CARDIAC AND NEUROLOGICAL ISCHEMIA AS A PRESENTATION OF CHURG-STRAUSS VASCULITIS

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BACKGROUND
Churg Strauss angitis (CSA) is a granulomatous necrotizing vasculitis affecting small - to medium-sized vessels, usually associated with asthma and peripheral eosinophilia. CSA is a rare disorder with an incidence between 1.3 to 6.8 cases per million patients per year. We report a patient who presented with an acute coronary syndrome and soon afterwards developed ischemic neurological manifestations as an overt expression of CSA.

CASE REPORT
A 58-year-old Hispanic man with a recent diagnosis of bronchial asthma was admitted after presenting with intermittent chest pressure of four days duration. Physical examination was noncontributory. Laboratory data abnormalities included eosinophilia with an absolute eosinophil count of 2968/mm³. The patient’s electrocardiogram revealed ST segment depression in inferolateral leads. Cardiac markers were positive with a troponin peak of 4.00 ng/ml. Cardiac catheterization did not reveal any obstructive disease. His hospital course was complicated by right upper extremity monoparesis and right facial droop that developed on the second day of hospitalization. MRI brain revealed multiple areas of ischemia and infarction in watershed distribution bilaterally. A CT chest revealed multiple patchy areas of ground glass opacification. Vasculitic work up showed an elevated ESR and a positive perinuclear anti-neutrophil cytoplasmic antibody (P-ANCA). Complement levels (C3 and C4) were within normal limits. Computed tomography of the lungs showed patchy ground glass opacities. Based upon these findings, a diagnosis of Churg-Strauss syndrome was made. In addition to standard treatment for acute coronary syndrome and stroke, the patient was treated with high dose corticosteroids. Improvement in his clinical condition was noted soon after initiation of therapy and sustained a year later.

DISCUSSION
CSA is a rare disorder with a reported incidence of 1.3-6.8 cases per million patients per year. Classically, CSA develops as adult-onset asthma that progresses to hyper-eosinophilia, before resulting in overt vasculitic manifestations. Cardiac involvement is seen in 13 to 47% of CSA. Myocardial infarction in CSA is typically related to inflammation of small distal coronary vessels. CNS involvement is infrequent (9-14%). Corticosteroids are the cornerstone of initial treatment of CSA. Immunosuppressive treatment with cyclophosphamide is also frequently utilized, especially in patients with poor prognostic factors (elevated serum creatinine, proteinuria, gastrointestinal tract involvement, cardiomyopathy and central nervous system involvement) as well as those with treatment failure or relapse. Anti B cell strategies using agents like rituximab are emerging as a novel therapeutic modality in ANCA-associated vasculitis. Unfortunately, both cardiac and neurological involvements result in severe morbidity. Untreated, the disease has fatal consequences. Early diagnosis and aggressive therapy are therefore crucial to avoiding an unfavorable outcome.

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