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CORRESPONDENCE

Chronic eosinophilic pneumonia (CEP) as a presenting feature of Churg-Strauss syndrome (CSS)

To the Editor:

We were very interested in the Case Report by HUETO-PEREZ DE HEREDIA *et al*. [1], since we have had the opportunity to observe three very similar cases.

Three patients (2 females and 1 male) with clinico-radiological features typical of chronic eosinophilic pneumonia (CEP) (mean 8 yrs) later developed systemic complications (cutaneous vasculitis and multiplex mononeuritis) coinciding with steroid weaning.

Diagnosis of Churg-Strauss syndrome (CSS) was assessed by skin biopsy and/or transbronchial biopsy, and treatment with high dose steroids and cyclophosphamide was successfully performed.

Interestingly, in our patients the overlap period was characterized by an increased polyclonal immunoglobulin A (IgA) level in serum (mean 470 $mg \cdot dl^{-1}$; normal level 75–350 $mg \cdot dl^{-1}$). The single previous report of an association between IgA and CSS described the identification of IgA in glomerular deposits [2].

According to the recent literature, a possible explanation for an increase in IgA could be a dysregulation of the CD4+ lymphocytes, Th2 [3]. Indeed, the Th2 lymphocyte pathway has been incriminated in several diseases, such as allergic rhinitis, asthma and atopic dermatitis. These cells produce different cytokines implicated in immunoglobulin E (IgE) production (interleukin-4 (IL-4)), IgA production (interleukin-5 (IL-5)), and eosinophil growth and activation (interleukin-5 (IL-5)) [4].

Another particular finding was that multisystemic involvement occurred at the same period of the year (May) in the three subjects. The role of an inhaled antigen in the CSS is accepted by the majority of authors [3]. Could it be that a Springtime respiratory antigen played a role as a trigger for the disease evolution in our patients? This question remains to be answered. To the best of our knowledge, three other case reports of CSS following CEP are reported in literature [5–7]; two of them also developed systemic manifestations in May or late April [6, 7].

In conclusion, we confirm that there is a possible evolution of some forms of CEP towards CSS. Elevated IgA in serum could be a biological marker of this transition.

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