Churg Strauss Syndrome after Polypectomy Procedure and On and Off Prescription of Monteleukast: Case Report

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Abstract
Churg Strauss syndrome is considered a systemic vasculitis in asthmatic patients with important diagnostic signs of hypereosinophilia, pulmonary infiltration, vasculitic skin lesions and nasal polyps and mononeuritis multiplex also supports this diagnosis. Here we present a patient with long history of asthma, polypectomy, abdominal pains and treatment by different brands of monteleukast.

Introduction
Churg Strauss syndrome (CSS) is considered a systemic vasculitis in asthmatic patients with important diagnostic signs of hypereosinophilia, pulmonary infiltration, vasculitic skin lesions and nasal polyps and mononeuritis multiplex also supports this diagnosis. Here we present a patient with long history of asthma, polypectomy, abdominal pains and treatment by different brands of monteleukast. The differential diagnosis includes Wegener’s granulomatosis, drug reaction, bronchogenic granulomatosis, fungal and parasitic infections, and malignancy [1].

LTRA’s (Leukotriene receptor antagonists) have important role in steroid sparing in the control of asthma. Yet, CSS has been reported as a possible side effect in people with moderate to severe asthma taking these medications. LT’s are also called slow reacting substance of anaphylaxis [2].

CSS has been reported in the general population with 20 times higher incidence in patients who have been treated with LTRA’s. This has been true for patients on previous glucocorticosteroid treatment or not, people with concurrent glucocorticosteroid treatment and individuals on glucocorticosteroid (GCS) taper while receiving LTRA’s [2,3].

Case Presentation
The patient is a 48 year old male with long standing history of asthma, nasal polypectomy 18 years ago who developed disseminated skin erythema after using monteleukast, eosinophilia and abdominal pains. Six months prior to this incidence, he had been using monteleukast of a different brand and had stopped. Diagnosis of Churg-Strauss was given two months prior and he was taking prednisolone medication. He has severe allergy to kiwi fruit and seasonal allergy in the season of spring.

He denied any severe allergies that had led to hospitalization. Patient notes productive cough, dyspnea (FC II) and blood clot in sputum in the past 2 days and melena. He complains of fever and chills with weight loss of 15 Kg in the past 3 months.

On physical exam, he was in well condition without respiratory distress or being ill and toxic. Vital signs were T 37.5°C oral, P 88, RR 20, BP 110/80 and pulse oximetry of 91% on room air. Head and neck exam was normal. Diffuse wheezing was ausculted on pulmonary exam. He had no murmurs, rubs or gallops. Abdominal exam was normal without organomegally. Examination of extremities showed edema in the left lower extremity and his right hand. Skin rash (erythematous papules) was observed in all extremities with numbness and tingling in acral areas of the lower extremity and hand.

Laboratory exam showed a WBC of 14700 with 14% eosinophilia and 60% neutrophilia, Hgb=14.9, Hct=42.6, ESR=42. Coagulopathy tests were normal. Creatinine was 1.2. Electrolytes and UA was normal. ANCA was negative. IGE was > 500.

NCV, EMG
Assymetric axonal and sensory peripheral neuropathy process that is indicative for mononeutitis multiplex.

Spiral CT scan of thorax
Patchy asymmetric bilateral pulmonary infiltrations

Coronal CT of sinuses
Showed thickening of maxillary, ethmoid and frontal sinuses.

During hospitalization patient underwent endoscopy were fungal esophagitis was noted.

Skin biopsy showed superficial mild perivascular neutrophilic infiltration. Sputum smear for AFB was negative and patient was started on methylprednisolone pulses, endoxan and fluconazole.

Melena resolved and he felt overall better and was discharged with medications prednisolone, fluconazole and to follow-up in clinics.

Discussion
With the advent of leukotriene antagonist medications for the treatment of asthma, many cases of CSS and hypereosinophilia have been reported particularly with zafirleukast (cysteinyl leukotriene antagonist) [4,5].

Leukotriene receptor antagonists have been popularly prescribed since 1990 for the treatment of asthma [6]. Receptors are located in small endothelial vessels and lead to upregulation of P-selectin expression which leads to inflammation in vasculature and it seems that cysteinyl leukotriene receptor 1 can lead to production of leukotrienes

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and cause symptoms of CSS syndrome via receptors that have not been blocked [7].

Incidence of CSS in the general population has been estimated at 4 cases/million individuals. This is while in the year 2000, FDA representatives reported adverse effects on 5 asthma medications and their report included 151 patients who met 2 ACR criteria for diagnosis of CSS while being treated with LT modifiers. Of these individuals, 88% were being tapered of glucocorticosteroids (GCS) and 12% had never or recently used GCS [3].

Review of medline of 62 cases in which CSS occurred after LTA treatment, 3 groups were identified all which suggested temporal relationship between LTA use and development of CSS: 1) group with no previous steroid treatment (7 individuals never got steroids) 2) group with steroid treatment pill or inhaler and no change in treatment when LTA was added 3) group with reduction in steroid treatment with addition of LTA to their regimen. Mean age was 47 years and 55% were female. Most patients developed CSS within 6 months of LTA treatment [8].

Even surgical procedures such as nasal polypectomy have been implicated. One of the theories in the occurrence of this syndrome includes decreased systemic glucocorticoids which have unmasked the CSS syndrome [9]. Nasal polypectomy and some surgeries increase cytokines such as Tumor necrosis factors (TNF’s), interleukins and interferons [10-12].

Leukotrienes were discovered nearly 60 years ago, also known as slow reacting substance of anaphylaxis (SRS-A) [11] which were later named cysteiny1 leukotrienes including LTC4, LTD4, LTE4 which contain the amino acid cysteine [13]. In vivo and vitro studies have shown that LTC4, LTD4, LTE4 as potent recruiters for eosinophils can cause pathologic changes in asthma including increased vascular permeability, chemotaxis and lead to contractibility in airway smooth muscle [14,15]. Eventhough CysLT1 function by activating CysLT1 receptors, yet LTC4, LTD4 are not indolent. As a result during the past few decades, drug companies have attempted to produce medications named LT-modifier drugs. These drugs have two pharmacologic components one which blocks 5-Lipoxygenase and other acts as leukotriene receptor antagonists. LTRAs block CysLT1 receptors and biologically inhibit LTC4, LTD4. Of the most popular approved in the US is montelukast and zafirleukast which due to bronchodilatory and anti-inflammatory effect may improve pulmonary function and decrease symptoms in asthmatic patients. The indication for use of LT modifiers has been noted to be replacement after use of inhaled corticosteroids, cromolyn or theophylline for individuals above the age of 12 who have uncontrolled mild to moderate persistent asthma. CSS generally occurs in patients who are steroid dependent.

LT modifiers are considered safe and effective in patients with mild to moderate persistent asthma in place of inhaled corticosteroids as maintenance medication, not rescue medication. Beta-2 agonists should be taken as rescue medication. Steroid taper while taking LT antagonist may cause adverse effect.

Why is incidence of asthma increasing? Many environmental triggers, pollution, seasonal allergy and etc should be addressed. Concomitant multiple medication prescription should be addressed. Teaching use of peak flow meters have long been strongly advised for asthmatics and attending their friendly emergency room physician upon signs of worsening asthma as well. There is a lot a friendly emergency room physician can do in addition to relieving anxiety which is certainly not good for asthma attacks as well as administering oxygen and appropriate medications. Although this excludes a patient who presents with first time attack of CSS without any prior warning signs.

References