

**Case Report**

Hypereosinophilic Syndrome - A Case with Churg Strauss Vasculitis

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Abstract: The hypereosinophilic syndromes represent a group of disorders marked by the sustained overproduction of eosinophils, with eosinophilic infiltration of tissues and mediator release, leading to multiple organic damages. Churg-Strauss syndrome is a rare disorder characterized by systemic small vessel vasculitis, extra vascular granulomas and hypereosinophilia, characteristically occurring in people with background late-onset asthma and allergic rhinitis. Its annual incidence is between 0 and 4 per million of population. The authors present the case of a 72 year old Caucasian female patient, admitted at the Hygeia Hospital Tirana due to a hypereosinophilic syndrome. The thorax CT scan verified bilateral pseudo nodular pulmonary infiltrates; the head MRI showed vascular bilateral lesions in periventricular white matter and centrum semiovale, right internal capsule and left caudate nucleus, and the nerve conduction study (NCS) revealed a distal mixed polyneuropathy. A fibro-gastroscopy with biopsy suggested an atrophic gastritis. The patient underwent a course of treatment with intravenous methylprednisolone, tapered thereafter to oral prednisone together with cyclophosphamide, with a prompt improvement of the clinical picture.

Keywords: Hypereosinophilia, Churg-Strauss Syndrome, Vasculitis, Systemic Changes

1. Introduction

The hypereosinophilic syndromes (HES) are a group of disorders marked by the sustained overproduction of eosinophils, in which eosinophilic infiltration and mediator release cause damage to multiple organs [1-7]. Hypereosinophilic syndrome (HES) is used to describe a group of heterogeneous clinical syndromes; it does not necessarily imply a primary hematologic or neoplastic disorder. Only the following two features are required:

- a Hyper-eosinophilia (i.e., absolute eosinophils count $\geq 1500/\text{microL}$) on at least two occasions;
- b Signs of organ dysfunction attributable to the eosinophilia [3, 5, 9].

Churg-Strauss syndrome (Eosinophilic granulomatosis with polyangiitis) is a rare disorder characterized by systemic small vessel vasculitis, extra vascular granulomas and

hypereosinophilia, which characteristically occurs in people with background late-onset asthma and allergic rhinitis [10]. Its annual incidence is between 0 and 4 per million population and prevalence is between 7 and 22 per million [12]. This is very rare for Albania, a country with 2891124 inhabitants [17]. It is reported the presence of perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) in 40% of the patients [13]. According to the American College of Rheumatology (ACR), there are four or more criteria out of six for the diagnosis to be made: asthma, eosinophilia ($> 10\%$ in peripheral blood), paranasal sinusitis, pulmonary infiltrates, and histological evidence of vasculitis with extra vascular eosinophils, and mononeuritis multiplex or polyneuropathy [11]. We present hereby a case of Churg Strauss vasculitis.

2. Case Report

We present the case of a 72 year old Caucasian female patient. She was admitted at Hygeia Hospital Tirana in August 2016 with a hypereosinophilic syndrome. The last three months she reported of extreme fatigue, anorexia, weight loss of 10 kg, persistent headache, pruritis, paresthesia in hands and foot, abdominal discomfort, photophobia, memory loss and pain in the right eye. She had a 6 year history of late onset asthma which improved within a year, anemia, renal failure, hemorrhagic gastric ulcer, microscopic hematuria and high inflammation tests. At that time she was attended by haematologist and pulmonologist because the CT scan showed bilateral subpleural infiltrates and lymph nodes enlargement. The lymph node biopsy 6 years ago was normal. We noticed a pale skin with no vasculitis signs, afebrile, marginal keratitis and corticonuclear cataract of right eye, no wheezing, normal cardiac sounds, and hypoesthesia gloves-stocks pattern with slight strength decrease in distal muscles, no signs of arthritis or oedema. Blood results revealed white blood cells (WBC) $14080/\text{mm}^3$, haemoglobin (Hb) 9.6 g/dl, creatinine 3.4 mg/dl, uraemia 117 mg/dl, uricemia 6.5 mg/dl, blood eosinophilia 30%, erythrocytation rate (ERS) 70 mm/h, C reactive protein (PCR) 37.5mg/L, albuminemia 2.8g/dL, 30 mg albumin in urinalyses, p-ANCA positive, 26% eosinophils in bone marrow aspiration, low complement C3 and C4, IgE 309 UI/mL; with negative test for parasites in stool analyses and Echinococcus. Tumor markers were within normal range, as well as echocardiography findings.

The thorax CT scan [Figure 1] verified bilateral pseudo nodular pulmonary infiltrates, the head MRI [Figure 2] showed vascular bilateral lesions in periventricular white matter and centrum semiovale, right internal capsule and left caudate nucleus, and the nerve conduction study (NCS) revealed a distal mixed polyneuropathy. A fibro-gastroscopy with biopsy suggested an atrophic gastritis. The stomach biopsy [Figure 3] detected moderate eosinophilic infiltrates and fibrosis in the antral mucosa. A differential diagnosis of the hypereosinophilic syndromes was performed and after we ruled out other diseases the diagnosis of Churg Strauss was made based on 4 ACR criteria [11].

We started a course of treatment with methylprednisolone 1g/d IV for three consecutive days and after that Prednisone 1mg/kg for one month together with cyclophosphamide 1000 mg once monthly IV. After three days therapy the number of eosinophils was 0 % in blood count and the patient clinical condition was good. The patient is on follow-up with a monthly therapy of 1g IV cyclophosphamide and tapering doses of prednisone.

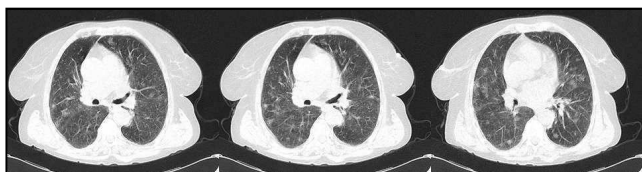


Figure 1. Chest CT with diffuse bilateral pseudo-nodular infiltrates.

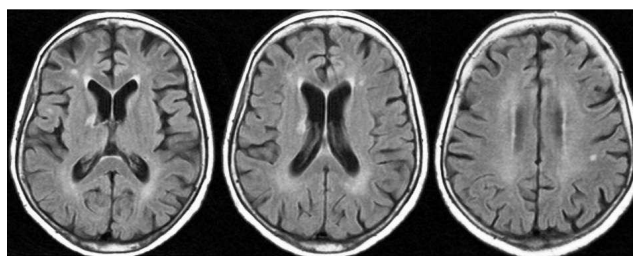


Figure 2. Head MRI. Vascular bilateral lesions in periventricular white matter and centrum semiovale, right internal capsule and left caudate nucleus.

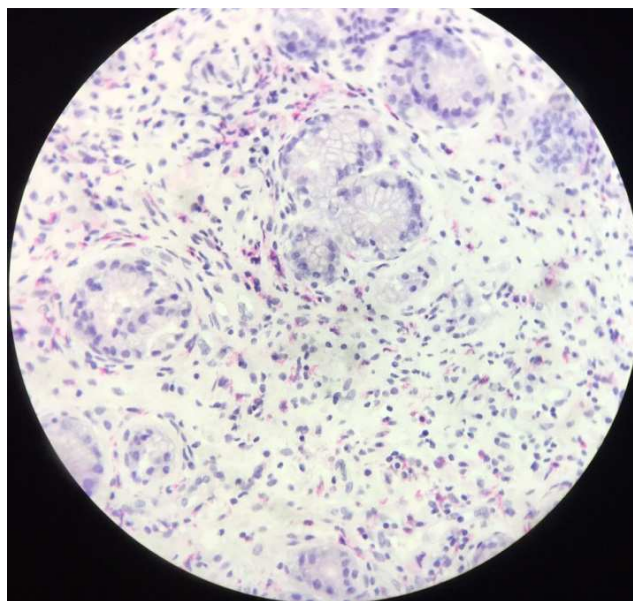


Figure 3. Gastric biopsy, with moderate eosinophilic infiltrates and fibrosis.

3. Discussion

Here we have presented a case that despite previous pulmonary, renal and gastrointestinal involvement, only the persistent blood eosinophilia raised suspicion of possible Churg Strauss vasculitis. A differential diagnosis of hypereosinophilic syndromes was made, with parasitic infectious (Echinococcus and nematodes), allergic, neoplastic, endocrine, Wegener's granulomatosis, drug reaction, bronchogenic granulomatosis, hematologic and pulmonary diseases [14]. In the differential diagnosis are included also immune-mediated diseases such as the idiopathic thrombocytopenic purpura, cryoglobulinemia, and collagen vascular diseases. Complement range are normal or elevated in vasculitis, whereas malignancies generally do not cause failure of major organs [16]. Polyarteritis nodosa (PAN) also involves small and medium sized vessels, and severe renal disease is common, but eosinophilia is rare [15]. Diagnosis is by clinical, pathological, and laboratory correlates. After we ruled out all these conditions associated with hypereosinophilia, the patient was diagnosed with Churg Strauss vasculitis.

The diagnosis was based on 4 out of 6 ACR criteria (history of asthma, pulmonary infiltrates, polyneuropathy and eosinophilia). We couldn't the suralis nerve biopsy;

meanwhile the stomach biopsy findings were not specific because vasculitis and granuloma can be present more often in small intestine and/or colon involvement [11]. The diagnosis was supported also by MRI findings, kidneys' involvement and the p-ANCA abnormal value.

4. Conclusions

Churg Strauss is a rare disorder, the least common of the ANCA associated vasculitis. Its annual incidence is between 0 and 4 per million population and prevalence is between 7 and 22 per million. It has a mean age of appearance between 45-50 years, with male female ratio of about 1. Eosinophilia is present in active untreated disease and often $>1500/\text{mm}^3$ (97% of patients) and serum IgE is raised in more than 75% of patients. As renal involvement is uncommon in Churg Strauss (usually $<25\%$ of patients), eosinophilia is a late finding and p-ANCA are positive only in 25-40% of patients, therefore a biopsy is very helpful [12]. Patient subgroups according to ANCA status are those with increased frequency of kidney involvement, constitutional symptoms, purpura, alveolar hemorrhage and nervous system involvement [13]. Renal abnormalities are presented with an ANCA-associated necrotizing crescentic glomerulonephritis; but the natural course of the treated disease, as well as the final outcome, usually are good [10].

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