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*Casuistics*

# A case of Churg-Strauss syndrome

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**Summary.** A 54-year-old woman died after being battered, but the forensic autopsy revealed that her injuries were not serious or lethal. A detailed histological examination revealed that the basic disease had been Churg-Strauss syndrome. The differentiation of this syndrome of pulmonary infiltration and eosinophilia (PIE syndrome) from other allergic and noninfectious granulomatoses needs cautions consideration.

**Key words:** Pulmonary infiltration and eosinophilia – Churg-Strauss syndrome

**Zusammenfassung.** Eine 45jährige Frau verstarb nach stumpfer Gewalt durch Schläge. Die gerichtliche Obduktion ergab jedoch, daß ihre Verletzungen nicht schwer oder tödlich waren. Durch ausführliche histologische Untersuchungen wurde als Grundkrankheit ein Churg-Strauss-Syndrom aufgedeckt. Dieses Syndrom der pulmonalen Infiltration und Eosinophilie (PIE-Syndrom) kann nur nach gründlicher Überlegung von anderen allergischen und nicht-infektiösen Granulomatosen abgegrenzt werden.

**Schlüsselwörter:** Pulmonale Infiltration und Eosinophilie – Churg-Strauss-Syndrome

## Introduction

The syndrome of pulmonary infiltration and eosinophilia (PIE syndrome) occurs only occasionally in the practice of forensic pathology. The PIE syndrome has numerous causes and is often a component of some illnesses, such as allergic bronchopulmonary aspergillosis, chronic eosinophilic pneumonia, drug reaction, and helminth infestation. Bacterial and fungal infections, sarcoidosis, and tumors are also infrequently associated with PIE syndrome. However, some forms of this syndrome seem to occur as a separate entity. One of these is the Churg-Strauss syndrome, a combination of vasculitis and respiratory tract

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granulomas. Bronchial asthma is usually present for several years before the full syndrome develops, which resembles polyarteritis nodosa and is accompanied by eosiniphilia. The main histological feature of this syndrome, the disseminated necrotizing vasculitis, was first described i 1951 by Churg and Strauss [2]. The vasculitis involving small arteries and veins may be present in any organ [1–4, 6, 9], and is manifested as fibrinoid necrosis, and an eosinophil-rich inflammatory infiltration, but around the larger muscular arteries the inflammation may be granulomatous. The early phase is characterized by eosinophil angiitis, followed by fibrinoid necrosis. In the next phase necrotizing granulomas with histiocytes and giant cells appear. In the last phase fibrosis and cicatrices are present. The Churg-Strauss syndrome has a high mortality of approximately 50% and a short survival period of 4–5 years. The cause of death depends on the location and may be bronchopneumonia, cardiac failure, renal failure, peritonitis, etc. Adequate therapy has not yet been found but corticosteroid treatment could be beneficial. The differential diagnosis of the Churg-Strauss syndrome is difficult. Besides the above mentioned diseases periarteritis nodosa, bronchial asthma, Löffler pneumonia, Wegener granulomatosis, eosinophilic granuloma, eosinophilic leukemia, and Goodpasture syndrome must also be considered [5–8, 10, 11]. The anamnesis together with the necropsy and the systemic histological examination is necessary to make a correct diagnosis.

## Case report

A 54-year-old woman was found dead at the bottom of the ladder to the loft in the yard of her house with numerous wounds on her body. The case was reported as battery by her husband, but the husband said she had fallen from the loft. Later on, he admitted only a mild battery with a cudgel. They had lived in very poor conditions and were frequently intoxicated. The woman had no medical records on her anamnesis so that a forensic autopsy was requested to establish the cause of death.

## Necropsy findings

There were numerous wounds on different parts of the body. These were all superficial abrasions and bruises in various shapes and sizes. There were no bone fractures or visceral injuries. At the necropsy the lungs showed the most apparent alterations. Dark grayish-red nodules varying in size from a miliary tubercle to leisons 1–2 cm in diameter were scattered through all the lobes of lungs. The picture resembled hemorrhagic infarction or blood aspiration, but pulmonary arteries were passable, and in the upper respiratory tract no traces of blood stains caused by basic fracture or nosebleed were found. The heart ventricles were found to be dilated and there was a mild pulmonary and a severe brain edema with impaction of the cerebellar tonsils. There were some small hemorrhages under the visceral layer of the pericardium and in the subcutis in locations other than those of the abrasions and bruises. The viscera did not show characteristic lesions. The cause of death was failure of the right ventricle of the heart.

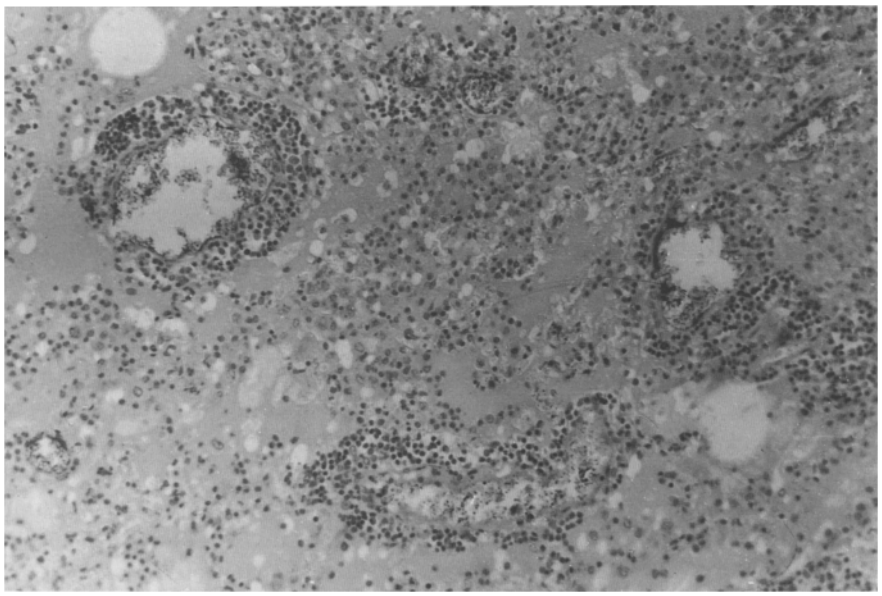

**Fig. 1.** Eosinophil-rich inflammatory infiltrate around the blood vessels of the lung

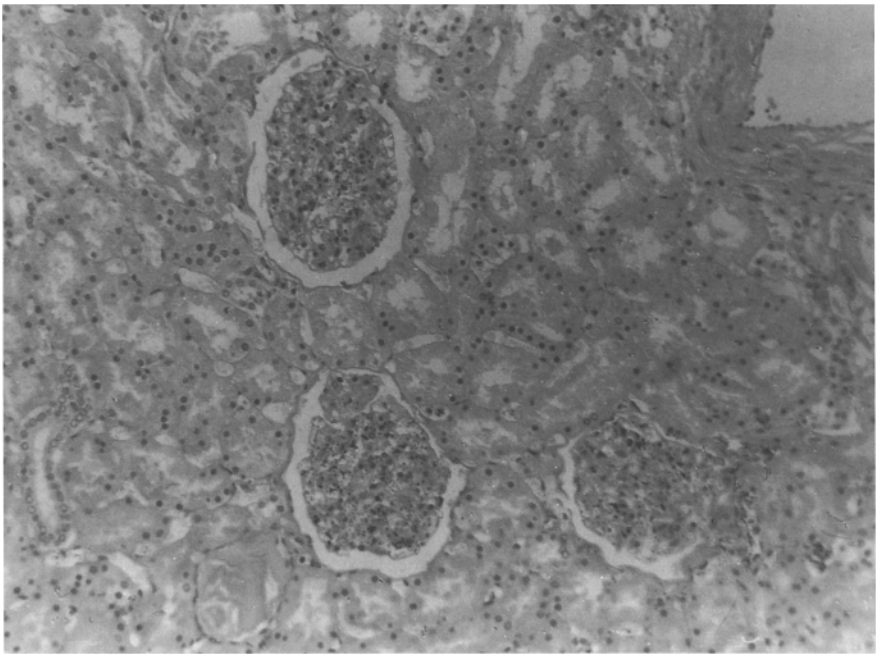

**Fig. 2.** Eosinophil-rich inflammatory infiltrate in glomeruli

## Histological findings

The microscopic examination showed vasculitis involving small arteries and veins in the lungs. The inflammatory infiltration was eosinophil-rich (Fig. 1). Fully developed granulomas were not present in the lungs. In the subcutis and the subpericardium there were similar but less frequent eosinophil infiltrations and small hemorrhages. Focal and segmental lesions of glomeruli caused by eosinophil infiltration in the kidneys (Fig. 2) were also found. In the liver small granuloma-like nodules were to be found around portal veins.

## Discussion

The Churg-Strauss syndrome is a manifestation of the syndrome of pulmonary infiltration and eosinophilia (PIE syndrome). It is of an uncertain origin but is often preceded by bronchial asthma or allergic diseases. The diagnosis of Churg-Strauss syndrome can only be established by detailed autopsy and histological examination. Depending on the phase of this disease, eosinophil infiltration or granulomas can be found in numerous organs. The cause of death is generally the failure of the most deteriorated organ. Comparison of the type and extent of organ lesions and the histological features helps to differentiate the Churg-Strauss syndrome from other allergic and non-infectious granulomatoses. The forensic autopsy of the body of a woman who died after being battered revealed that there was no connection between the battery and death. The cause of death was of natural origin. The histological examination revealed the basic disease to be an early-stage Churg-Strauss syndrome. In similar cases only a thorough autopsy, histological examination, and an extensive knowledge of the pathology of rare diseases can clarify the cause of death.

## References

1. Chumbley LC, Harrison EG, DeRemee RA (1977) Allergic granulomatosis and angiitis (Churg-Strauss Syndrome): Report and analysis of 30 cases. Mayo Clin Proc 52:477–487
2. Churg J, Strauss L (1951) Allergic granulomatosis, allergic angiitis, and periarteritis nodosa. Am J Pathol 27:277–289
3. DeRemee RA (1980) Respiratory vasulitis. Mayo Clin Proc 55:492–499
4. Fauci AS, Haynes BF, Katz P (1978) The spectrum of vasculitis: Clinical pathologic, immunologic, and therapeutic considerations. Ann Intern Med 89:660–669
5. Friedman PJ, Liebow AA, Sokoloff J (1981) Eosinophil granuloma of the lung. Medicine 60:385–392
6. Kissane JM (eds) (1985) Anderson's Pathology. Mosby, St. Louis Toronto Princeton, pp 886–887
7. Koss MN (1981) Allergic granulomatosis (Churg-Strauss Syndrome): Pulmonary and renal morphologic findings. Am J Surg Pathol 5:21–27
8. Liebow AA (1973) Pulmonary angiitis and granulomatosis. Am Rev Respir Dis 108:1–12
9. Sale S, Patterson R (1981) Recurrent Churg-Strauss vasculitis. Arch Int Med 141:1263–1266
10. Spencer H (1977) Pathology of the lung. Pergamon Press, Oxford New York Toronto Sydney Paris Frankfurt, pp 706–724
11. Wilson KS, Alexander HL (1945) The relation of periarteritis nodosa to bronchial asthma and other forms of human hypersensitiveness. J Lab Clin Med 30:195–203

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