

## CASE REPORT

# Spinal hematoma complicating a Churg-Strauss syndrome patient: a previously unreported association

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## INTRODUCTION

Churg-Strauss syndrome (CSS) is a systemic disease characterized by an eosinophil-rich and granulomatous inflammation involving the respiratory tract, necrotizing eosinophilic vasculitis affecting small- to medium-sized vessels, and an association with asthma and eosinophilia (1,2). Neurological involvement in CSS occurs in approximately 62% of patients, usually as peripheral neuropathy (3). Nevertheless, central nervous system (CNS) involvement is rare. Cerebral infarctions, subarachnoid hemorrhage, intracerebral hemorrhage and optic neuritis are reported events (4-8).

Spinal hematoma is a rare pathology [Kreppel et al. identified only 613 cases over 170 years of medical literature (9)]. The classic presentation is an acute onset of severe, radiating back pain followed by the signs and symptoms of spinal cord compression, which develop minutes to days later (9-11). The true etiology remains unknown, but associations with predisposing conditions, such as blood dyscrasias and arteriovenous malformation, have been reported (9,12). No previous spinal hemorrhage has been described in association with CSS.

In this case report, we describe a 40-year-old woman with CSS presenting with a spinal hematoma.

## CASE DESCRIPTION

A previously healthy 40-year-old woman was admitted to the general ward due to recent asthma symptoms, purpuric skin lesions, and intermittent paresthesia and paresis in both arms and legs, which were suggestive of multiplex mononeuritis. She also complained of diminished visual acuity. The relevant initial laboratory tests revealed a slightly increased creatinine value (1.55 mg/dl) and elevated leukocytes (24,520/mm<sup>3</sup>) due to eosinophilia (15,000/mm<sup>3</sup>). A urinary sediment evaluation demonstrated microscopic hematuria (>100 red cells per high-power field) and leukocyturia (30 leukocytes per high-power field). A coagulation evaluation revealed no remarkable contribution (platelets 459,000/mm<sup>3</sup>, prothrombin activity 75%, partial

thromboplastin time of 30 s). Perinuclear anti-neutrophil cytoplasmic antibodies and low titers of FAN-HEP2 were detected in her serum. A computed tomography (CT) scan exhibited bilateral maxillary, ethmoid and sphenoid sinus opacification and diffuse lung infiltrates. A skin biopsy demonstrated predominant extravascular eosinophils and necrotizing vasculitis (Figure 1). A diagnosis of Churg-Strauss syndrome was confirmed, and corticosteroid treatment was initiated.

The next morning, the patient awoke with urinary incontinence and an inability to move her legs. She maintained consciousness and had no pain or respiratory or cardiac distress. Her vital signs were normal. A neurological examination revealed complete paraplegia. Neurological and neurosurgical consultants were contacted, and emergency magnetic resonance imaging (MRI) was ordered. MRI of the thoracic spine revealed a hyperintense mass on the posterior topography, causing spinal cord compression from T5-T6 to T7-T8 (Figure 2). The compressed spinal cord already demonstrated signal alteration on T2-weighted images. Based on her physical examination and her image findings, a hematoma of the thoracic spine was suspected, and she was taken to the operating room for an emergency decompression laminectomy. During the operation, a subdural hematoma was discovered and removed. The pathologic report revealed a hematoma, but no blood vessels were present in the specimen to analyze whether vasculitis was present. After the surgery, she was admitted to the Intensive Care Unit (ICU). However, the patient was unable to move or feel her legs, and the urinary incontinence persisted. The patient continued on 1 g of methylprednisolone intravenously for three days.

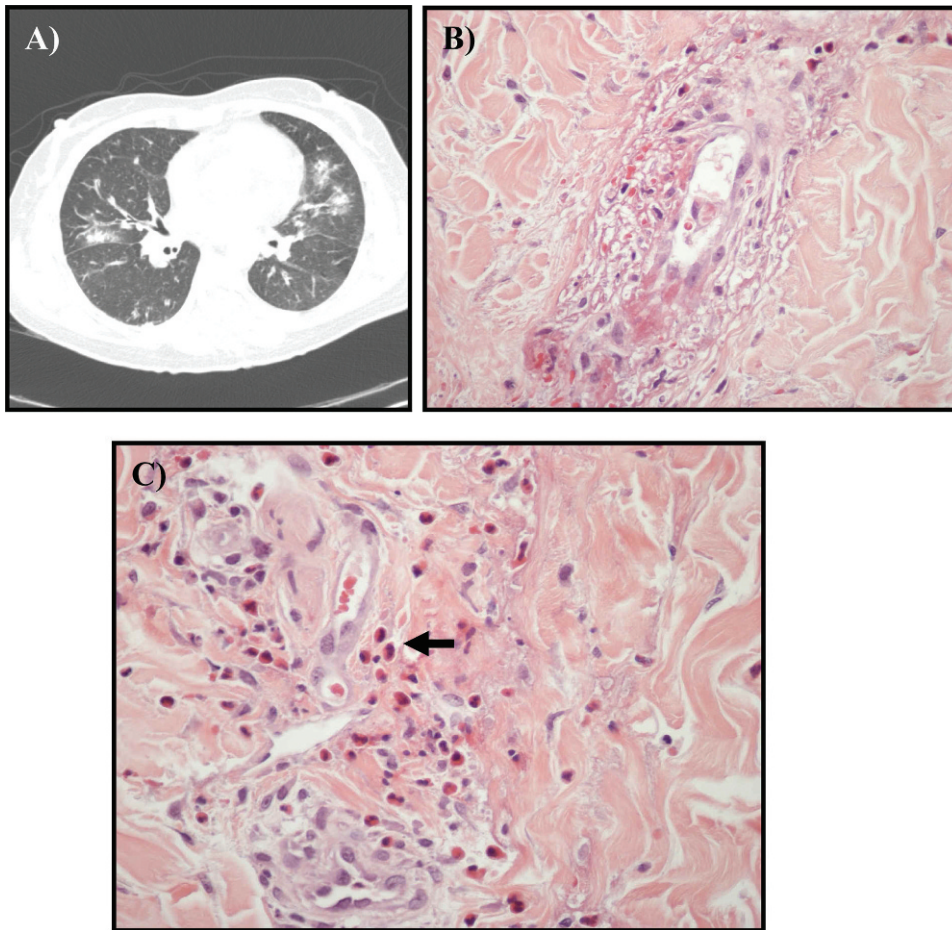
Her postoperative course was further complicated on the 6<sup>th</sup> day, when she developed surgical site infection and meningitis. She received appropriate antibiotic therapy. No microorganisms were identified. No new neurological symptoms appeared, and she was discharged without paraplegia improvement. After six months, she still had not recovered from the paraplegia or recovered from urinary retention. The asthma symptoms, eosinophilia, visual complaints, renal function (0.65 mg/dl) and paresthesia in her arms improved.

## DISCUSSION

The American College of Rheumatology has established six criteria for the classification of CSS in a patient with docu-

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No potential conflict of interest was reported.



**Figure 1** - Panel displaying the thoracic computed tomography findings and the skin histopathology. (A) The thoracic computed tomography scan demonstrates small nodular lesions with discrete ground-glass attenuation. (B) The skin biopsy (hematoxylin-eosin) demonstrates small-vessel vasculitis with eosinophils. (C) Magnification of the inflammatory infiltrate demonstrates the presence of numerous eosinophils (small arrow).

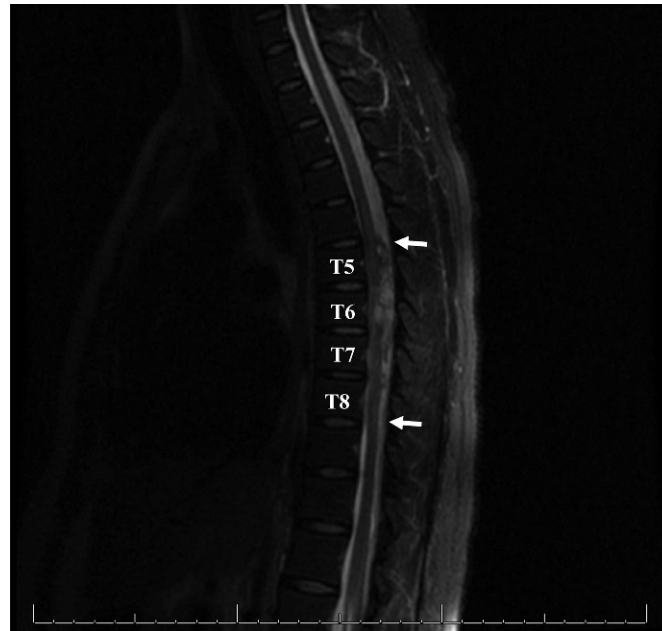
mented vasculitis: asthma, >10% eosinophilia in a differential white blood cell count, mononeuropathy (including multiplex) or polyneuropathy, non-fixed pulmonary infiltrates on radiography, paranasal sinus abnormality, and a biopsy showing blood vessels with extravascular eosinophils. The presence of four or more of these criteria reportedly yields a sensitivity of 85% and a specificity of 99.7% (13). Our patient could not be evaluated with a nerve conduction study to further characterize the symptoms clinically attributed to mononeuritis multiplex before the surgery and the corticosteroid treatment. Nevertheless, she displayed at least five criteria (excluding the neuropathy). The visual alterations might be related to optic neuritis, which improved after corticosteroid treatment, as did the other CSS-related symptoms. Unfortunately, she also could not undergo any ophthalmologic diagnostic study to conclude that she indeed had optic neuritis before treatment. The overall prognosis for CSS is good, and treatment with prednisone is usually successful (1).

Although neurological involvement in CSS is common, with peripheral neuropathy present in 53–78% of the patients (1,3), central nervous system involvement is uncommon (6–10%) and usually presents as cerebral infarction and rarely with subarachnoid hemorrhage, intracerebral hemorrhage, optic neuritis, convulsions, coma or psychosis (1,2). To the best of our knowledge, no previous case of CSS associated

with spinal hematoma has been documented. Only three cases of subdural hematoma with ANCA-associated vasculitis (Wegener's Granulomatosis) have been reported, and only one presented with a spinal subdural hematoma (14-16).

The majority of spinal hematomas (29.7%) have no etiological factor identified (9). After idiopathic spinal hematoma, cases related to anticoagulant therapy and vascular malformations represent the second and third most common categories (9). The present case had no history of trauma or other precipitating factors and no coagulation abnormality. Additionally, no vascular malformations were observed using MRI or CT. Unfortunately, the absence of vessels in the spinal specimen sent for pathologic evaluation precluded adequate study for evidence of vasculitis or eosinophilic infiltration. Nevertheless, because both CSS, characterized by an incidence of 0.11 to 2.66 new cases per million population per year, and spontaneous spinal hematoma are rare disorders [an estimated incidence of approximately one case per million patients per year (18)], we believe that a coincident association between these two conditions is unlikely.

In conclusion, we described a CSS case presenting with a novel neurological association, a spinal subdural hematoma, which, at the time of publication, had never been documented.



**Figure 2** - Magnetic resonance imaging of the thoracic spine. Sagittal T2-weighted imaging reveals a hyperintense posterior mass extending from T5-T6 to T7-T8 (white arrows), which compresses the spinal cord anteriorly.

## AUTHOR CONTRIBUTIONS

All of the authors participated in manuscript preparation, the literature search, data collection and data interpretation. All of the authors read and approved the final manuscript. This study was performed at the Emergency Department and Dermatology Department, Hospital das Clínicas, University of São Paulo Medical School, São Paulo, Brazil.

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