcoid process, no matter what the instigating factor(s) might be, expresses itself, in part at least, as an immunologic disorder. The well-known manifestations of iritis, arthralgia, erythema nodosum, skin anergy, and hyperglobulinemia are evidence for this. The occurrence of ITP with sarcoidosis, reported herein, is yet another clinical expression of an immunologic disturbance. Of course, the possibility exists that the prior respiratory infection or aspirin ingestion played a significant causative role in the appearance of the thrombocytopenia, but nonetheless, we feel that the association of ITP with a documented case of sarcoidosis may have greater significance in this instance. In this context, the elevated IgM immunoglobulin level in our patient is worthy of note. Further study of immunoglobulin behavior in both sarcoidosis and ITP may be rewarding.

Finally it seems significant that, although our patient’s thrombocytopenic purpura responded well to cortico-steroids, there was no discernible change in the mediastinal lymph node size. We interpret this as confirmatory to the contention that the thrombocytopenia was immunologic in nature and, as such, a correctable by-product of the sarcoid process.

REFERENCES