Letters to the Editor

A retrospective evaluation of the treatment of 7 patients with Churg-Strauss syndrome

Sir,

Since the description of the clinical entity named Churg-Strauss syndrome (C-S syndrome) by Jacob Churg and Lotte Strauss in 1951 (1) the optimal treatment of the disease has been debated. The main treatment options are the use of corticosteroids alone or the use of corticosteroids in combination with cytotoxic drugs such as cyclophosphamide. Some reports have suggested that the use of cytotoxic drugs together with steroids should be confined to those patients with the most severe prognosis (2, 3).

Guillivin and co-workers have suggested a useful denominator of severe disease, the so-called "five-factor score", which defines factors of particular importance for the prognosis. These are cardiomyopathy, CNS involvement, gastrointestinal tract involvement, renal failure and proteinuria > 1 g/day (4). In this respect it was of interest to determine whether considerations concerning the prognosis of the disease as described by the "five-factor score" had influenced the treatment of patients at our clinic.

To study this point, 7 patients diagnosed as having C-S syndrome between 1993 and 1995 were evaluated. Of the 7 patients, 2 had severe organ involvement as defined by the "five-factor score" (see Table I). One had gastrointestinal (GI) involvement as well as myocardial infarction and the other had CNS involvement, i.e. migraine. The other 5 patients did not develop signs of severe organ involvement but had other symptoms and signs compatible with the C-S diagnosis such as asthma, sinusitis, pulmonary infiltrates, polyneuropathy, blood eosinophilia and a positive skin biopsy revealing eosinophilic infiltrates together with vasculitis and/or granuloma (5).

Somewhat disappointingly, it was not possible to discern a pattern in the treatment strategies. The initial treatment in 4 of the 7 patients was prednisone in combination with cyclophosphamide. One of patients had GI and myocardial involvement indicating severe disease with a more negative prognosis. The other 3 treated with the combination of prednisone and cyclophosphamide did not, however, display symptoms included in the "five-factor score". Of the remaining 3 patients, 2 were treated with corticosteroids alone, even though one of them developed migraine which is compatible with CNS involvement. One patient was treated with the combination of corticosteroids and azathioprine. Neither did this patient develop severe organ symptoms. The reason for the use of azathioprine was its feasibility as a steroid-saving drug.

It may thus be concluded that: (i) no clear strategy was used at our clinic between 1993 and 1995 in the treatment of C-S disease with reference to the severity and prognosis of the disease; and (ii) use of the "five-factor score" offers an option to divide patients into two categories with different prognoses. With this in mind prospective studies of C-S syndrome may be undertaken in which different treatment strategies are used and the long-term outcome is evaluated.

P. LARSSON
I. LUNDBERG
Department of Rheumatology, Karolinska Hospital, Stockholm, Sweden.
Please address correspondence and reprint requests to: Per Larsson, MD, PhD, Dept of Rheumatology, Karolinska Hospital, S-17176 Stockholm, Sweden.
E-mail: plarsson@rheum.ks.se

References

1. CHURG J, STRAUSS L: Allergic granulomatosis, allergic angiitis, and periarteritis nodosa. *Am J Pathol* 1951; 27: 277-301.
2. FAUCI AS, HAYNES BF, KATZ P, WOLFF SM: Wegener's granulomatosis: Prospective clinical and therapeutic experience with 85 patients for 21 years. *Ann Int Med* 1983; 98: 76-85.
3. LHOTE F, GUILLEVIN L: Polyarteritis nodosa, microscopic polyangiitis and Churg-Strauss syndrome. Clinical aspects and treatment. *Rheum Dis Clin North Am* 1995; 21: 911-48.
4. GUILLIVIN L, LHOTE F, GAYRAUD M *et al*.: Prognostic factors in polyarteritis nodosa and Churg-Strauss syndrome. A prospective study in 342 patients. *Medicine* 1996; 75: 17-28.
5. MASI AT, HUNDER GG, LIE JT *et al*.: The American Collage of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). *Arthritis Rheum* 1990; 33: 1094-100.

Table I. Summary of clinical findings in 7 patients with Churg-Strauss syndrome.

Modalities Sex, age (yrs.)	Case 1 M, 36 yrs.	Case 2 M, 61 yrs.	Case 3 F, 25 yrs.	Case 4 F, 36 yrs.	Case 5 F, 46 yrs.	Case 6 M, 56 yrs.	Case 7 F, 70 yrs.
Juvenile onset asthma	Yes. Resolved at age 16	No	Yes	Yes	No	No	No
Adult onset asthma	Yes. Symptoms 2 years before diagnosis	Yes. symptoms 20 years before diagnosis	Asthmatic symptoms since 9 yrs. of age	Asthmatic symptoms since 15 yrs. of age.	Yes. Symptoms 1 1/2 yrs. before diagnosis.	Yes. Symptoms 10 years before diagnosis	Yes. Symptoms 10 years before diagnosis
Results of asthma therapy	No improvement	Not evaluated	Not evaluated	Not evaluated	No improvement	No improvement	No regular therapy prescribed
Sinusitis	Yes	Uncertain	Yes	Yes	Yes	Yes	No
Polyneuropathy	No	Yes	Yes	Yes	No	Yes	Yes
Pulmonary infiltrates	Yes	Yes	Yes	No	Yes	No	Yes
Skin biopsy-proven vasculitis or granuloma	Yes	Not performed	Yes	Yes	Not performed	Yes	Not performed
Severe organ involvement[1]	Yes, heart and GI involvement at presentation	No	No	Suspected miagraine 4 mos. after presentation	No	No	No
"Five factor" score (15)	2	0	0	1	0	0	0
Initial treatment	I.v. pred. + oral CYC	Oral pred. + oral CYC	Oral pred.	Oral pred.	Oral pred. + oral CYC	Oral pred. + i.v. CYC	Oral pred.+ oral AZA

Pred. = Prednisolone ; CYC = Cyclophosphamide ; AZA = azathioprine