Churg-Strauss syndrome in a pediatric patient

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Background

The term 'vasculitis' denotes a pathological proces of inflammation, vessel wall destruction and tissue necrosis. Vasculitis can occur secondary to drugs, infections and/or other diseases like rheumatoid arthritis, and may also occur without known underling cause ('primary vasculitis').
Churg-Strauss syndrome (CSS), also called allergic granulomatosis and angiitis, is an uncommon multisystem disorder characterized by this vasculitis and is associated with asthma and eosinophilia. CSS in childhood is rare and the clinical presentation can be quite diverse.

Methods

We report on a 12-year-old boy with asthma and deterioration of general condition, who was eventually diagnosed with an ANCA-negative Churg-Strauss syndrome. Patient characteristics and clinical characteristics are summarized.

Results: medical history

Eleven months before clinical deterioration, the child presented with his first acute asthmatic exacerbation, which was treated with frequent nebulisation of ipratropium and salbutamol, and temporarily extra oxygen administration.
Laboratory and pulmonary investigations showed total IgE 228 kU/l. Allergic tests were positive for grass and tree pollen. Spirometry demonstrated a reversible airway obstruction, steroid naive fractional exhaled nitric oxide (FeNO) was increased (38.5 ppb), and decreased under maintenance therapy with salmeterol/fluticasone (5.6 ppb).

Results: presentation at emergency department

During summer holidays abroad, his general condition deteriorated progressively. Additional examinations revealed leukocytosis (26.5x109/L) with nearly 50% eosinophils, whereas total IgE was increased to 2901 kU/L. One day later, he presented himself at the emergency department and was hospitalized for further investigations and therapeutical intervention.

Physical examination revealed cachexia, several skin lesions on the elbows, back and feet (fig. 1a), and two palpable subcutaneous noduli on the back (fig. 1b). Breathing frequency was 23 breaths per minute, transcutaneous oxygen saturation was more than 97%. Auscultation of lungs, heart and abdomen were normal. There was no hepatosplenomegaly. Neurological examination was normal.

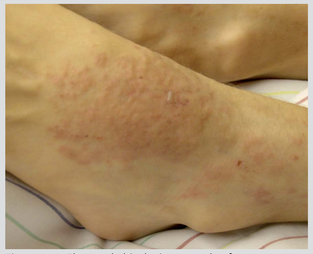

Figure 1a: Elevated skin lesions on the feet, compatible with vasculitis

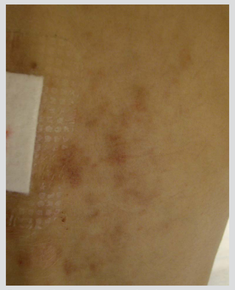

Figure 1b: Purpura and subcutaneous noduli on the back

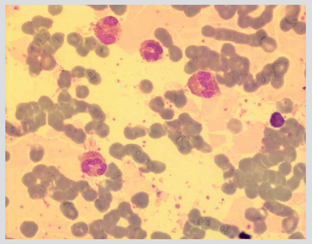

Figure 2: Peripheral blood eosinophilia with 4 eosinophils and 1 basophil

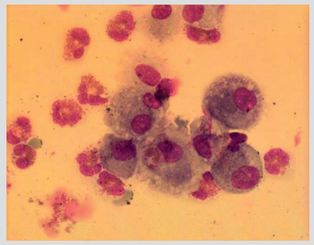

Figure 4: Bronchoalveolar fluid demonstrating eosinophils and macrophages

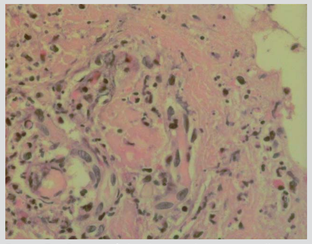

Figure 5a: Biopsy of skin lesion, showing capillaritis with fibrinic thrombi and eosinophilic inflammation

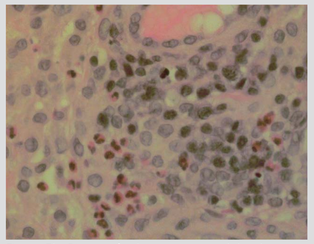

Figure 5b: Biopsy of subcutaneous nodule, showing multinodular basophilic necrosis with eosinophilic inflammation

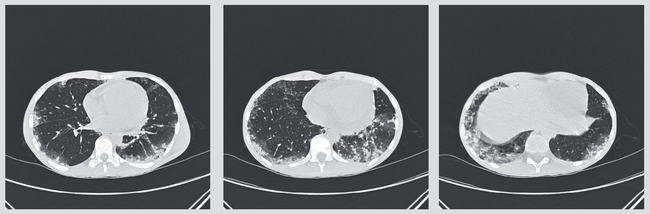

Figure 3a-3c: Chest computed tomography revealing bilateral pulmonary infiltrates with lower lobe predominance, peripheral consolidations and some pericardial effusion

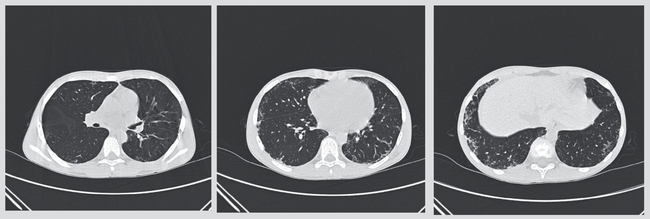

Figure 6a-6c: Chest computer tomography after six days, revealing substantial improvement with less interstitial peripheral abnormalities

Results: additional investigations

White blood cell differentiation and blood smear confirmed leukocytosis (31.9x109/L) and hypereosinophilia (12.40x109/l, 39% eosinophils) (fig. 2). C-reactive protein and erythrocyte sedimentation rate were raised (66 mg/L and 64 mm/hr respectively). Immunoglobin (Ig) levels were elevated for IgG (27.7 g/L) and IgE (2445 kU/l). Complement C3 and C4 were normal. Anti-nuclear antibody, anti-streptolysine-O and anti-DNA antibodies, as well as antineutrophilic cytoplasmatic antibody (p-ANCA, MPO-ANCA) were all negative. Rheumatic factor was minimally elevated (22 U/L). Urinalysis, renal and liver function tests were normal.

Chest radiography and computed tomography (CT) revealed bilateral infiltrates with lower lobe predominance and peripheral consolidations, as well as some pericardial effusion (fig. 3a-3c). Bronchoalveolar lavage fluid demonstrated leukocytes of 1.8x109/l with 76% eosinophils (fig. 4). Biopsy of the skin lesion showed capillaritis with fibrinic thrombi and eosinophilic inflammation.
Biopsy of the subcutaneous nodule showed multinodular basophilic necrosis with eosinophilic inflammation. Both biopsies were suggestive of eosinophilic vasculitis (fig 5a-5b).

Results: therapeutical intervention

Because of prominent peripheral blood eosinophilia, pulmonary involvement, cutaneous involvement and a history of asthma, Churg-Strauss syndrome was considered. High dose prednisone was started.

Symptoms improved rapidly. Blood eosinophilia declined rapidly within one week from 12.40*109/l to 0.04*109/l . Chest CT was repeated after six days and revealed substantial improvement with less interstitial peripheral abnormalities (fig 7a-7c), and confirmed rapid steroid responsiveness.

Extensive cultures and serologic tests were negative for infectious diseases caused by bacteria, viruses, parasites or fungi. Ziehl-Neelsen stain showed no acid-fast bacteria, tuberculosis culture was negative.

After nine days of hospitalization, the child was discharged to home. He was treated with oral prednisone (2 mg/kg/day) maintenance therapy for ten weeks. Thereafter, the dose was slowly tapered down to 0.3 mg/kg every other day twenty-six weeks after diagnosis.

Conclusion

This case report describes the rapid steroid-responsiveness in a 12-year old boy. Despite the low incidence of CSS in children, physicians must be aware of this rare disease in order to provide adequate therapy.