

CASE REPORT

Montelukast-associated Churg–Strauss syndrome

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Abstract

A 68-year-old asthmatic presented markedly unwell with arthralgia, mononeuritis multiplex, peripheral neuropathy, and eosinophilia. His past medical history included perennial rhinitis, and nasal polyps. Three months prior to admission his prednisolone was stopped and Montelukast was started. The diagnosis of Montelukast-associated Churg–Strauss syndrome was made. The drug was stopped and steroids started with general improvement and reduction of eosinophilia; however, the neurological deficit persisted.

Keywords: *Churg–Strauss syndrome, Montelukast, leukotriene receptor antagonists*

Case report

A 68-year-old male, presented with bilateral shoulder and elbow pain for 6 days, and numbness of the right lateral three fingers, followed by numbness of the tips of the left medial three fingers. He had no cardiac, gastrointestinal, or urinary symptoms apart from frequency and urgency.

His past medical history included asthma, perennial rhinitis and nasal polyps, hypertension, and minor stroke several years previously with no residual deficit. His medication included beclomethasone, ipratropium, and salbutamol inhalers. Three months prior to admission prednisolone was stopped and he was started on Montelukast (leukotriene receptor antagonist). He was also on aspirin and nifedipine.

He had been exposed to asbestos, was a non-smoker, and did not drink alcohol.

On examination he was conscious and orientated. Cardiovascular, respiratory, and abdominal examination revealed no abnormality apart from sinus tachycardia at 115 beats per minute and slight pyrexia at 37.5 °C. There was no lymphadenopathy, and there was no evidence of arthritis. On neurological examination, tone and power testing were normal. The reflexes were normal apart from the right brachioradialis and the left ankle, which were diminished. The plantar reflexes were downgoing. There was diminished pain and touch sensation in the distribution of the ulnar nerve in the left hand, and in the tips of the lateral three fingers of the right hand. There were also diminished pain and touch sensations in the distal legs and feet. Co-ordination and gait were normal.

Full blood count showed WBC 27.3×10^9/l (neutrophils were 10.9, eosinophils 14.2, and lymphocytes 1.1), Hb 150 g/l, and platelets 373×10^9. WBC increased 5 days later to 46.7×10^9 (32.6 were eosinophils). The muscle enzymes were slightly elevated: CK 592 u (N < 175), AST 34 u (N < 45), LD 711 u (N < 465). CRP was 68.7 mg/dl. Kidney and liver functions were repeatedly normal. Urine and stool analyses were normal. Rheumatoid arthritis screen, ANA, pANCA and cANCA were negative. CXR showed calcified pleural plaques affecting the right hemithorax.

He was diagnosed as Montelukast-associated Churg–Strauss syndrome. Montelukast was stopped and prednisolone started with general improvement and reduction of eosinophil count, however the neurological deficit remained.

The patient was followed up for 2 years, during which the neurological deficit did not improve till he died from myocardial infarction complicated by a cerebrovascular accident.

Discussion

Churg–Strauss syndrome was first described by Churg and Strauss in 1951. It is a necrotizing vasculitis affecting arterioles, venules and medium sized muscular arteries.

The American College of Rheumatology, in 1990, developed criteria for classification of Churg–Strauss syndrome. These are: asthma, eosinophilia >10% on differential WBC, mononeuropathy (including mononeuropathy multiplex) or polyneuropathy, non-fixed pulmonary infiltrate on CXR, paranasal sinus abnormality, and a biopsy containing a blood vessel with extravascular eosinophils. The presence of four or more of these six criteria yielded a sensitivity of 85% and a specificity of 99.7%. Our patient fulfilled four of these criteria.

Several cases of Churg–Strauss syndrome associated with Montelukast use have been reported in the UK [1], and with Zafirlukast in the US [2]. A suggested explanation for this is the unmasking of previously unsuspected Churg–Strauss syndrome by oral steroid reduction. In the cases reported, like in our case, Churg–Strauss syndrome became apparent within days to months of stopping oral prednisolone [3]. However, leukotriene modifier associated Churg–Strauss syndrome was also reported in steroid naïve asthmatics [4] and in a patient with no recent use of oral steroids [5]. This might indicate an allergic response to the drug [6].

Asthma may be the presenting manifestation of Churg–Strauss syndrome. Churg–Strauss syndrome could be fatal, and early recognition and treatment are essential and could be life saving. Doctors should monitor asthmatic patients who are started on leukotriene modifiers. The development of systemic symptoms should prompt further investigations and Churg–Strauss syndrome has to be considered.

Key points

- Diagnosis of Churg–Strauss syndrome is considered when a patient has four of the six American College of Rheumatology criteria.
- Churg–Strauss syndrome rarely develops in patients with moderate to severe asthma when started on leukotriene modifiers.
- A suggested explanation: the withdrawal of steroids afforded by leukotriene modifiers may lead to unmasking of a vasculitic process previously recognised as asthma. Allergic drug reaction is an alternative explanation.
- Churg–Strauss syndrome is rare; however, it could be fatal. Doctors need to be vigilant when starting anti-leukotrienes for asthma patients.

References

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