
Correspondence

MALIGNANT IDIOPATHIC ANAPHYLAXIS DOES EXIST IN BRAZIL

To the Editor:

Idiopathic anaphylaxis (IA) was first reported in 1978.¹ IA is an acute syndrome with no defined etiology in which all or some of the following symptoms may be present: urticaria, angioedema, airway obstruction with laryngeal edema, bronchospasm, gastrointestinal manifestations, and cardiovascular events including shock leading to a potentially life-threatening medical emergency. No exogenous antigenic or physical stimulus, such as exercise, can be detected, but prognosis continues to remain good with the majority of patients achieving remission with corticosteroid therapy.² Malignant IA is the most severe form of this clinical entity, in which episodes of potentially fatal anaphylaxis are not controlled when prednisone is reduced below 60 mg every other day or 20 mg daily.³

A 66-year-old Brazilian nonatopic male presented with a history of four recurrent episodes of life-threatening anaphylaxis for the last 2 years. The clinical presentations during these events included: sudden malaise, weakness, sweating, difficulty in breathing, nausea, vomiting, increased peristalsis with diarrhea, anxiety, hyperinflation, altered consciousness, syncope, marked hypotension, tachycardia, shock, transient pulmonary edema (adult respiratory distress syndrome) requiring emergency treatment in the intensive care unit with assisted ventilation and cardiovascular support, reversed with epinephrine, antihistamines, and large doses of intravenous methylprednisolone. Associated medical conditions included: type 2 diabetes mellitus controlled with oral hypo-

glycemic agents, arterial hypertension stabilized with calcium channel inhibitors and α -blockers, a successful coronary artery bypass surgery after a myocardial infarction, and a mild benign prostatic hypertrophy. Laboratory data showed 12% blood eosinophilia ($1,296 \text{ cells/mm}^3$), and a bone marrow aspirate demonstrated the presence of eosinophilia without mastocytosis. The following extensive work up was entirely negative or within normal limits: rest of the CBC, platelets, ESR, CD19, CD3, CD4, CD8, serum tryptase, VDRL, anti-HIV 1 and 2, antiphospholipid, anti-La, anti-Ro, anti—double-stranded DNA, antithyroid antibodies, ANA, rheumatoid factor, hepatitis A, B, and C antigen markers, serum immunoglobulins (IgG, IgA, IgM, IgE), C1q, C2, C3, C4, CH50, C1-esterase inhibitor, routine biochemistry including serum transaminases and LDH, iron, TIBC, ferritin, complete coagulogram, free T4, TSH, 24-hour urinary level of 5-HIAA, UA, stool guaiac, ova and parasites (four specimens), chest x-rays, and routine allergy prick skin tests to inhalant and food antigens. The patient was instructed on how to use epinephrine auto-injector and placed on maintenance treatment with oral prednisone 20 mg, daily, switched later to 40 mg every other day to be tapered 5 mg every 2 weeks; daily oral fexofenadine 120 mg; ranitidine 300 mg; ketotifen 4 mg; and oral albuterol 2 mg, three times daily. When the alternate day prednisone dose reached 25 mg, another severe, similar life-threatening anaphylactic episode occurred requiring tracheal intubation and ventilatory assistance. He was then placed on a fixed, daily AM oral prednisone dose of 20 mg. For the last 15 months he is well controlled and completely asymptomatic on this established therapeutic regimen.

It is possible that immediate allergic reactions to self-peptides as in an autoimmune process might be present in the pathogenesis of IA. Various histamine-releasing factors might also be released in an uncontrolled manner.¹ Acute IA patients compared with those in remission have a significantly higher percentage of CD3⁺HLA-DR⁺ cells. Patients with acute IA on prednisone as well as IA patients in remission have a significantly higher percentage of CD19⁺CD23⁺ cells than in normal subjects.⁴ This suggests that there are more activated T cells in patients with acute IA than in patients in remission. IA is therefore an immunologic disorder with activated T cells that are effectively suppressed by steroids.

To our knowledge this is the first Brazilian report on IA and additional cases will probably follow.

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