ASTHMA IN A FOURTEEN YEAR-OLD GIRL

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An online version of this case can be found at:
http://www.eaaci.net/site/content.php?l1=17&sel=668
This is a case report of a fourteen-year-old girl diagnosed with asthma since the age of twelve, medicated with budesonide (400 µg) and salbutamol. At the age of thirteen years and ten months she was admitted with an asthma crisis and medicated with clarithromycin, prednisolone, salbutamol, and ipratropium bromide. After this, she began taking montelukast (10 mg/day). One month later, she was admitted again and treated with the same drugs. She had multiple emergency admissions with asthma crisis and received similar treatment. At the age of fourteen, she was admitted with mottled reticulated erythematous violaceus pattern on the skin (Fig. 1), myalgias, and refused to walk, developing a neuropatic gait. At this point, she exhibited eosinophilia higher than 10%.

Based on the medical history and the laboratory study on admission, which are the differential diagnosis hypotheses?
Several differential diagnosis were considered such as hypereosinophilia, polyartheritis nodosa, Wegener granulomatosis, and Churg Strauss Syndrome.

Based on the differential diagnosis hypotheses, which clinical exams we can perform?
Laboratory studies showed the following results: Leucocytosis of 24500 with 57% eosinophils, erythrocyte sedimentation rate (ESR)60 mm/h, renal and liver function were normal. The IgE was 2568 KU/L, antinuclear and antineutrophilic cytoplasmic antibodies were negative. Viral serologies and parasitologic studies were negative.
An imagiologic study was also made, chest radiography, chest CT scan, abdominal and renal ecography were normal and cerebral MRI showed pansinusitis (Fig. 2). Neurophisiologic showed acute axonal polyradiculoneuropathy (mainly motor). Microscopy revealed extravascular eosinophilic infiltrates without vasculitis (Fig. 3).

**Fig. 2**

**Fig. 3**

**BASED ON THE RESULTS OF THE CLINICAL EXAMS, WHAT IS YOUR DIAGNOSIS NOW?**

CSS is an uncommon entity that is rarely reported in pediatric patients. The diagnosis is based in at least four of six diagnostic criteria, including: asthma, eosinophilia>10%, mononeuropathy or polynuropathy, pulmonary infiltrates, paranasal sinus abnormality, and extravascular eosinophilic infiltration in biopsy. Based on these criteria, this adolescent had four of the six diagnostic criteria: eosinophilia >10%, pansinusitis, polynuropathy, and extravascular eosinophilic infiltration in biopsy. The diagnosis of Churg Strauss Syndrome was made.

**WHAT ACTION SHOULD BE TAKEN?**

This adolescent had diagnostic criteria for CSS, so she started prednisolone (2mg/kg/day). Inhaled budesonide was maintained and montelukast was suspended. Methotrexate (10 mg/m2/day) was initiated as a steroid potential sparing agent. Subsequently, her corticosteroids
were weaned to a dose of 1 mg/kg/alternate day, but when reduction of corticosteroids was tried she exhibited multiple asthma exacerbations. Currently, her asthma is controlled with prednisolone 0.5 mg/kg/alternate day, methotrexate 7.5 mg/m²/week, budesonide 800 µg DPI, and salbutamol 400 µg DPI in crisis.

**DISCUSSION**

This case described an adolescent with a history of asthma, sinusitis, peripheral eosinophilia, and eosinophilic infiltrates in skin biopsy. Secondary causes of eosinophilic syndromes were ruled out.

Churg Strauss Syndrome was described by Churg Strauss in 1951, the original report was based on autopsy data. Reports of CSS occurring in children consist of single-case reports. It is conceivable that patients may be under diagnosed because of the lack of specificity, a low index of suspicion, and the invasiveness often involved with pathologic confirmation. The symptoms in pediatric patients demonstrate that CSS in children may present findings typical of adults. Only ANCA seems to be significantly different between children and adults, as there are very few case reports of CSS ANCA positive in children.

Specific clinical criteria developed by the American College of Rheumatology (ACR) for the classification of CSS include a history of asthma, eosinophilia>10%, mononeuropathy, polyneuropathy, non-fixed pulmonary infiltrates, paranasal sinus abnormality, and a biopsy with extravascular eosinophils. The presence ≥ 4 of the six criteria are thought to have a diagnostic sensitivity of 85% and a specificity of 99%. This patient presented four of the six criteria: the history of asthma, sinusitis, eosinophilia>10%, and skin biopsy revealing extravascular eosinophilia. The cornerstone of treatment remains corticosteroid therapy. Methotrexate was required in this case as a steroid sparing agent to reduce the lateral effects of corticotherapy. While there are no studies about prognostic factors in children, cardiac and renal involvement decreases the survival of affected adult patients and this seems to be similar in paediatric patients.

**CONCLUSION**

The triggering cause of CSS remains unknown. Several asthma medications have been suspected. Leukotriene antagonists allowing reduction of CCT would unmask incipient CSS as well as CCT tapering by itself. Macrolides have also been associated. This patient had a small course of two weeks of montelukast without CCT reduction. She was also treated with macrolides. Could this be a coincidence or a true association between these drugs and the etiopathogeny of CSS?

**References**

5. McDanel D, Muller B: The linkage between Churg Strauss Syndrome and leukotriene receptor antagonists: fact or fiction? Therapeutics & Clinical Risk Management, 2005;125-140
SUMMARY
We describe a case report of Churg Strauss Syndrome (CSS), which is uncommon in paediatric patients.

KEY WORDS
Churg Strauss Syndrome, asthma