Churg-Strauss angiitis in a 64-years old woman with gait difficulty

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ABSTRACT: Peripheral neuropathy is often a feature of systemic diseases. We report a case of a 64-year-old woman with gait difficulty due to multifocal sensorimotor polyneuropathy which was diagnosed as Churg-Strauss syndrome and presented clinical improvement after oral prednisolone administration. The clinical suspicion of this condition is crucial to early and correct diagnosis.

Keywords: Churg-Strauss, sensorimotor polyneuropathy
Churg-Strauss syndrome. A skin and muscle biopsy showed perivascular infiltrations of eosinophils (fig 1b), so the diagnosis of Churg-Strauss angiitis was confirmed according to American College of Rheumatology diagnostic criteria. Other primary systemic vasculitis were excluded due to the presence of asthma and blood hypereosinophilia. Furthermore the presence of asthma excluded the diagnosis of idiopathic hyper-eosinophilic syndrome. Prednisolone 1mg/kg per day was administered with dose tapering within few weeks. The patient mildly improved directly and after one month she was walking without support, showing impressive clinical improvement.

DISCUSSION
Churg-Strauss angiitis is a rare disorder with prevalence between 10.7-13 per 1000000 adults. The diagnosis is very difficult. The presence of asthma, rhinitis or sinusitis associated with peripheral eosinophilia and symptoms suggesting vasculitis, supports the diagnosis. Tissue biopsy should be obtained. ANCA are positive in only 40% of patients. ANCA status is associated with a particular clinical phenotype and, possibly, with a different pathogenesis and a different genetic background. [2,3] Our patient was ANCA negative but had other abnormal immunological factors such as CRP, ESR and ANA. Our patient fulfilled five out of six diagnostic criteria of American College of Rheumatology (polyneuropathy, asthma, eosinophilia >10%, pulmonary infiltrates and extravascular eosinophilic infiltration in tissue biopsy), while only four are required to establish the diagnosis of Churg-Strauss angiitis. In order to lead to the final diagnosis of this rare form of angiitis one should take into consideration the longitudinal medical history of pulmonary disease.

Figures: 1a. Chest CT scan showing fibrotic atelectatic features bilaterally. 1b. Skin muscle biopsy showing perivascular infiltrations of eosinophils.
Αγγειώτιδα Churg-Strauss σε 64χρονη γυναίκα με διαταραχή βάδισης

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Περίληψη: Η περιφεριακή πολυνευροπάθεια αποτελεί συχνά εύρημα συστηματικών παθήσεων. Παρουσιάζουμε περιπτώσεις σε 64χρονη γυναίκα με διαταραχή βάδισης λόγω πολυεστιακής αισθητικοκινητικής πολυνευροπάθειας που διεγνώθηκε ως σύνδρομο Churg-Strauss και παρουσιάζει βελτίωση μετά την από το στόματος χορήγηση πρεδνιζολόνης. Η κλινική υποψία του συνδρόμου αυτού είναι κρίσιμη για την έγκαιρη διάγνωση και την ουσιώδη θεραπεία του.

Αξέχαστα Κλειδιά: Churg-Strauss, αισθητικοκινητική πολυνευροπάθεια

REFERENCES