Reumatol Clin. 2010;**6(2)**:76–81

Reumatología Clínica

www.reumatologiaclinica.org

Original article

Churg-Strauss Vasculitis. Description of 9 cases

Emma García-Melchor,* Sonia Mínguez Blasco, Anna Moltó Revilla, Lourdes Mateo Soria, Susana Holgado Pérez, and Alejandro Olivé Marqués

Sección de Reumatología, Hospital Universitari Germans Trias i Pujol, Badalona, Barcelona, Spain

ARTICLE INFO

Article history:
Received September 25, 2008
Accepted February 11, 2009

Keywords:
Vasculitis
Churg-Strauss syndrome

ABSTRACT

Introduction: Churg-Strauss Syndrome (CSS) is a necrotizing vasculitis affecting small to medium-sized vessels, characterized by lung involvement, asthma and peripheral blood eosinophilia, and pathologically by the presence of granulomas and eosinophilic infiltrates.
Objectives: This report analizes the characteristics of 9 patients with SCS diagnosed in an university referral center.
Patients and methods: Retrospective study. Between 1984 and 2007 nine patients with SCS were diagnosed in our center. Epidemiological, clinical, laboratory test as well as pathologic studies and treatment required were retrospectively analyzed.
Results: Nine patients (7 males). The mean age at the time of diagnosis was 51 years (range 23-76 years). Eight of these patients had history of asthma. The more frequent organs involved were the skin (66%), musculoesqueletical system (66%), peripheral nervous system (55%) and the lung (55%). All patients presented peripheral eosinophilia. ANCA positivity was demonstrated in 6 patients (66%), most of the patients with the p-ANCA pattern. All patients were treated with corticosteroids, and in 8 immunosupressant treatment was required, mainly cyclophosphamide.
Conclusions: In this report, 9 patients with SCS are presented. Clinical characteristics are similar with the observed in other reports. We observed a major positivity of ANCA. Most of the patients were treated with corticosteroids and inmunosupresants, but the treatment should be tailored depending on the involvement of the patient.

Vasculitis de Jacob Churg y Lotte Strauss. Descripción de 9 casos

Palabras clave:
Vasculitis
Síndrome Churg-Strauss
Anticuerpos anticitoplasma de neutrófilo

RESUMEN

Introducción: El síndrome de Churg-Strauss (SCS) es una vasculitis que afecta a vasos de pequeño y mediano calibre, caracterizándose clínicamente por afectación predominante del aparato respiratorio, asma y eosinofilia periférica y anatomopatológicamente por la presencia de granulomas y la infiltración tisular por eosinófilos.
Objetivos: En el presente trabajo se detallan las características de una serie de 9 pacientes con SCS diagnosticados en un centro universitario de referencia.
Pacientes y métodos: Estudio retrospectivo: entre 1984 y 2007 se diagnosticaron 9 pacientes con SCS en nuestro centro. De todos ellos se obtuvieron de forma retrospectiva datos epidemiológicos, clínicos, analíticos, estudios anatomopatológicos y tratamiento recibido.
Resultados: De estos 9 pacientes, 7 eran hombres. La edad media en el momento del diagnóstico fue de 51 años (rango 23-76 años). Ocho de los pacientes tenían antecedente de asma bronquial. Las manifestaciones clínicas más frecuentes fueron las cutáneas (66%), musculoesqueléticas (66%), del sistema nervioso periférico (55%) y las pulmonares (55%). Todos presentaban eosinofilia periférica. Los anticuerpos anticito-

* Corresponding author.
E-mail address: emmitagm@gmail.com (E. García-Melchor).

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plasma de neutrófilo (ANCA) fueron positivos en 6 pacientes (66%), la mayor parte de ellos con patrón perinuclear en inmunofluorescencia (p-ANCA). Todos los pacientes recibieron tratamiento con glucocorticoides y 8 de ellos requirieron, además, tratamiento inmunosupresor, principalmente ciclofosfamida.
Conclusiones: En este trabajo se presentan 9 pacientes diagnosticados de SCS. Las manifestaciones clínicas no difieren de las observadas en el resto de series publicadas. Sin embargo, en esta serie se observa una mayor positividad de ANCA. La mayoría de los pacientes fueron tratados con glucocorticoides e inmunosupresores, debiéndose individualizar el tratamiento de cada paciente según el grado de afectación.

Introduction

Churg-Strauss syndrome (CSS) is a vasculitis of small and medium-caliber vessels that affects arterioles, capillaries and venules. It is clinically characterized by predominant involvement of the respiratory system, asthma and peripheral eosinophilia. Pathologically, the presence of granulomas and tissue infiltration by eosinophils is seen. Although respiratory manifestations are the most common, it is a systemic vasculitis and may present skin, cardiovascular, neurological, gastrointestinal or kidney involvement.[1]

This syndrome was first described in 1951 by Jacob Churg and Lotte Strauss as a result of the necropsy study of 13 patients with asthma and systemic vasculitis.[2] However, by 1939 Rackemann and Greene had observed that patients with asthma had a special type of polyarteritis nodosa.[3] Churg and Strauss called this new entity 'allergic granulomatous angiitis'. The diagnosis was based on the presence of asthma, peripheral eosinophilia and systemic vasculitis, with necrotizing vasculitis, an inflammatory infiltrate composed of eosinophils, and extravascular granulomas observed in the histological study.

Subsequently, Chumbley questioned the fact that the presence of granulomas was a prerequisite to diagnosing this entity.[4]

Lanham[5] proposed less stringent criteria than those by Churg and Strauss (Table 1), since it was not necessary to confirm the disease through biopsy, proposing: the presence of asthma, peripheral eosinophilia and vasculitis of two or more organs, making it able to perform a bedside diagnosis.

In 1990 the American College of Rheumatology (ACR) established a set of criteria for each type of vasculitis[6] in order to create homogeneous groups of patients in studies (Table 1). In the case of CSS, in order to establish the diagnosis, 4 of these 6 criteria are needed, reaching 85% sensitivity and a specificity of 99.7%.

At the conference in Chapel Hill, vasculitidies were classified according to the histopathologic characteristics based on the size of vessel affected.[7] CSS was defined as a small vessel granulomatous vasculitis with an inflammatory infiltrate, predominantly eosinophils, which affects the upper respiratory tract in association with asthma and peripheral eosinophilia. Within the group of small vessel vasculitis, it was grouped with Wegener's granulomatosis and microscopic polyangiitis by its association with neutrophil cytoplasmic antibodies (ANCA).

The following describes the epidemiological, clinical, laboratory data and treatment characteristics of a series of 9 patients with CSS diagnosed in our center.

Table 1
CSS diagnostic criteria[2,5,6]

Churg and Strauss criteria (1951)
1. Asthma
2. Peripheral eosinophilia
3. Necrotizing vasculitis, infiltration of eosinophils and granulomas
Lanham criteria (1984)
1. Asthma
2. Peripheral eosinophilia (> 1,500 eosinophils/mm^3)
3. Vasculitis of two or more extrapulmonary organs
ACR criteria (1990)
1. Asthma
2. Peripheral eosinophilia (> 10% of total WBC)
3. Sinus involvement
4. Pulmonary infiltrates
5. Mononeuritis multiplex
6. Histology compatible with vasculitis
The diagnosis is established when four of these six criteria are present

ACR indicates American College of Rheumatology; CSS, Churg-Strass Syndrome.

Patients and methods

Between January 1984 and December 2007, nine patients were diagnosed with CSS at the Hospital Universitari Germans Trias i Pujol, which covers a reference area of 800,000 inhabitants.

Patients were identified retrospectively in the CIPER database, which contains all patients seen by the Section of Rheumatology at the center since 1984. Eighty eight percent of patients met the ACR or Lanham diagnostic criteria.

We collected epidemiological data of all of them, as well as clinical and laboratory tests results such as blood count (Coulter), ESR, biochemical and immunological tests: antinuclear antibodies (ANA) (Hep-2), rheumatoid factor (nephelometry) and ANCA (direct immunofluorescence Menarini® kits). We calculated the Five Factor Score[15] for each and collected information about the treatment they received.

Results

Between 1984 and 2007, 354 cases of vasculitis were diagnosed in our department, 9 of which (2.5%) were of CSS (Figure). Seven patients were men (77%) and 2 women (22%). The mean age at diagnosis was 51 years (range 23-76 years). Eight of the 9 patients (88%) met the Lanham criteria for classification, while 6 (66%) fulfilled the ACR criteria.

The retrospective study of medical records did not find any triggering infections, or history of vaccination, or the prior treatment with new drugs. It is noteworthy that one case was diagnosed during the postpartum period. Clinical manifestations are summarized in Tables 2 and 3.

Eight of the patients had a history of asthmatic bronchitis (88%), and the interval between onset of asthma and the diagnosis of vasculitis was 12.6 years (range 1-40 years). A history of atopy (eczema, drug allergy, allergic rhinitis, urticaria) was present in 2 cases (22%).

Sinus involvement was seen in 4 patients (44%), two of them diagnosed through a sinus CT. The remaining 2 had no imaging evidence, but one showed clinical manifestations compatible with recurrent sinusitis and the other had presented a single episode of sinusitis. Two patients (22%) had history of nasal polyps that had required surgery. After asthma, the most frequent clinical

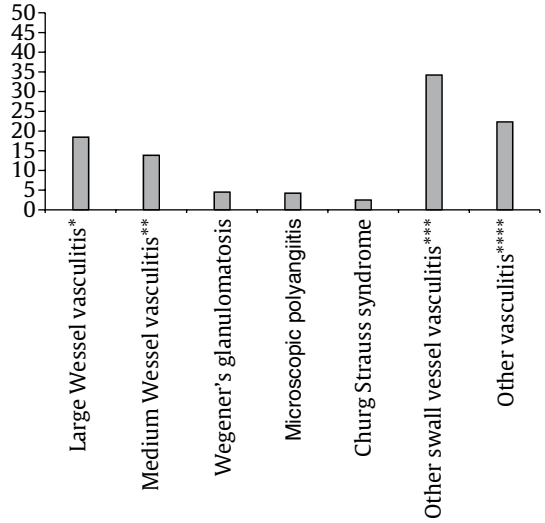

Figure. Percentages of different vasculitis in our service.
* Large vessel vasculitis: giant cell arteritis and Takayasu arteritis.
** Medium caliber Vasculitis: polyarteritis nodosa and Kawasaki disease.
*** Other small-vessel vasculitis: cryoglobulinemic vasculitis, hypersensitivity vasculitis and Henoch-Schönlein purpura.
**** Other vasculitis: Behçet disease, erythema nodosum, paraneoplastic, serum sickness.

Table 2
Main characteristics of the 9 patients

	n (%)
Asthma	8 (88%)
Fever	8 (88%)
Sinusitis	4 (44%)
Atopy	2 (22%)
Nasal polyps	2 (22%)
Skin	6 (66%)
Palpable purpura	3/6 (50%)
Exanthema	2/6 (33.3%)
Nailbed hemorrhage	2/6 (33.3%)
Subcutaneous nodules	1/6 (16.6%)
Erythematous macules	1/6 (16.6%)
Musculoskeletal	6 (66%)
Muscle pain	4/6 (66.6%)
Joint pain	4/6 (66%)
Arthritis	2/6 (33.3%)
Lung	5 (55%)
Pleural effusion	3/5 (60%)
Pulmonary infiltrates	1/5 (20%)
Pulmonary hemorrhage	1/5 (20%)
Peripheral nervous system	5 (55%)
Gastrointestinal	3 (33%)
Abdominal ischemia	1/3 (33%)
Granulomatous ileitis	1/3 (33%)
Colitis	1/3 (33%)
Renal	2 (22%)
Focal and segmental glomerulonephritis	
Central nervous system	1 (11%)

manifestations were skin, musculoskeletal, peripheral nervous system and pulmonary.

Six patients (66%) had skin lesions classifiable as palpable purpura, which was the most common dermatological manifestation; others had erythematous rash, subcutaneous erythematous nodules, splinter haemorrhages and nailbed erythematous macules on the back of the hands and feet. The pathology of these lesions showed an eosinophil-rich infiltrate, leukocytoclastic vasculitis, with granulomas in only one case.

Joint involvement consisted of joint pain, myalgia, and ankle arthritis in one patient, and another with symmetrical arthritis of the small joints of the hands. The peripheral nervous system manifestations were present in five patients (55%) and in all cases it was a multiple mononeuritis confirmed by electromyogram.

Radiological lung involvement was seen in 5 patients (55%), 3 with eosinophil-rich pleural effusions (right sided in two cases and one left sided), and one case with transient alveolar infiltrates and a case of alveolar hemorrhage confirmed by bronchoscopy. Gastrointestinal symptoms, heart, kidney and central nervous system were less frequent.

Gastrointestinal involvement was present in 3 patients. One of these patients had an intestinal occlusion due to granulomatous ileitis. The second case was diagnosed with colitis by colonoscopy and had a biopsy compatible with vasculitis, following a study due to bloody diarrheal stools. The third patient had clinical manifestations compatible with intestinal angina. One patient had cerebellar cortical infarcts. Two patients had renal involvement in the form of proteinuria; both cases underwent renal biopsy with results consistent with focal segmental necrotizing glomerulonephritis. Finally, one patient showed repeated episodes of pericarditis during follow-up, with no accompanying eosinophilia, which was not resolved with glucocorticoids, but rather with non-steroidal anti-inflammatory drugs, so it was classified as viral pericarditis, more than as a manifestation of vasculitis.

Regarding laboratory tests, all patients had peripheral eosinophilia at the time of diagnosis, with a median of 15.4×10^9 eosinophils/l (range 1.2 to 28×10^9 eosinophils/l). The median ESR was 40 mm in the 1st h (range 18-118 mm). Were determined IgE levels in 8 patients, and found them elevated in 6, with average values of 1.256 UI/ml (range 102-45.550 UI/ml. ANA were positive in one patient, with a titer of 1/160 and a speckled pattern. ANCA were determined in all patients, and were positive in 6 (66%), two of them with a cytoplasmic pattern and 4 with a perinuclear pattern.

A total of 23 biopsy samples were obtained, all except one showed changes consistent with the diagnosis of CSS. Skin biopsies (4 total) showed leukocytoclastic vasculitis with an infiltrate composed of eosinophils. Only one of the patients presented granulomas.

We performed two kidney biopsies in patients with proteinuria, which showed focal segmental necrotizing glomerulonephritis. One of them also had necrotizing tubulointerstitial nephritis with diffuse eosinophilia.

Of the six muscle biopsies performed, one was normal, the remaining showed necrotizing vasculitis of small and medium size arteries with an inflammatory infiltrate with predominance of eosinophils.

One patient had an intestinal perforation secondary to obstruction requiring surgery. A sample of ileus was obtained showing intra and extravascular granulomas with necrosis and eosinophilic infiltrates.

Another patient had bloody diarrheal stools, with a biopsy of the colon mucosa showing leukocytoclastic capillaritis, eosinophilia vascultitis of the lamina propria and submucosa with fibrinoid necrosis. In one patient a temporal artery biopsy showed signs of necrotizing vasculitis with a moderate infiltrate of eosinophils.

In another case, while monitoring after a prostatectomy due to prostatic syndrome, an adenomatous prostate specimen with eosinophilic infiltration was documented.

We calculated the Five Factor Score[16] of all of the patients, with 5 patients having 0, 1 in 2 patients and 2 in 2 patients.

Table 3
Characteristics of patients in the series

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Gender	Male	Male	Female	Male	Male
Age at diagnosis, years	56	51	23	75	57
Interval asthma-vasculitis, years	2	1	1	19	5
History	Asthma Sinusitis	Asthma Sinusitis	Asthma Nasal polyps	Asthma AAS allergy	Asthma Nasal polyps
Clinical vasculitis	Glomerulonephritis Multineuritis	Muscle pain Pleural effusion Abdominal angor Multineuritis	Joint pain, muscle pain Ankle arthritis Palpable purpura Pleural effusion Multineuritis	Exanthema Granulomatous ileitis	Muscle pain, joint pain Palpable purpura Nailbed hemorrhages Pulmonary hemorrhage Multineuritis
Eosinophils, cels/l	13.3×10^9	28×10^9	25.8×10^9	8.1×10^9	15.4×10^9
ANCA	Positive (p-ANCA)	Negative	Positive (c-ANCA)	Negative	Positive (p-ANCA)
IgE, UI/ml	441	339	102	996	4,550
Biopsies	Renal: necrotixing focal and segmental glomerulonephritis Bone marrow: eosinophilia	Muscle and nerve: vasculitis with eosinophilic infiltrate Bone marrow: eosinophilia	Skin: leokocytoclstic vasculitis with eosinophilia Muscle: medium caliber vessel vasculitis with eosinophilia Bone marrow: eosinophilia	Skin: leukocytoclstic vasculitis Ileus: necrotizing vasculitis with intra and extravascular granulomas and eosinophilic infiltrate Bone marrow: eosinophilia	Skin: leukocytoclastic eosinophílic vasculitis Muscle: necrotizing small caliber vessel vasculitis with eosinophils Bone marrow: eosinophilia
Induction therapy	PDN 1 mg/kg/day v.o. CF 100 mg/day v.o.	PDN 1 mg/kg/day v.o. CF 100 mg/day v.o.	MTP 1 g/day 3 days ev. CF 2 mg/kg/day v.o. PDN 1 mg/kg/day v.o. Gammaglobulin 400 mg/kg/day 5 days ev.	MTP 1 g/day 3 days ev. PDN 1 mg/kg/day v.o.	MTP 1 g/day 3 days ev. PDN 1 mg/kg/day v.o. CF ev.

	Patient 6	Patient 7	Patient 8	9
Gender	Female	Male	Male	Male
Age at diagnosis, years	28	69	76	27
Interval asthma-vasculitis, years	Unknown	40	20	Non asthmatic
History	Asthma Sinusitis	Asthma Atopy	Asthma	Allergic rhinitis
Clinical vasculitis	Joint and muscle pain Erythematous maculas Nailbed hemorrhages Anterior pyramidal syndrome D6 Ischemic cerebral and cerebellar lesions	Mononeuritis	Arthritis Erythematous nodules Lung infiltrate Pleural effusion	Joint pain Purpura Lung infiltrate Glomerulonephritis Colitis
Eosinophils, cels/l	17.4×10^9	23.2×10^9	1.3×10^9	5.8×10^9
ANCA	Negative	Positive (p-ANCA)	Positive (p-ANCA)	Positive (p-ANCA)
IgE, UI/ml	20	50	Undetermined	1,110
Biopsies	Muscle: fibrinoid necrosis of medium and small caliber vessels Bone marrow: eosinophilia	Muscle: necrotizing vasculitis of medium caliber vessels Temporal artery: necrotizing vasculitis with eosinophils Bone marrow: eosinofilia	Skin: granulomatous interstitial dermatitis with eosinophils	Skin: leukocytoclastic vasculitis with eosinophilia Renal: necrotizing focal and segmental glomerulonephritis with interstitial nephritis and diffuse eosinophilia Colon: leukocytoclastic capillaritis and eosinophilia of the lamina propia, vasculitis with fibrinoid necrosis of the submucosa
Induction therapy	MTP 500 mg/day 3 days ev. PDN 1 mg/kg/day v.o. CF ev.	MTP 500 mg/day 3 days ev. PDN 1 mg/kg/day v.o. CF ev.	PDN 1 mg/kg/day v.o. Methotrexate 10 mg/week v.o.	PDN 2 mg/kg/day CF ev.

AAS indicates aspirin; ANCA, anti-neutrophil cytoplasm antibodies; c-ANCA, anti-neutrophil cytoplasm antibodies with a cytoplasmic pattern under immunofluoresence; CF, cyclophosphamide; ev., intravenous; MTP, methylprednisolone; p-ANCA, anti-neutrophil cytoplasm antibodies with a perinuclear pattern under immunofluoresence; PDN, prednisone; v.o., orally.

Table 3
Main series of patients with CSS[8-11]

	Guillevin et al (1976-1982)	Solans et al (1977-1999)	Della Rosa et al (1989-2000)	Mi-Jung Oh et al (1995-2004)	Submitted series (1984-2006)
n	96	32	19	17	9
M/F	0.84	0.39	0.9	1.1	3.5
Average age	48	42	46	36	51
Asthma, %	97.9	100	100	100	88
Skin	49	68.8	68.4	58.8	66
Muscleskeletal	54.2	37.5	52.6	–	66
Mononeuritis	77.1	43.8	57.8	64	55
Lung	40.6	75	57.8	64.7	55
Gastrointest.	31.2	37.5	47.3	17.6	33
Heart	12.5	28.1	31.5	17.6	0
Renal	26	12.5	15	–	22
Eosinophilia	83.3	100	100	88	100
ANCA	47.6	53.8	35	5.9	66

ANCA indicates association with neutrophil cytoplasmic antibodies; CSS, Churg-Strauss Syndrome; F, female; M, male.

All patients received glucocorticoids, six of them in the form of an intravenous bolus. Eight patients required immunosuppressive therapy (88%): 7 with cyclophosphamide (4 intravenous and 3 oral) and one with methotrexate due to polyarthritis. One patient with multineuritis and was treated with intravenous immunoglobulin.

Discussion

CSS is a necrotizing vasculitis of small and medium vessels, which particularly affects the respiratory system, but can manifest in any organ, as shown in this series.

The demographic characteristics are similar to those of other series[8-11], although we noted the increased prevalence of men, and a somewhat higher average age in this study.

Lanham, in 1984, proposed less stringent classification criteria than those of Churg and Strauss, as his did not require histological confirmation.[5] About 66% of our patients met the ACR criteria, while 88% met the Lanham criteria. The difference lies mainly in those patients without involvement of the respiratory system, such as sinusitis or pulmonary infiltrates, which can be diagnosed with CSS according to the Lanham criteria but not with the ACR criteria. Only one patient met none of the criteria. This was a patient who was asthmatic and had no involvement of paranasal sinuses, but a history of allergic rhinitis, eosinophilia, palpable purpura, renal involvement and colon biopsy consistent with the diagnosis of CSS.

Although this vasculitis has been related to some drugs, especially with leukotriene antagonists used in the treatment of asthma,[12] the present study did not identify any precipitating factor.

According to Lanham, asthmatic bronchitis not associated with vasculitis usually begins before age 21, while in CSS patients, episodes of bronchospasm appear in adults.[5] In all of the subjects of this study, asthma respiratory symptoms began in patients over 21 years of age, with a mean age of onset of asthma of 45 years, supporting the hypothesis of Lanham. Particularly striking is the low incidence of rhinitis in this series, probably because it is a retrospective analysis and these symptoms may not be reflected in the patient's medical history. The other clinical manifestations were often similar to those observed in other published series (Table 4).

There is controversy over whether CSS should be considered an ANCA-associated vasculitis, as this association is not as strong as in the case of Wegener's granulomatosis, in which over 90% of patients had positive ANCA.[13] In fact, in a series published by Mi-Jung et al,[11] only 5.9% of patients had positive antibodies. The results of this study showed a higher positivity of ANCA compared to other European series,[8-10] with 66% of patients with positive ANCA.

Within the group of ANCA positive patients in our series, 66% showed a perinuclear immunofluorescence pattern (p-ANCA) with specificity against myeloperoxidase. In more recent series, there is a greater prevalence of this pattern over the cytoplasmic one, with 92.8% of patients presenting p-ANCA in a series by Solans[9] and 100% in the series by Della Rosa.[10]

Sable-Fourtassou et al observed, in a cohort of 112 patients, that there were differences between ANCA positive and ANCA negative patients.[14] The clinical manifestations were more clinically vasculitic (glomerulonephritis, and mononeuritis), whereas negative clinical manifestations were characterized by eosinophil tissue infiltration (heart, lung). Furthermore, in ANCA positive patients, the biopsy tended to have higher diagnostic performance, with a larger probability of finding vasculitis . Although it's difficult to extrapolate data, it is noteworthy that in the present study, the two patients with renal involvement were ANCA positive.

Regarding the usefulness of the biopsies, a total of 23 were performed and all but one showed changes consistent with CSS. Granulomas were observed only in 2 cases (22%), confirming the hypothesis by Chumbley that granulomas are not essential for diagnosis.[4]

Typically, CSS treatment consisted of corticosteroids. In fact, in the present study, all patients received this treatment. But now there is a tendency to be more aggressive and to use other immunosuppressants such as cyclophosphamide. Eight of our patients required immunosuppressive therapy with cyclophosphamide and methotrexate. The Five Factor Score (FFS, 'Index of prognostic factors')[15] is a set of five factors that associate this vasculitis to a worse prognosis, being: proteinuria > 1 g/24 h, renal insufficiency (serum creatinine > 1,6 mg/dl), cardiac, gastrointestinal and central nervous system involvement. According to the French group led by Guillevin, if a patient has none of these factors, treated with only glucocorticoids is acceptable, whereas if any of them should be present, immunosuppressive treatment should be added. In this series, of the 5 patients with no poor prognosis risk factors, all required immunosuppressive therapy due to mononeuritis multiplex, pulmonary hemorrhage or polyarthritis. Therefore, we must bear in mind that there are clinical manifestations that due to thedisability they produce (mononeuritis) or their severity (pulmonary hemorrhage), and although they are not included in the index of prognostic factors, should be treated aggressively.

The change in treatment strategy, together with a greater understanding of this condition by physicians and, therefore, an earlier diagnosis, are probably responsible for the fact that the prognosis of these patients has changed. From survival rate of 50% at 3 months in patients without treatment[1], we are now seeing a survival of more than 70% at 5 years.[8]

The limitations of this study are the retrospective nature of data collection and the limited number of patients, which did not allow for obtaining results in terms of clinical differences between ANCA positive and ANCA negative patients, or prognostic factors.

In conclusion, this paper presents 9 patients with CSS. The clinical features are not different from those observed in other published series. However, in this series there is a greater frequency of ANCA positivity. Most patients were treated with corticosteroids and immunosuppressants, making it necessary to individualize treatment for each patient according the degree of organ involvement.

Disclosures

The authors have no disclosures to make.

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