## H79 Cardiac Manifestations of Churg-Strauss Syndrome: A Case Report and Literature Review

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After attending this presentation, attendees will be familiar with the characteristics of the rare disease eosinophilic granulomatosis with polyangiitis, formerly known as Churg-Strauss Syndrome, and the life-threatening cardiomyopathy associated with the syndrome.

This presentation will impact the forensic science community by emphasizing the differential diagnosis of eosinophilic syndromes and their life-threatening sequelae in young populations suspected of substance abuse.

A 22-year-old man with a history of long-standing asthma called 911 with complaints of chest pain and shortness of breath. He had presented to the emergency room on multiple occasions for the past two years, had been admitted eight times, and had undergone nerve biopsies for foot drop without diagnostic findings; he was believed to have mononeuritis multiplex. Earlier in the year, after an episode of eosinophilic pneumonia with leukocytosis, he was newly diagnosed with Crohn's disease. During his longest hospital admission in the last six months, Computed Tomography (CT) scan had diagnosed multiple hepatic abscesses, believed to be due to high-intensity steroid therapy. He was never out of hospital for more than a few days before returning with vague complaints of abdominal or chest pain that did not result in clear diagnoses. Medical records documented growing concern with his narcotics dependence and demands for stronger pain medications.

On the day of his death, the man appeared acutely ill at home. Police identified white powder at the scene. The man displayed combative behavior in the ambulance; with oxygen, he improved enough to transfer himself from the ambulance gurney to the emergency room bed. Within 15 minutes, he suddenly lost pulse and blood pressure and could not be resuscitated. Drug overdose was suspected, versus acute asthma exacerbation.

Autopsy showed "bread and butter" pericarditis and diffuse myocardial lesions without clear etiology that did not correspond to the coronary artery distribution. There were multiple recent wedge-shaped renal infarcts in the absence of any thromboembolic disease. Modest pulmonary hyperinflation was not suggestive of a fatal asthma attack; the lungs showed palpable granularity. The gastrointestinal tract was abnormal, but showed no gross evidence of Crohn's disease; there were no hepatic abscesses.

Histology identified Eosinophilic Granulomatosis with Polyangiitis (EGPA), formerly known as Churg-Strauss disease, in the heart, lungs, kidneys, gastrointestinal tract at multiple levels, and retroperitoneal vessels. There was extensive myocardial scarring, with both acute and resolving lesions. Histology confirmed no Crohn's disease. Toxicology was deemed non-contributory.

Cardiac involvement in EGPA includes carditis (myo-, endo-, and pericarditis), pericardial effusion, rhythm abnormalities, cardiac tamponade, and dilated cardiomyopathy, overall causing half of deaths attributable to EGPA. While only 25% have clinical cardiac symptoms, cardiac evaluations of patients with EGPA yield abnormal results in up to 62% of cases, highlighting the importance of early cardiac evaluation in patients with chronic asthma, gastrointestinal inflammation, and neuropathy.[1] The French Vasculitis Study Group has engendered a five-point system for assessing mortality risk, which includes reduced renal function, proteinuria, gastrointestinal hemorrhage, involvement of the central nervous system, and cardiomyopathy.[2,3] This patient fulfilled five of six diagnostic criteria, and three of five high-mortality criteria, with cardiomyopathy carrying the most significant prognostic weight.

Due to his narcotics dependence and steroid therapy masking his peripheral eosinophilia, his life-threatening EGPA cardiac involvement was not identified prior to autopsy. If cardiac imaging is initiated early in the disease process, and myocardial EGPA is treated aggressively, outcomes improve.[1] Autopsy findings such as those in this case can help guide future diagnoses.

**Reference(s):**

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2. Guillevin L., Lhote F., Gayraud M., Cohen P., Jarrousse B., Lortholary O., Thibult N., Casassus P. Prognostic factors in polyarteritis nodosa and Churg-Strauss syndrome. A prospective study in 342 patients. *Medicine (Baltimore)*. 75 (1996); 17-28.
3. Bourgarit A., Le Toumelin P., Pagnoux C., Cohen P., Mahr A., Le Guern V., Mouthon L., Guillevin L., French Vasculitis Study Group. Deaths occurring during the first year after treatment onset for polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome: A retrospective analysis of causes and factors predictive of mortality based on 595 patients. *Medicine (Baltimore)*. 84 (2005); 323-330.

**Churg-Strauss Syndrome, Refractory Asthma, Allergic Eosinophilia**



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