Usefulness of serum eosinophil cationic protein (ECP) in predicting relapse of Churg and Strauss vasculitis

Sirs,

Eosinophil cationic protein (ECP), a cytotoxic granule protein of eosinophils known to play a major role in defence against parasites (1), has been shown to increase in various disorders (2-6) including acute flares of Churg and Strauss syndrome (CSS) (7, 8). We report a case of CSS with elevation of serum ECP months before onset of symptoms on two occasions over 5 1/2 years. High ECP titers preceded blood eosinophilia. C-ANCA titers did not rise before onset of symptoms. This 52-year-old asthmatic white male was diagnosed with CSS in February 1987. He then fulfilled the 1990 criteria for the classification of CSS (2). Treatment was successful and medication was discontinued in October 1993. ECP measurements were begun at that time using a fluorescent enzyme immunoassay (Pharmacia, Sweden) according to the manufacturers’ instructions with a normal value of < 12 µg/L.

Self-limited cutaneous relapses occurred in March 1994, April 1995 and July 1995. ECP titers rose moderately from January 1994 to March 1995, then dramatically with titer increases, reaching 92 µg/L by October 5 (Fig. 1). At that time, an episode of fever, epigastralgia and pruriginous erythema was complicated by renal, cardiac and CNS involvement with vasculitic lesions on a cerebral MRI. Arrhythmia prompted admission to intensive care. Treatment was successful and followed by a normalisation of ECP titers. Before this flare, neither eosinophil counts nor CRP had started rising until after serum ECP had increased notably. Titors remained low throughout 1996. One episode of skin rash was noted and attributed to contact dermatitis on the basis of history. In June 1997, an episode of productive coughing with normal chest X-ray and eosinophil counts improved with inhaled salmeterol. ECP levels rose during this episode and fell on remission, but did not reach normal values. From then until April 1998, there were several episodes of self-limited erythema without eosinophilia that were attributed to allergy but not documented as such. A second episode of coughing responded to inhaled steroids.

In August 1998, a productive cough with decreased peak expiratory flow rate, erythema and a normal chest X-ray required the admission to intensive care. Treatment was successful and followed by a normalisation of ECP titers. Before this flare of CSS, neither ANCA titers nor CRP were found to be useful in the clinical follow-up. ECP, on the other hand, seemed to match disease activity closely and its rise preceded symptoms. ECP monitoring has been suggested in asthma with the caution that it may not add information to clinical and lung function parameters (9). Clinical monitoring of CSS is often less clear than that of asthma and its pathogenesis is more clearly dominated by eosinophil activation. The lack of a complete correlation between ECP and eosinophilia also makes this marker interesting. It has been suggested elsewhere that ECP may reflect eosinophil activation (10). Monitoring ECP in asymptomatic CSS patients could have real clinical utility if it proves to be a sensitive marker of disease activity. Prospective studies would be necessary to determine if this is indeed the case.

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References

Fig. 1. Variations of biological markers and treatment during the course of disease. Initial doses are indicated. Pulse iv methylprednisolone was administered on re-introduction of steroid therapy in October 1995. AZ (Asathioprine), MP (Methylprednisolone), PN (Prednisone), CY (Cyclophosphamide).
Pericarditis as a presenting feature of Henoch-Schönlein purpura

Sirs,

Henoch-Schönlein purpura (HSP) is an inflammatory vascular disease, characterized by involvement of the skin, gastrointestinal tract, joints and kidneys (1). The presenting triad in childhood is usually represented by non-thrombocitopenic purpura, abdominal pain, and arthritis or arthralgia. The pediatric form is generally considered a benign and self-limited disorder, and cardiac involvement is exceptionally rare. When present, cardiac involvement with or without pericarditis is usually an isolated finding. HSP presenting with myocardial complications is extremely rare, with to our knowledge very few reports published so far (2-6).

Pericarditis can be associated with systemic infections such as infections (viral, bacterial or fungal), neoplasms and endocrine/metabolic disorders, or autoimmune diseases. In particular, it is frequent in the context of systemic lupus erythematosus, rheumatic fever, familial Mediterranean fever and systemic onset juvenile arthritis. However, in HSP cardiac involvement is extremely rare, with to our knowledge very few reports published up to now (2-6). Only in one case has pericarditis been described (6), the other reports describing patients with myocarditis/heart failure.

We conclude that HSP should be added to the differential diagnosis of pericarditis of unknown cause in childhood.

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Fig. 1. Cardiac ultrasound showing a pericardial effusion anteriorly and at the apex.