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Single Case

Churg-Strauss Syndrome or Eosinophilic Granulomatosis with Polyangiitis: Exuberant Classic Clinical Picture of a Rare Disease

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Keywords
Churg-Strauss syndrome · Allergic granulomatous angiitis · Vasculitis

Abstract
The authors present a classic case of Churg-Strauss syndrome with an exuberant clinical picture in a 34-year-old woman. She showed the following diagnostic criteria: asthma, polyneuropathy, rhinopathy, marked eosinophilia, positive p-ANCA with a perinuclear pattern, and skin histopathology results suggestive of vasculitis with eosinophils. There was a good response to prednisone, dexamethasone pulse therapy, and cyclophosphamide.

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Introduction

Churg-Strauss syndrome (CSS), or eosinophilic granulomatosis with polyangiitis, or allergic granulomatous angiitis, is a rare autoimmune disorder characterized by excess circulating tissue eosinophils and vasculitis, which affects the lung and skin. It is a primary vasculitis characterized by late onset of asthma, multisystem involvement, and eosinophilic vasculitis of small- and medium-sized vessels [1, 2]. This syndrome was first described by Churg and Strauss in 1951. It is a rare disease with an incidence of 0.5–2.7 cases per 1 million, affecting both sexes equally, and preferably middle-aged patients [3]. The disease usually begins with respiratory disorders like asthma or rhinitis, followed by, or simultaneous with, vasculitis [4].

Case Report

A 34-year-old woman with a history of asthma of difficult control and recent onset but without a history of atopy complaining of rhinitis, arthralgia, and skin lesions presented with palpable purpura on the lower limbs and bullae that evolved into necrosis in the acral regions (Fig. 1, 2).

A neurologic examination showed an abolished Achilles reflex and reduction of muscular strength in the left side of the body.

With a diagnostic suspicion of CSS, the patient was submitted to a skin biopsy, the histopathologic result of which disclosed epidermal necrosis (Fig. 3) and a dense dermal infiltrate occupying the interstice in a linear trail, with presence of eosinophils and extravasation of red blood cells (Fig. 4).

Rhinopathy and discrete bilateral polypoid degeneration were noted. Tomography of the facial sinuses demonstrated shrouded paranasal sinuses. The remaining examinations showed mild proteinuria with good kidney function, as well as polyneuropathy, marked eosinophilia (6,900), and positive p-ANCA 1/40 with a perinuclear pattern.

This case presented five clinical manifestations (Table 1) [5]: asthma, polyneuropathy, rhinopathy, marked eosinophilia (46%), and a histopathology of a cutaneous biopsy suggestive of vasculitis with the presence of eosinophils; additionally, there was positive p-ANCA with a perinuclear pattern, which occurs in 70% of patients and indicates disease activity.

The patient was treated with prednisone, pulses of dexamethasone, and cyclophosphamide, with complete regression of the cutaneous lesions and consequent improvement of the remaining complaints.

Discussion

CSS typically affects middle-aged patients and usually begins with respiratory asthmatiform manifestations or rhinitis, usually followed by manifestations of vasculitis, as in our case.

Cutaneous alterations occur in approximately 70% of the cases and manifest as hemorrhagic lesions: palpable purpura, petechiae, ecchymosis, and hemorrhagic bullae in 48%; subcutaneous nodules, frequently located on the scalp or distributed bilaterally over the extensor

surfaces of the extremities, in 30%; maculopapular eruption in 25%; and urticariform eruptions and livedo reticularis in 25% of cases [2, 4, 5].

Peripheral neuropathy is expressed as mononeuritis multiplex, occurring in 60% of patients [4]. Central nervous system signs such as headache, convulsion, hemiplegia, and brainstem signs are relatively rare [6].

Although in most cases this syndrome is idiopathic, inhaled antigens, vaccination, desensitization, infection, and suppression of oral corticoids have been considered as triggering factors [7].

The probable pathogenesis involves immediate hypersensitivity and cytotoxic reactions with activation of Th2 lymphocytes, followed by activation/degranulation of mastocytes and eosinophils, and activation of ANCA-dependent neutrophils [3].

It was suggested that IgE levels and eosinophilia play a central role in the syndrome's pathogenesis, being good parameters for follow-up of the disease's activity and response to therapy [1].

CSS typically presents in three distinct evolutionary phases: (1) allergic rhinitis, nasal polyposis, and asthma, persisting for years or decades; (2) eosinophilic pneumonia, gastroenteritis, and peripheral eosinophilia, with frequent recurrences; and (3) systemic vasculitis with granulomatous inflammation, occurring on average 3 years after the initial manifestations, although it is also possible for it to present up to 30 years later [4].

There are six diagnostic criteria for CSS: asthma, peripheral eosinophilia exceeding 10%, mono- or polyneuropathy, transitory pulmonary infiltration, abnormality of the paranasal sinuses, and histopathology revealing vessels with tissue eosinophilia. The presence of at least four of the six provides a diagnostic sensitivity of 85% and a specificity of 99.7% (Table 1) [5, 7].

The occurrence of p-ANCA ranges between 60 and 70%, and a perinuclear pattern is most frequently found. It is considered an indicator of disease activity, as well as peripheral eosinophilia [7].

Prednisone is the indicated treatment (60–100 mg/day) until no more disease activity is detected (eosinophilia, ESR, and p-ANCA), at which point a gradual reduction of the dosage is initiated. In cases of rapid worsening and involvement of multiple organs, pulse therapy with steroids and immunosuppression with cyclophosphamide should be used [2, 7].

Another possibility to be added to the arsenal in CCS is warfarin therapy. Najmi et al. [6] followed a patient for about 5 months, checking the INR at 2- to 3-week intervals; they found that there were no new attacks but reported that further studies are necessary to confirm the efficacy of anticoagulation in severe cases of CSS.

Conclusion

Our patient was diagnosed with a classic and exuberant clinical course of CSS, a rare disease. In her case, we found the following diagnostic criteria: (1) asthma, (2) polyneuropathy, (3) rhinopathy, (4) marked eosinophilia and a histopathology of vasculitis with eosinophils; and (5) positive p-ANCA with a perinuclear pattern. She exhibited complete regression of the cutaneous lesions and improvement of all the remaining manifestations with prednisone, pulses of dexamethasone, and cyclophosphamide.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors have no conflicts of interest to disclose.

References

1 Choi JH, Ahn IS, Lee HB, Park CW, Lee CH, Ahn HK. A case of Churg-Strauss syndrome. Ann Dermatol. 2009 May;21(2):213–6.
2 Marques CC, Fernandes EL, Miquelin GM, Colferai MM. Cutaneous manifestations of Churg-Strauss syndrome: key to diagnosis. An Bras Dermatol. 2017;92(5 Suppl 1):56–8.
3 Chung L, Kea B, Fiorentino DF. Cutaneous vasculitis. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. Dermatology. St. Louis: Elsevier; 2008. p. 347–67.
4 Brandt HR, Arnone M, Valente NY, Sotto MN, Criado PR. Vasculites dos médios e grandes vasos. An Bras Dermatol. 2009 Jan-Feb;84(1):55–67.
5 Masi AT, Hunder GG, Lie JT, Michel BA, Bloch DA, Arend WP et al. The American College of Rheumatology 1990 criteria for the classification of Churg-Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum. 1990 Aug;33(8):1094–100.
6 Najmi S, Ghareaghaji-Zare A, Ghazanfari-Amlashi S. Churg-Straus syndrome: A case report. Iran J Neurol. 2017 Jul;16(3):159–61.
7 Rochael MC, Caldas ML, Martinho DL. Vasculites. In: Ramos-e-Silva M, Castro MC, editors. Fundamentos de Dermatologia. Rio de Janeiro: Atheneu; 2010. p. 685–722.

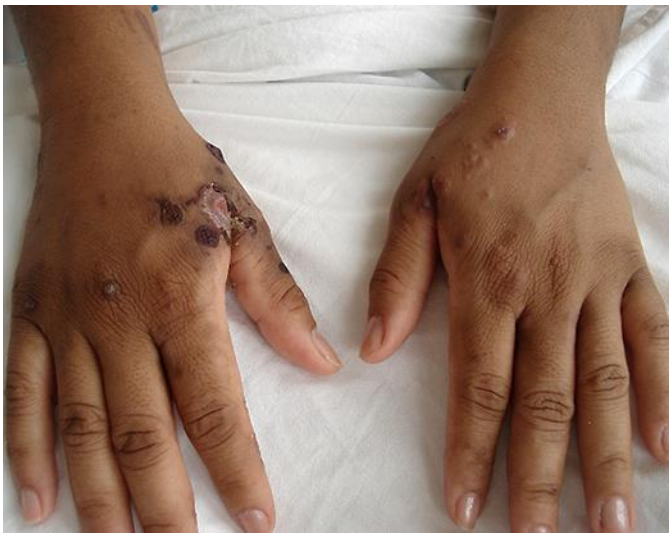

Fig. 1. Palpable purpura, bullae, and necrosis on the hands.

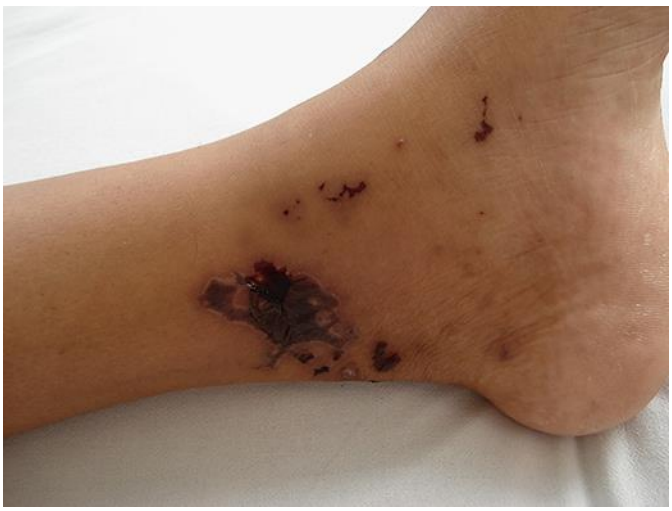

Fig. 2. Palpable purpura on the ankle that evolved into necrosis.

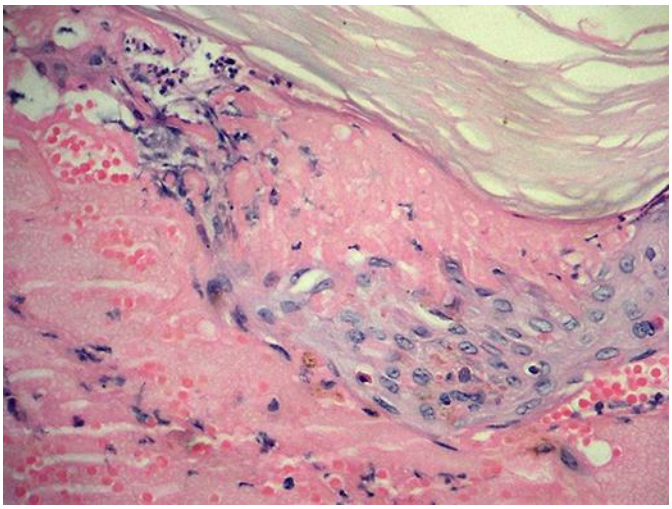

Fig. 3. Epidermal necrosis. H&E. ×100.

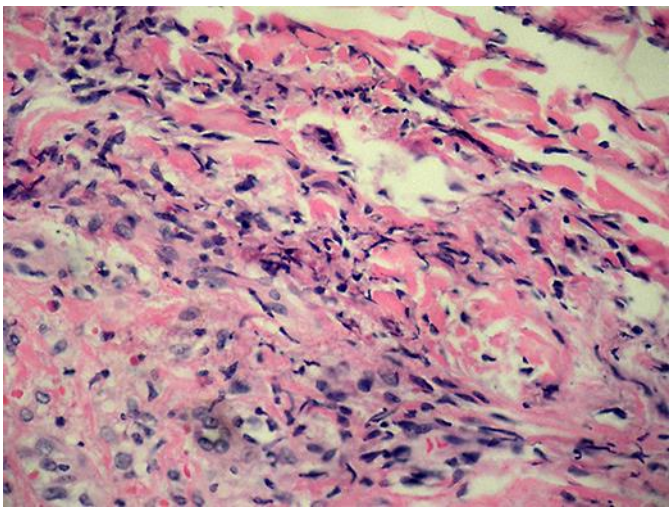

Fig. 4. Dense dermal infiltrate with eosinophils and red blood cell extravasation. H&E. ×100.

Table 1. Criteria for the classification of Churg-Strauss syndrome – American College of Rheumatology 1990 [5]

Asthma: history of wheezing or diffuse high-pitched expiratory rhonchi
Eosinophilia: eosinophilia >10% on differential white blood cell count
Mono- or polyneuropathy: development of mononeuropathy, multiple mononeuropathies, or polyneuropathy (glove/stocking distribution) attributable to systemic vasculitis
Pulmonary infiltrates, non-fixed: migratory or transitory pulmonary infiltrates (not including fixed infiltrates) attributable to vasculitis
Paranasal sinus abnormality: history of acute or chronic paranasal sinus pain or tenderness or radiographic opacification of the paranasal sinuses
Extravascular eosinophils: biopsy including artery, arteriole or venule showing accumulations of eosinophils in extravascular areas