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SUCCESSIVE TREATMENT OF CHURG-STRAUSS SYNDROME WITH CYCLESONIDE AFTER ORAL STEROIDS DISCONTINUATION DUE TO SIDE EFFECTS

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Churg-Strauss syndrome, ANCA-associated is characterized by history of asthma, peripheral blood and tissue eosinophilia with particular predilection to lungs in context of multisystem disorder manifested by skin involvement, mesenteric ischemia, peripheral neuropathy, and myocarditis. We would like to present a case of 35 year-old patient with severe asthma, referred to our hospital with symptoms of the status astmaticus. On admission, the patient was in severe state, physical examination suggested bilateral pneumonia, confirmed by chest X-ray, blood gas analyze showed features of a respiratory failure. Patient was successfully treated with epidephrine, predonisone, three antibiotics, and oxygen. On subsequent hospitalization one month later, also due to asthma exacerbation he presented with fever, cough, dyspnea, mialgia, and symptoms of sinusitis. Chest X-ray revealed profuse pulmonary infiltrates, peripheral blood smear showed eosinophilia 50%. The history of severe asthma and peripheral blood eosinophilia made us suspect Churg-Strauss syndrome. Further investigations revealed negative test for both antineutrophil cytoplasmatic anybodies in a perinuclear distribution and antibodies against myeoperoxidase, granulomas in the bronchi, which could correspond to Wegener's granulomatosis. The therapy with oral steroids was started with resolution of symptoms. After 10 months of oral steroids use, the patient again developed dyspnea, fever, marked obturation, hypoxemia, disseminated patchy infiltrates seen on chest X-ray and sinusitis with nasal polyps. Ultimately the diagnosis of Churg-Strauss syndrome was established on clinical basis and cyclophosphamide was added to the corticosteroids. Clinical improvement in the patient's general health and normalization of laboratory tests were achieved again. The patient was under surveillance for 2 years and over this time symptoms and lesions to the lungs did not recur. Cyclophosphamide was discontinued after 12 months of use. The patients decided to withdraw oral steroids because of whole range of adverse symptoms: significant weight gain, striae of the skin on abdomen, and aseptic necrosis of both femoral heads and diabetes mellitus. To prevent disease recurrence the new regimen with ciclesonide 2 x 320 μg was introduced. Today the patient is in good health with no symptoms, the laboratory findings show eosinophilia of 6%, which is the lowest in his disease course. The usage of cyclesonide is not recommended for Churg- Strauss syndrome, but in the case of the patient with mainly pulmonary manifestations who discontinued oral steroids due to side effects proved successful. Further investigations are needed to find out whether cyclesonide my replace oral steroids in long term control of Churg- Strauss syndrome with mainly pulmonary manifestations.