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CASE STUDY

Cocaine-induced Churg-Strauss vasculitis

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Cocaine-induced Churg-Strauss vasculitis. R. Orriols, X. Muñoz, J. Ferrer, P. Huget, F. Morell. ©ERS Journals Ltd 1996.

ABSTRACT: A freebase cocaine-smoking woman developed relapsing fever, bronchoconstriction, arthralgias and weight loss. Pulmonary infiltrates, arthritis, microhaematuria, pruriginous skin rash and mononeuritis multiplex were later added to the clinical picture. Both skin and muscle biopsies showed eosinophilic angiitis. Improvement or worsening of her clinical picture repeatedly coincided with avoidance or use of smoked cocaine, respectively.

We suggest that Churg-Strauss vasculitis may be a complication of smoking freebase cocaine.

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Various lung diseases related to the use of freebase cocaine have been reported [1, 2]. Pulmonary infiltration with eosinophilia is an uncommon presentation of cocaine smoking [3–6]. We report a case of Churg-Strauss vasculitis in a patient whose clinical symptoms were clearly related to cocaine smoking. To our knowledge, this effect of cocaine abuse has not been described previously.

Case report

A 39 year old woman had been a 40 cigarette·day^{-1} smoker since the age of 16 yrs. She had been using cocaine for the last 14 yrs and denied other toxic exposures and *i.v.* drug abuse. Her initial cocaine use had been limited to intranasal administration. Since then, she had occasionally suffered from wheezing which remitted with the inhalation of adrenergic β_2-agonists. She had begun to use freebase "smoked" cocaine 8 months earlier. Shortly afterwards, she presented with dyspnoea on effort, wheezing, 20 kg weight loss and polyarthralgias. The patient was admitted because of sweats, 38°C fever, dry cough and right pleuritic pain of 1 week's duration.

Physical examination showed right inspiratory crackles and diffuse expiratory wheezes. Nasal examination was normal. Chest radiography disclosed an alveolar infiltrate in the right lower lobe. Electrocardiography (ECG) revealed tachycardia at 110 beats·min^{-1} with diffuse repolarization changes. Pulmonary function testing showed a forced vital capacity (FVC) of 2.6 L (79% of predicted) forced expiratory volume in one second (FEV_1) 2.0 L (70% pred) and FEV_1/FVC 77%. Arterial blood gas measurements performed with the patient breathing room air were: pH 7.42; arterial carbon dioxide tension (P_{a,CO_2}) 5.6 kPa (42 mmHg) and arterial oxygen tension (P_{a,O_2}) 6.9 kPa (52 mmHg). Leucocyte count was 11.9×10^9 cells·L^{-1} with 3% eosinophils, and serum immunoglobulin E (IgE) was 346 IU·mL^{-1}. Erythrocyte sedimentation rate (ESR) was 99 mm·h^{-1}. All bacteriological studies were negative. Testing for human immunodeficiency virus was also negative.

The patient stopped smoking cocaine and was treated with bronchodilators and antibiotics. Marked clinical and radiological improvement was observed within a few days and the patient was discharged. Temporal relationship between cocaine exposure and laboratory data is shown in figure 1.

Six months later, the patient began to smoke cocaine again and presented with three episodes of fever, cough, wheezing, arthralgias and peripheral lung infiltrates (fig. 2),

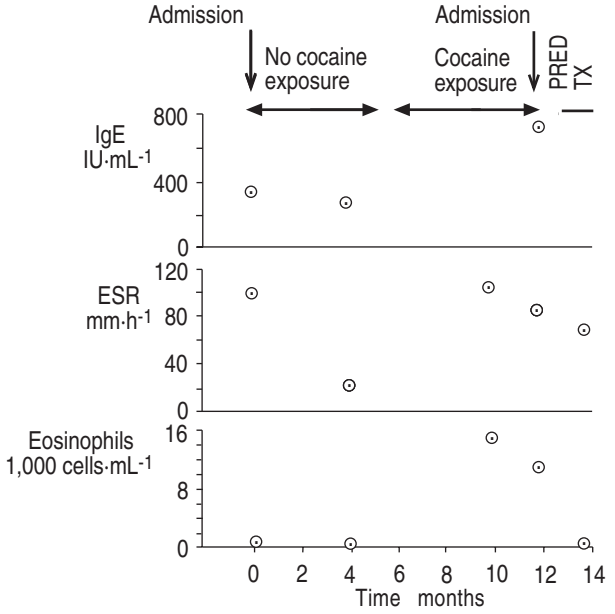

Fig. 1. – Temporal relationship between cocaine exposure and laboratory data. PRED TX: prednisone treatment; IgE: immunoglobulin E; ESR: erythrocyte sedimentation rate.

one of which showed cavitation (fig. 3). These episodes disappeared or improved when exposure to cocaine was stopped. The patient was treated on each occasion with β_2-agonists, budesonide and antibiotics, but never with oral corticosteroids.

The patient was readmitted 12 months after the first discharge because of fever, dyspnoea with wheezes, skin rash with scabs on all extremities, arthritis, paraesthesia and hypoalsthesia in the external side of the right lower limb. Chest radiography showed patchy right pulmonary infiltrates. ECG again revealed tachycardia at 120 beats·min^{-1} with diffuse repolarization changes.

Pulmonary function testing showed a FVC of 2.38 L (72% pred), FEV1 2.0 L (70% pred), FEV1/FVC 83%, residual volume (RV) 2.46 L (157% pred), total lung capacity (TLC) 4.75 L (96% pred), single breath transfer factor for carbon monoxide ($T_{L,CO}$) 70% pred and transfer factor/alveolar volume ($T_{L,CO}/V_A$) 107%. Bronchodilator test was positive with a FVC and FEV1 improvement of 13 and 16%, respectively. Arterial blood gas measurements performed with the patient breathing room air were: pH 7.41; P_{a,CO_2} 5.6 kPa (42 mmHg) and P_{a,O_2} 6.1 kPa (46 mmHg). Leucocyte count was 21.9×10^9 cells·L^{-1} with 48% eosinophils, serum IgE was 730 IU·mL^{-1} and ESR 82 mm·h^{-1}. Urine sediment showed microhaematuria of 25 erythrocytes·field^{-1}. Negative investigations at that time included antinuclear antibody (ANA), rheumatoid factor, antinuclear cytoplasmic antibody (ANCA), hepatitis B surface antigen, aspergillus precipitins, syphilis serology, human immunodeficiency virus serology, parasite study and blood, sputum, stool and urine cultures.

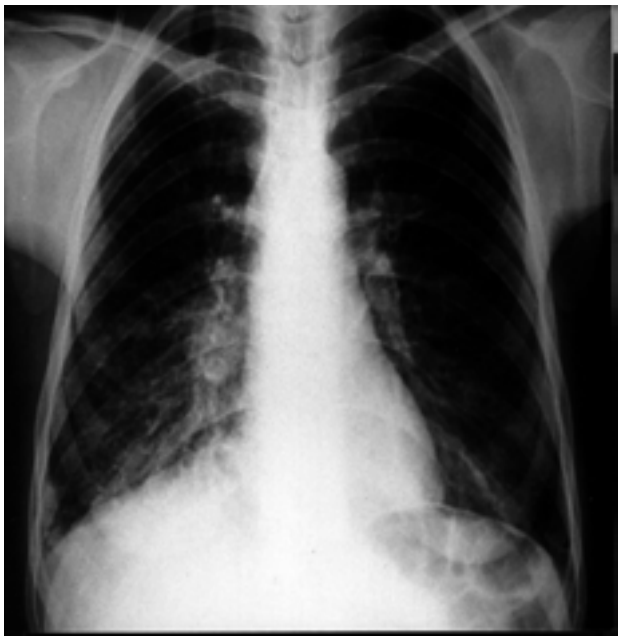

Fig. 2. – Chest radiograph showing bilateral pulmonary infiltrates.

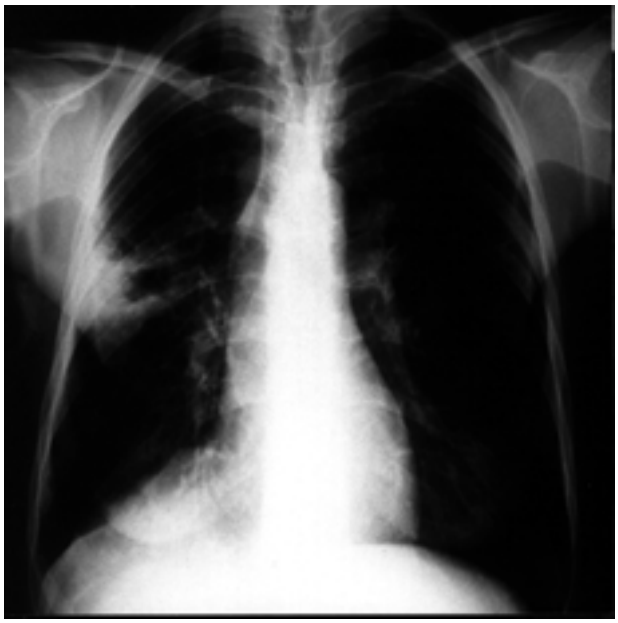

Fig. 3. – Chest radiograph showing pulmonary infiltrate with cavitation.

Neurophysiological studies showed mononeuritis multiplex with diminished amplitude of motor and sensory potentials of sural and right common peroneal nerves. Severe denervation of the extensor digitorum brevis muscle was also observed. Skin and muscle biopsies showed eosinophilic infiltrate of moderate intensity in small arterioles and venules with fibrinoid necrosis (fig. 4). Muscle fibres presented degenerative changes, basophilia and myophagia in areas of vascular involvement.

The patient stopped smoking cocaine and treatment with prednisone 1 mg·kg^{-1}·day^{-1} was started, with rapid improvement in symptoms and normalization of the radiographic changes. The pulmonary function testing showed a FVC of 3.03 L (92% pred), FEV1 2.64 L (92% pred), FEV1/FVC 87%, $T_{L,CO}$ 106% and $T_{L,CO}/V_A$ 131% pred. Arterial blood gas measurements performed with the patient breathing room air were: pH 7.38; P_{a,CO_2} 5.5 kPa (41 mmHg) and P_{a,O_2} 12 kPa (90 mmHg). While

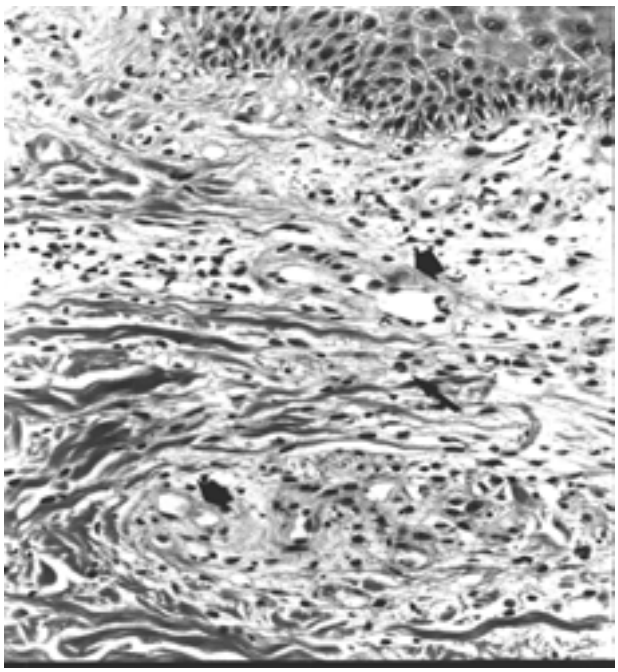

Fig. 4. – Inflammatory infiltrate with eosinophils in the dermis (thin arrow). Vessels with focal fibrinoid necrosis (thick arrow). (Haematoxylin and eosin stain; original magnification x250).

under treatment, the patient resumed her cocaine smoking habit for two seperate periods of time. Once again, her fever, arthralgia, arthritis, skin rash and mononeuritis multiplex improved or worsened with avoidance or use of cocaine, respectively.

Discussion

Our patient developed features typical of Churg-Strauss vasculitis, including late-onset asthma, heavy blood eosinophilia, pulmonary infiltrates, mononeuritis multiplex and raised serum IgE. Skin and muscle eosinophilic infiltration and necrotizing angiitis were demonstrated. Granulomas were not found. However, the three histological components of the disorder often do not coexist temporally or spatially, and are found together only in a minority of cases [7]. The relationship to cocaine use is supported by the reappearance of the syndrome during periods of cocaine smoking. Thus, it seems evident that Churg-Strauss vasculitis might have been induced by a hypersensitivity reaction to smoked freebase cocaine.

Aetiological or precipitating factors of Churg-Strauss vasculitis are not established in the majority of cases. However, allergic desensitization in 25 reported cases [8, 9], and biliary tract infection by ascaris in one patient [10], have been implicated as precipitating factors of Churg-Strauss vasculitis. Inhaled antigens have also been suggested as possible causes. GUILLEVIN *et al*. [11] described a patient who developed Churg-Strauss syndrome after exposure to pigeons.

Some reports suggest that cocaine could cause vasculitis. BACHARACH *et al*. [12] reported a case of thoracoabdominal aortitis with aneurysm formation, consistent with possible cocaine-induced vasculitis. In another report, KAYE and FAINSTAT [13] suggested that cocaine might cause cerebral vasculitis. Whilst histological confirmation was not established in this case, the angiographic findings were consistent with vasculitis and the patient responded to steroid therapy. KRENDEL *et al*. [14] reported the cases of two cocaine users who developed transient blindness, persistent headache and progressive widespread cerebral dysfunction while smoking freebase cocaine. Brain biopsy showed vasculitis involving small vessels in both patients.

The mechanism by which cocaine may interact with the vascular epithelium and provoke vasculitis remains unknown. As in our case, IgE production, eosinophilic chemotaxis and subsequent release of mediators with vascular injury may be induced by inhaled cocaine.

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References

1. Ettinger NA, Albin RJ. A review of the respiratory effects of smoking cocaine. *Am J Med* 1989; 87: 664–668.
2. Warner EA. Cocaine abuse. *Ann Intern Med* 1993; 119: 226–235.
3. Oh PI, Balter MS. Cocaine-induced eosinophilic lung disease. *Thorax* 1992; 47: 478–479.
4. Murray RJ, Albin RJ, Mergner W, Criner GJ. Diffuse alveolar hemorrhage temporally related to cocaine smoking. *Chest* 1988; 93: 427–429.
5. Kissner DG, Lawrence WD, Selis JE, Flint A. Crack lung: pulmonary disease caused by cocaine abuse. *Am Rev Respir Dis* 1987; 136: 1250–1252.
6. Nadeem S, Nasir N, Israel R. Löffler's syndrome secondary to crack cocaine. *Chest* 1994; 105: 1599–1600.
7. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: a clinical approach to the Churg-Strauss syndrome. *Medicine* 1984; 163: 65–81.
8. Phanuphak P, Khocer PF. Onset of polyarteritis nodosa during allergic hyposensitization treatment. *Am J Med* 1980; 68: 479–485.
9. Guillevin L, Guittard TH, Bletry O, Godeau P, Rosenthal P. Systemic necrotizing angiitis with asthma: causes and precipitating factors in 43 cases. *Lung* 1987; 165: 165–172.
10. Chauhan A, Scott DGI, Neuberger J, Gaston JSH, Bacon PA. Churg-Strauss vasculitis and ascaris infection. *Ann Rheum Dis* 1990; 49: 320–322.
11. Guillevin L, Amouroux J, Arbeille B, Boura R. Churg-Strauss angiitis: arguments favouring the responsibility of inhaled antigens. *Chest* 1991; 100: 1472–1473.
12. Bacharach JM, Colville DS, Lie JT. Accelerated atherosclerosis, aneurysmal disease and aortitis: possible pathogenetic association with cocaine abuse. *Int Angiol* 1992; 11: 83–86.
13. Kaye BR, Fainstat M. Cerebral vasculitis associated with cocaine abuse. *J Am Med Assoc* 1987; 258: 2104–2106.
14. Krendel DA, Ditter SM, Frankel MR, Ross WK. Biopsy proven cerebral vasculitis associated with cocaine abuse. *Neurology* 1990; 40: 1092–1094.