

Pulmonary arterial hypertension and orbital involvement in a patient with eosinophilic granulomatosis with polyangiitis

Sirs,

A 28-year-old male presented with an 8-year history of painless bilateral proptosis. He acknowledged sinusitis that was treated with surgery one year after the onset of the orbital symptoms. At that time he was diagnosed with Wegener's granulomatosis and was treated for 6 months with cyclophosphamide pulse therapy. On admission he was being treated with oral methotrexate. Ophthalmic examination showed gross bilateral proptosis (Hertel readings OD = 35 mm and OS = 30 mm), full eye motility and temporal optic disc palor in both eyes. Visual acuity was 20/40 OD, 20/60 OS. Magnetic resonance imaging revealed that both orbits were diffusely infiltrated (Fig. 1). A biopsy of the lacrimal gland and intraconal fat was performed. The results of the biopsy indicated polyclonal lymphoid hyperplasia. The patient was treated with orbital radiotherapy only. Two years later he presented again with severe bronchospasm requiring admission to the intensive care unit. He was intubated, treated with corticosteroids, antibiotics and vasoactive drugs due to septic shock. There was marked peripheral blood eosinophilia (36% of eosinophils – 4,960 cells/mL). Four months later he had no respiratory symptoms, but computed tomography of the lung showed diffuse ground-glass opacities and a mosaic pattern. Bronchoscopy and bronchoalveolar lavage were performed, revealing 79.4% eosinophils. His pulmonary function test showed moderate restriction but no obstruction (total lung capacity: 76.1% of predicted; forced expiratory volume in the first second: 66.1%; forced vital capacity: 68.3%) and his arterial blood gases were normal; c-ANCA, p-ANCA and HIV tests were negative. As the orbital infiltration persisted, another orbital biopsy was performed. Histopathological examination showed inflammatory infiltration and granulomatous inflammation with many eosinophils and small vessel vasculitis. In light of the clinical and histopathological diagnosis of eosinophilic granulomatosis with polyangiitis (EGPA), an echocardiogram revealed normal left chambers, mild to moderate tricuspid dysfunction and pulmonary hypertension (estimated systolic pulmonary artery pressure: 77 mmHg). Right heart catheterisation showed moderate pulmonary hypertension (mean pulmonary arterial pressure: 39 mmHg), normal venocapillary pressure and negative vaso-reactivity test. Computed tomography angiography of the lungs showed no evidence of pulmonary embolism (PE). Currently the patient is being treated with monthly

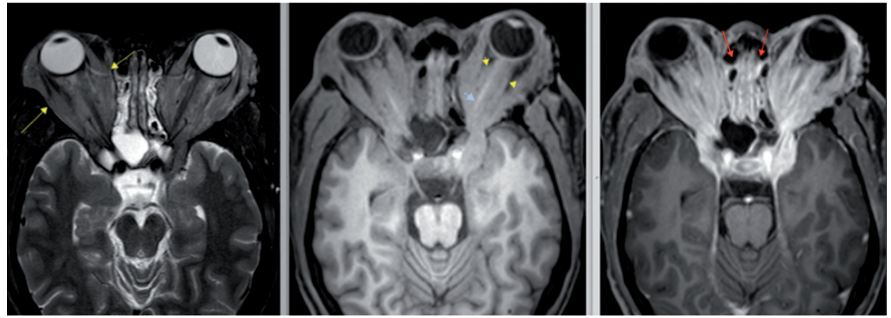


Fig. 1. Axial magnetic resonance imaging (T2 weighted) shows diffuse thickening of extrinsic orbital muscle (yellow arrows - left); optic nerve sheath thickening (yellow arrow head - centre), optic nerve (blue arrow - centre) and diffuse rhinosinusitis (red arrows - right).

cyclophosphamide pulse therapy and has no respiratory or cardiovascular symptoms, although proptosis persists. EGPA (formerly known as Churg-Strauss syndrome) is a rare small-vessel vasculitide associated with antineutrophil cytoplasmic antibodies (ANCA) (1). The specific roles of vasculitis and hypereosinophilia are unknown and usually there is considerable overlapping of both characteristics. Although any tissue can be affected by the disease, the most common manifestations are weight loss (49.3%), mononeuritis multiplex (46%), sinusitis (41.8%), skin lesions (39.7%) and lung infiltrates (3.6%) (2). Among connective tissue diseases and vasculitides, pulmonary arterial hypertension (PAH) is found mostly in scleroderma (incidence: 7–12%) (3). Although EGPA patients have a greater risk of PE (7.6% of patients) and cardiomyopathy (16.4% of patients), we are unaware of any report of pulmonary arterial hypertension secondary to EGPA, with no evidence of left chambers dysfunction or chronic thromboembolic pulmonary hypertension. It is unlikely that pulmonary hypertension was due to parenchymal lung disease, once there was little spirometric impairment and no abnormalities on arterial blood gases. Orbital inflammatory disease is found in 0.7% of EGPA patients at diagnosis (4). To our knowledge, this is the third report of bilateral exophthalmos and the second in which there was confirmation of extraocular myositis with eosinophilic infiltration in a patient with EGPA (5). In summary, this is an atypical case of EGPA with pulmonary arterial hypertension and very rare orbital involvement.

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