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Eosinophilic fasciitis and eosinophilic colitis: a rare association

Sir, Although eosinophilic fasciitis (EF) was originally considered to be a disease predominantly localized to the fascia, several case reports have subsequently reported other manifestations [1–7]. Aplastic anaemia, haemolytic anaemia, thrombocytopenia, lymphoproliferative disorders, thyroiditis, pulmonary


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FIG. 1. Histology of deep skin biopsy showing features consistent with eosinophilic fasciitis. There is an inflammatory cell infiltrate in the deep subcutaneous tissue composed of lymphocytes, neutrophils and prominent eosinophils.
fibrosis, Sjögren’s syndrome, Raynaud’s phenomenon, myositis, medium vessel vasculitis, pericarditis, colitis and glomerulonephritis are some of the manifestations that have been reported in patients with EF. To our knowledge, the association of eosinophilic colitis (EC) and EF has been reported only once before [6]. Here, we describe an additional patient with EF who developed EC.

A 38-yr-old previously fit and healthy man initially presented to the general physicians in December 2000 with a 2-month history of fatigue and generalized myalgia that was worse after exertion. The onset of his illness was acute following several hours of extreme physical exertion at work. He denied any exposure to toxins, drugs or nutritional supplements prior to the onset of his illness. Physical examination was entirely unremarkable at this stage. In particular, he had normal power in all muscle groups. Routine laboratory testing revealed normal values for erythrocyte sedimentation rate, C-reactive protein and creatine kinase, but eosinophil count was elevated at $1.8 \times 10^9/l$ (normal range $0–0.4 \times 10^9/l$). Over the next several weeks, he developed increasing swelling in his ankles, erythema over his lower legs and restricted range of movements in his wrists and ankles. His skin was then noted to be indurated not only in his lower legs, but also in his forearms. His eosinophil count had steadily risen to $4.8 \times 10^9/l$ during this time. There were no clinical features suggestive of systemic sclerosis such as sclerodactyly, Raynaud’s phenomenon or nailfold capillary abnormalities. Pulmonary function tests and echocardiogram were entirely unremarkable. The relevant serological tests (antinuclear antibody and antibodies to extractable nuclear antigens) were negative.

A presumptive clinical diagnosis of eosinophilic fasciitis (EF) was made by the dermatologist and was confirmed on full thickness skin biopsy (down to superficial muscle) taken from his forearm (Fig. 1). Histological examination of the biopsy specimen showed an inflammatory cell infiltrate, predominantly composed of eosinophils, in the deep subcutaneous tissue and fascia with associated fibrosis. There was patchy, mild perivascular inflammation in the skin, but there was no definite abnormality in the skeletal muscle. The general physicians commenced him on 40 mg of prednisolone/day in May 2001, but it appeared that he did not derive any benefit from it. His eosinophil count fell to less than $0.2 \times 10^9/l$ even before prednisolone was commenced and he eventually reported subjective improvement in symptoms following intense physiotherapy. This was associated with softening of the affected areas on his limbs. The dose of his prednisolone was rapidly tapered after September 2001 and he was receiving only 5 mg/day when he first came to our attention in October 2001.

Soon thereafter, he developed persistent bloody diarrhoea and was referred for specialist gastroenterology opinion. Flexible sigmoidoscopy revealed severe colitis. Colonic biopsy samples showed active inflammatory reaction with focal clusters of eosinophils in lamina propria, consistent with eosinophilic colitis (Fig. 2). No ischaemic or vasculitic features were identified. In view of these findings, the dose of his prednisolone was increased and he was commenced on mesalazine. Cimetidine was also added later in view of anecdotal evidence of improvement in symptoms of EF (see below) [7]. Although there was good initial response with resolution of bowel symptoms with the above

![Fig. 2. Histology of colonic biopsy specimen showing features consistent with eosinophilic colitis. The lamina propria contains varying numbers of eosinophils in focal clusters.](image-url)
measures, he was being maintained on 7 mg of prednisolone/day (with appropriate bone protection) at the time of writing this letter, as his bowel symptoms flared each time the dose was reduced below this level. Methotrexate was being considered as a steroid-sparing agent to control his bowel disease.

Extracutaneous and visceral manifestations are increasingly recognized in patients with EF. Colitis has, however, been reported in such patients only twice, and EC only once. The only other EF patient who developed EC also had multisystem involvement with pericarditis, thyroiditis and monoclonal gammopathy [6]. In the other patient, the features of colitis were considered to be consistent with Crohn’s disease [7]. In our patient, EC occurred as an isolated visceral manifestation several months after the diagnosis of EF was made. It is difficult to prove that this association was not coincidental, but it may be possible to speculate that the mechanisms underlying the association are immune mediated. Although the exact immunological mechanisms have not been identified, there is some evidence for the role of T lymphocytes in the causation of EC. In a murine model of oral antigen-induced diarrhoea associated with colonic inflammation, colonic T cells have been shown to transfer the disease to naive mice through a STAT6-dependent mechanism [8]. The occurrence of hypergammaglobulinaemia and inflammatory cell infiltrate in the fascia and the association with autoimmune haematological disorders, Sjögren’s syndrome and thyroiditis support the role for immune-mediated mechanisms in the causation of EF. It should also be noted that EC in adults could be secondary to drug reactions and parasitic infections, and toxins such as L-tryptophan [9] and infection by Borrelia burgdorferi [10] have been reported as causative agents in EF.

In view of the rarity of this syndrome, evidence-based management drawn from controlled trials cannot be offered for patients with EF. Removal of the inciting agent (if one is identified) is the most important measure. Prednisolone is often used as most patients show partial or complete response. We, however, did not feel that prednisolone helped the EF component in our patient. Hydroxychloroquine, methotrexate and cimetidine have also been reported to be beneficial, of which the latter was used in our patient. Cimetidine is thought to act through blockage of lymphocyte function and it should be noted that a subpopulation of lymphocytes possesses H2 receptors [11]. In fact, it is believed that lymphocytes (and not eosinophils) play a primary role in the causation of EF. Therapy for EC in our patient was along the lines of inflammatory bowel disease, which is why mesalazine was considered suitable for the patient. Indeed, it was thought that prednisolone helped the EF component in our patient. Hydroxychloroquine, methotrexate and cimetidine have also been reported to be beneficial, of which the latter was used in our patient. Cimetidine is thought to act through blockage of lymphocyte function and it should be noted that a subpopulation of lymphocytes possesses H2 receptors [11]. In fact, it is believed that lymphocytes (and not eosinophils) play a primary role in the causation of EF. Therapy for EC in our patient was along the lines of inflammatory bowel disease, which is why mesalazine was considered suitable for the patient. Indeed, it was thought that prednisolone helped the EF component in our patient.

In conclusion, it is important to appreciate the widening spectrum of EF and carefully look for internal organ involvement. The pathogenesis of the association between EF and EC is unknown, but is possibly immune mediated. Treatment for these conditions is largely based on only anecdotal evidence.

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About the difficulty in interpreting ultrasonographic images of temporomandibular joint

Sir, We have read with interest the paper ‘A comparison of ultrasonography and magnetic resonance imaging in the evaluation of temporomandibular joint involvement in rheumatoid arthritis and psoriatic arthritis’ by Melchiorre et al. [1].

We would like to dwell upon the method of ultrasonographic (US) examination, unfortunately not described in the paper, for a few observations that, in our opinion, could help to better understand the images obtained with this technique.

The echographic study of the temporomandibular joint (TMJ) consists of different scans in coronal, axial and oblique planes [2, 3]. The exploration of the bone profile of the mandibular condyle can give very precise information about the condition of this structure, which is visualized through a window of 120° in the coronal scans and 40° in the axial scans. Further information can be obtained with the dynamic scans: they allow us to observe the condyle posterosuperior surface, to measure the anterior translation of the condyle, having as a landmark the tragus cartilage, and finally to study the articular and peri-articular soft tissues, included part of the morphology and the movement of the disc (Figs 1 and 2).

As for the study of the articular space in the axial scans, our experience suggests that the image of the capsulolonyovial thickening should be considered as the slightly echoic structure between...