DOI: http://doi.org/10.4038/jccp.v49i1.7845

Case report

Sensory neural hearing impairment in a patient with eosinophilic granulomatosis with polyangitis (Churg-Strauss Syndrome)

Bandara L M H[1], **Hittarage A**[1]

Journal of the Ceylon College of Physicians, 2018, **49**, 35-36

Introduction

Eosinophilic granulomatosis with polyangitis (EGPA) is a rare vasculitic disease, previously known as Churg-Strauss syndrome[1,2]. It is characterized by disseminated vasculitis associated with eosiophilia and extravascular granuloma[1]. Cranial nerve involvement is rare. We present a case of EGPA presenting with peripheral neuropathy and bilateral sensory neural hearing loss (SNHL).

Case report

A 71-year-old retired government officer from Ganemulla, presented in August 2017 with progressive dyspnoea, wheezing, lower limb numbness, low grade fever and anorexia for 2 weeks.

He is a non-smoker, does not take alcohol, He did not have diabetes and had no significant family or drug history. He had been diagnosed as having late onset bronchial asthma (BA) 5 years back. He suffered several exacerbations of BA and used inhaled corticosteroids and a short acting beta-agonist. His hearing has been declining during the last year. There was no history of ear trauma or discharge. There was no history of sinusitis, skin rashes, Raynaud's phenomenon, photosensitivity or joint involvement.

He had bilateral foot and leg numbness, and during hospital stay developed bilateral foot drop. He had no fever, jaundice, pallor, clubbing or oedema. Cardiovascular and abdominal examination was normal. Respiratory examination, revealed fine crepitations in right mid zone. Stocking type sensory impairment of both legs was present which progressed to weakness of legs distally. Hearing was severely affected in both ears and there were no other cranial nerve involvement. He did not have any tinnitus or vertigo.

Investigations showed leukocytosis of 29×10^3/µl with 55% of eosinophils (15,950/µl), haemoglobin of 11.5 g/dl, platelets of 360×10^3/µl. Blood picture revealed severe eosinophilia. Urine full report showed albuminuria of 2+, 40-50 red blood cells with granular casts. Urine protein creatinine ratio was 34 mg/mmol, serum creatinine 102 µmol/l. Serum transaminase, bilirubin and protein levels were normal. C-reactive protein was 11.6 mg/dl, erythrocyte sedimentation rate was 86 mm. Chest X-ray showed right mid zone reticular nodular shadowing. There was eosinophiluria and more than 10% eosinophils in sputum. Serum immunoglobulin E (IgE) level of 10,000 IU/ml was detected while filarial antibody, toxoplasma antibody and *Aspergillus fumigatus* specific IgE antibodies were negative. Cultures didn't yield any bacterial growth. Stool examination didn't detect parasites or ova and sputum fungal culture didn't yield any fungi. Retroviral and syphilis screening were negative.

Perinuclear/myeloperoxidase-antineutrophil cytoplasmic antibodies (P-ANCA/MPO-ANCA) was positive while cytoplasmic-ANCA (C-ANCA/PR3-ANCA) was negative. Contrast enhanced computed tomography (CT) and high-resolution CT showed evidence of mild pneumonitis in perihilar region without significant lymphadenopathy or organomegaly. Repeat chest X-ray showed cleared shadows. CT of nasal sinuses showed no sinus infiltration. Echocardiography showed mild aortic sclerosis and trivial mitral regurgitation. Bone marrow biopsy showed elevated eosinophil precursors and no evidence of lymphoma. Sural nerve and inguinal lymph node biopsy showed perivascular aggregates of eosinophils extending to vessel wall of small and medium sized vessels (Figure 1). Hearing assessment showed bilateral profound SNHL.

The patient was started on 50 mg of prednisolone daily with osteoporosis prophylaxis. A rapid eosinophilic response was seen with absolute eosinophil count reduced to 275/µl in 6 hours. Sensory impairment started improving in one week. Hearing assessment in three weeks showed improvement. Motor weakness was recovering with physiotherapy.

[1] *Colombo North Teaching Hospital, Ragama, Sri Lanka*

Corresponding author: *Bandara L M H*

E-mail: 1harshanabandara@gmail.com

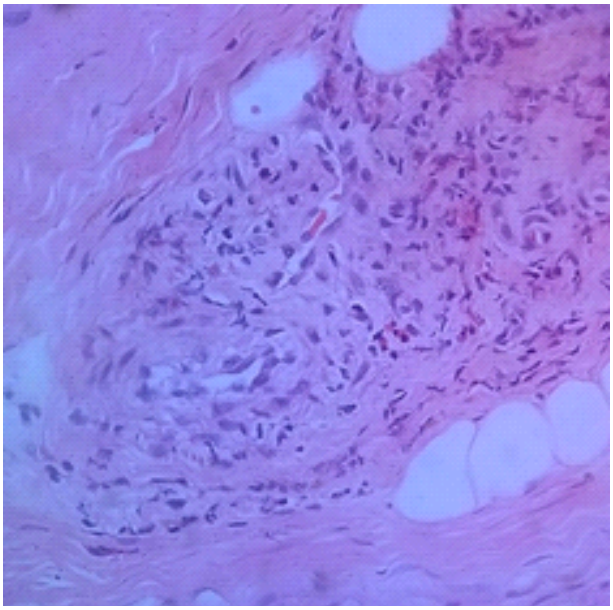

Figure 1. Microscopic view of sural nerve biopsy.

Discussion

EGPA is classified under small vessel vasculits associated with presence of P-ANCA/MPO-ANCA in 30-38% of cases[1]. Three phases in pathogenesis are allergic, eosinophilic and vasculitic. Clinical manifestations include history of severe asthma, sinus/ upper respiratory tract and pulmonary inflammation, skin and peripheral nerve involvement[3]. Renal, cardiac and gastrointestinal involvement carries a poor prognosis (Five-Factor Score)[3]. American College of Rheumatology has suggested six criteria for EGPA requiring four to diagnose with 85% sensitivity and 99.7% specificity[4]. Criteria include 1) History of asthma, 2) Eosinophilia of >10% from total leucocytes, 3) Mononeuropathy (including multiplex) or polyneuropathy, 4) Migratory or transient pulmonary opacities, 5) Paranasal sinus abnormality and 6) Biopsy containing a blood vessel showing the accumulation of eosinophils in extravascular areas[4]. In our patient four criteria were positive including histology with P-ANCA/MPO-ANCA positivity. Treatment of EGPA involves prednisolone 1mg/kg/day which is tapered over one year and immunosuppression with cyclophosphamide, ciclosporin or interon-alfa in advanced cases[1,2].

References

1. Noth I, Strek ME, Leff AR. Churg-Strauss syndrome. *Lancet* 2003; **361**(9357): 587-94. doi:10.1016/S0140-6736(03)12518-4
2. Greco A, Rizzo MI, De Virgilio A, Gallo A, Fusconi M, Ruoppolo G, Altissimi G, De Vincentiis M. Churg-Strauss syndrome. *Autoimmun Reviews* 2015; **14**(4): 341-8. doi:10.1016/j.autrev.2014.12.004
3. Mouthon L, Dunogue B, Guillevin L. Diagnosis and classification of eosinophilic granulomatosis with polyangiitis (formerly named Churg-Strauss syndrome). *J Autoimmun.* 2014; **48-49**: 99-103. doi:10.1016/j.jaut.2014.01.018
4. Byun J-H, Lee J-H, Choi I-S. Churg-Strauss Syndrome presenting with Hearing Impairment and Facial Palsy. *Annals of Rehabilitation Medicine* 2014; **38**(6): 852-5. doi:10.5535/arm.2014.38.6.852
5. Ishiyama A, Canalis RF. Otological manifestations of Churg-Strauss syndrome. *Laryngoscope* 2001; **111**(9): 1619-24. doi:10.1097/00005537-200109000-00024