Eosinophilic Granulomatosis with Polyangiitis (EGPA) presenting with acute motor polineuropathy

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Introduction.

Churg–Strauss syndrome (CSS), also known as eosinophilic granulomatosis with polyangiitis (EGPA), was first described in 1951 by Churg and Strauss as a small vessel necrotizing vasculitis with extravascular granulomas occurring among patients with asthma and tissue eosinophilia and associated with antineutrophil cytoplasmic antibodies (ANCAs).
Clinical manifestation evolve through a prodromic phase, dominated by asthma or rhino-sinusitis, an eosinophilic phase, marked by peripheral eosinophilia and organ involvement, and a vasculitic phase, with constitutional symptoms (fatigue, fever, weight loss..) and manifestation of the small-vessel vasculitis (peripheral neuropathy, renal involvement, skin lesions).

The diagnosis of Churg Strauss Syndrome is commonly based on ACR (American College of Rheumatology), 1990 criteria (Table 1). Diagnosis is probable when four of six criteria are present: asthma, eosinophilia > 10%, neuropathy, non-fixed pulmonary infiltrates, paranasal sinus abnormality and extravascular eosinophil infiltration on bioptic findings. Because of the use of steroid therapy, identification of granulomas in biopsy is rare.
Therapeutical approach to EGPA has to be tailored to individual patient because depends on his prognostic profile. Usually clinical remission is obtained with administration of high dose of corticosteroid. In addition to that, immunosuppressant therapy with azatioprine or metotrexate can be used.

Case presentation.

A 68y old woman was admitted to our department for bilateral distal lower limb paraesthesia and progressive weakness. Ten days before admission she reported a general malaise with marked generalized weakness and diarrhea without fever, with progressive worsening of symptoms.
On first examination the patient showed upper and lower limb distal hypoesthesia and lower limb weakness with hypo/areflexia.

Based on clinical presentation, a Guillain Barrè Syndrome (GBS) was first considered, also supported by a hypo/a-reflexia in the lower limbs.
Nerve Conduction Studies (NCS) showed a slight motor symmetric polyneuropathy with tibial conduction block and multifocal absent F-waves. However, cerebrospinal fluid analysis was normal and the symptoms showed no further progression after admission. The patient had a past history of asthma and a salivary gland cancer, treated with surgery and radiotherapy; blood findings showed high level of eosinophiles (>50%).

A diagnosis of Churg-Strauss syndrome with peripheral nervous system involvement was then hypothesized. The patient underwent further examinations (including echocardiography, renal function tests, chest radiography and other lab tests including immuno-rheumathological exams) without evidence of other organ involvement. p-ANCA were reported positive (antibodies to MPO, with a perinuclear fluoroscopic pattern). High-dose (1mg/kg/day) prednisone treatment was started with rapid and significant improvement of muscle strength.

Table 1. ***Diagnostic Criteria of EGPA***

❖ **Lanham et al.** Asthma Eosinophilia > 1,5 x 10^9 /liter Clinical or pathological evidence of vasculitis involving at least two organs
❖ **ACR (American College of Rheumathology), 1990** Asthma Eosinophlia > 10% Neuropathy (mono- or poly-neuropathy) Non-fixed pulmonary infiltrates Paranasal sinus abnormalities Extravascular eosinophil infiltration on biopsy
❖ **Chapel Hill Consensus Conference ,1994** Eosinophil-rich and granulomatous inflammation involving the respiratory tract, necrotizing vasculitis affecting small to medium-sized vessels and associated with asthma and eosinophilia

Discussion.

The patient presented with ascending weakness and hyporeflexia, more commonly observed in GBS; however, the history of asthma and the significant persistent eosinophilia raised the suspect of a Churg-Strauss Syndrome. Even though this patient does not fully meet criteria for EGPA diagnosis (expecially because of lacking of bioptic confirmation), clinical features and history, ANCA positivity and therapy response make this condition highly probable.
Peripheral nervous system involvement is quite common in patients with EGPA, reported in up to 60% at onset, especially in patients with positive ANCAs. Mononeuritis multiplex, multineuropathy or symmetric polyneuropathy are described, with the last one presenting in about 20% of cases.

Conclusion.

EGPA and other vasculitides should always be considered in the differential diagnosis of patients presenting with acute or subacute ascending polyneuropathy, mimicking Guillan Barrè Syndrome, mainly for the different therapeutic approach. The history of asthma and the findings of eosinophilia can help to address the suspect of a Churg-Strauss Syndrome and should drive to antibody testing and biopsy (sural nerve or skin biopsy).

References.

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