A Diagnostic Uncertainty- a Case of Eosinophilic fasciitis

Dr Sameena Khalid, Dr Gillian Smith (Rotherham General Hospital), Prof. Ariane Herrick (Salford Royal Hospital)

Background: Eosinophilic fasciitis is a rare disorder characterised by thickened, indurated skin that can be mistaken for systemic sclerosis

Clinical case: TH, a 59 year old gentleman, presented to his GP in July 2016 with a few weeks history of dry cough with gradual onset pain and stiffening in his legs and shoulders; paraesthesia in his lower limbs; poor grip strength and, unsteadiness on his feet. There was a history of proctitis in 2015 treated with mesalazine and possible pulmonary sarcoid at age twenty. C-Reactive Protein (CRP) was 85 mg/L and Erythrocyte Sedimentation Rate (ESR) was 25 mm/h. His GP had commenced prednisolone 30mg/day after making a presumptive diagnosis of polymyalgia rheumatica. The patient took prednisolone for only five days and then discontinued the medication due to ineffectiveness.

He was referred to the Rheumatology Department and seen in clinic in December 2016. The clinical features included significant skin thickening over the upper arms, forearms and thighs and lower legs with thickened tendons and mild flexion contractures of his fingers and weak grip. Investigations revealed Haemoglobin 139 g/L, a lymphopaenia of 0.6 $x10^9$/L and normal eosinophils. CRP 102 mg/L and ESR 34 mm/h. Rheumatoid factor, ANA, ENA and anti-centromere antibody were negative. Creatinine Kinase (CK), immunoglobulins and bone profile were normal.

In February 2017, a working diagnosis of diffuse systemic sclerosis was made, however, the absence of certain clinical features of established systemic sclerosis such as sclerodactyly and Raynaud's phenomenon with a normal auto-immune profile prompted an early revision of the diagnosis and there was concern about eosinophilic fasciitis. The MRI of lower legs was suggestive of inflammatory change in the lower limbs which could be in keeping with EF. Unfortunately, the skin biopsy did not contain sufficient deep fascia to allow for a histological diagnosis, although interstitial lymphoplasmocytic infiltrate was identified. TH then developed a sudden episode of thrombocytopaenia with platelet count 24 $x10^9$/L. A repeat FBC a few days later showed considerable improvement without any treatment.

The diagnosis was changed to EF in March 2017. Prednisolone was increased to 40mg od for one month and TH improved symptomatically and the CRP normalised. Mycophenolate Mofetil was commenced and the dose increased to 1g tds. The intention is to gradually reduce the dose of prednisolone over the next few months.

Conclusion: After working diagnoses of polymyalgia rheumatic and then systemic sclerosis, this patient was eventually diagnosed with EF. Although TH did not have hypergammaglobulinaemia or eosinophilia, the clinical pattern fit with a diagnosis of EF. The mainstay of treatment is with steroids although a number of immunosuppressive agents have been used. There is also a risk of haematological involvement, namely aplastic anaemia in patients with EF, although our patient only had a short lived decrease in platelet count only.

References:

1. Wright, NA, Mazori, DR, Patel, M, Merola, JF, Femia, AN, Vleugels, RA. Epidemiology and Treatment of Eosinophilic Fasciitis: An Analysis of 63 Patients From 3 Tertiary Care Centers. *JAMA Dermatol.* vol. 152. 2016 pp. 97-9.
2. Lebeaux D, Francès C, Barete S, Wechsler B, Dubourg O, Renoux J, et al. Eosinophilic fasciitis (Shulman disease): new insights into the therapeutic management from a series of 34 patients. *Rheumatology (Oxford)*. 2012 51(3):557-61
3. de Masson A, Bouaziz JD, Peffault de Latour R, Benhamou Y, Moluçon-Chabrot C, Bay JO, et al. Severe aplastic anemia associated with eosinophilic fasciitis: report of 4 cases and review of the literature. *Medicine (Baltimore)*. 2013 Mar. 92(2):69-81