CASE REPORT

Twenty-Nail Dystrophy Associated with Hematologic Abnormalities

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This report describes a 15 1/2 year old white male with twenty-nail dystrophy who has had recurrent episodes of immune thrombocytopenic purpura, autoimmune hemolytic anemia, and mild depression of immunoglobulin levels. The concurrence of these events suggests that each shares a common pathophysiologic mechanism, possibly an autoimmune process. Key words: twenty-nail dystrophy, immune thrombocytopenic purpura, hemolytic anemia.

Twenty-nail dystrophy of childhood is a disorder in which the nails of the hands and feet exhibit excessive longitudinal ridging, brittleness, and thickening without associated abnormalities of other ectodermal tissues (1). The occurrence of twenty-nail dystrophy in a patient with a selective deficiency of IgA suggested that the disorder may be secondary to an immunologic abnormality (2).

A variety of immunodeficiencies have been demonstrated in patients with autoimmune hematologic disorders (3, 4). In some of these subjects, the hematologic abnormalities may precede the clinical evidence of an immunodeficient state.

CASE REPORT

The patient, a 15 6/12 year old white male, presented with severe bruising and gingival bleeding at 2 3/12 years of age. There was marked thrombocytopenia and a moderate leukopenia and anemia. A bone marrow aspirate was consistent with idiopathic thrombocytopenic purpura. After initiation of prednisone therapy the platelet count returned to normal. Subsequent episodes of thrombocytopenia, usually associated with viral illnesses, also responded to prednisone as well as intravenous immune globulin (see Fig. 1). Over the years the patient had several episodes of hemolysis associated with a positive direct Coombs' test. Except for occasional episodes of leukopenia, the leukocyte counts were normal or elevated, a pattern that paralleled the course of the platelet counts.

Serial tests for ANA (anti-single and double-stranded DNA and precipitins) and rheumatoid factor were negative. C-3 and C-4 levels were normal. At age 6 the IgG level was decreased with normal levels of IgA and IgM. When first measured at age 9, the platelet-associated IgG was positive and anti-platelet antibodies were detected. T-cell studies while on steroids revealed: OKT-3 of 56%, OKT-4 of 38%, and OKT-8 of 28%, all of which were normal; mitogen assays (phytohemagglutinin, concanavalin A and pokeweed) were normal; titers for tetanus antibodies were 0.08 (normal > 0.01) and for diphtheria, 0.1 (normal > 0.01).

At age 10 the patient developed twenty-nail dystrophy (Fig. 2) which first affected the fingernails and later the toenails. There was longitudinal ridging with layering, fragility, and dullness. Fungal cultures were negative. Tapering of systemic corticosteroids was associated with an exacerbation of the nail dystrophy. The texture and appearance of the skin, hair, teeth, and oral mucosal surfaces have remained normal. Family history for hair or skin disorders,
allergies, or autoimmune diseases is negative. Extensive investigations for autoantibodies have been negative, including antibodies to thyroid, adrenal, parietal cells, and neutrophils. He has had no increased incidence of bacterial and/or viral infections except for a recent episode of Herpes zoster.

Quantification of immunoglobulin levels over the years revealed the highest values for IgG, IgM, and IgA to be 6.92 (4.41–11.35), 0.91 (0.47–2.00), and 1.30 (0.56–3.52) g/l respectively; lowest values were 3.43, 0.25, and 0.46 g/l. The immunoglobulin subtypes on two separate occasions were: IgG₁ = 3.38 and 3.54 g/l (4.70–13.00), IgG₂ = 0.92 and 0.75 g/l (1.15–7.50), IgG₃ = 0.78 and 0.67 g/l (0.20–1.30), IgG₄ = 0.036 and 5 g/l (0.02–1.65). At these times the IgG values were 5.45 g/l and 4.59 g/l respectively.

DISCUSSION

The nail abnormalities in this patient are similar to those occurring in children with twenty-nail dystrophy (1). The etiology of the disorder is unknown. However,
autoimmune processes and immunologic abnormalities have been implicated in other conditions with similar nail changes; e.g. alopecia areata, and psoriasis (4, 5).

In our patient evidence for an autoimmune mechanism as a cause of twenty-nail dystrophy was the occurrence of chronic immune thrombocytopenic purpura, autoimmune hemolytic anemia, and persistently low levels of immunoglobulins, a combination of events that has been described in other patients with immunodeficiency disorders (3). The thrombocytopenia and anemia are secondary to autoantibodies. The intermittently low levels of immunoglobulins may be secondary to a similar mechanism, although corticosteroid mediated suppression of T-cell function may also be a factor. However, immunoglobulins continued to be decreased, even when corticosteroid therapy was discontinued for approximately eight months.

Alopecia areata has been observed in a patient with immune thrombocytopenia and autoimmune hemolytic anemia (4). Also autoimmune mechanisms have been implicated in the pathogenesis of alopecia areata, lichen planus, and psoriasis, conditions of ectodermal derivatives that are often associated with twenty-nail dystrophy (4, 5). Twenty-nail dystrophy of childhood may also be an autoimmune disorder in which the antibody is directed against some component of keratin. Anti-keratin antibodies have been demonstrated in the serum of patients with psoriasis, rheumatoid arthritis, systemic sclerosis, ankylosing spondylitis, and systemic lupus erythematosus (6). However, no anti-keratin antibodies could be detected in our patient’s serum utilizing immunoblot techniques. Possibly the antibody is against some other component of the nail or a cellular rather than a humoral mechanism may be operative.
REFERENCES


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