Primary synovial non-Hodgkin’s lymphoma in association with ankylosing spondylitis

Sir, A 65-yr-old gentleman presented with a 3-month history of a painful, swollen left knee associated with left inguinal lymphadenopathy. There had been no symptoms of weight loss or fevers. There was a background of longstanding ankylosing spondylitis associated with inflammation of his left knee, requiring arthroscopy in 1984. This had shown extensive synovitis but nothing more specific on biopsy. The knee symptoms resolved over a period of months.

A plain film of the left knee had shown some bony destruction associated with an apparent large effusion. Clinically there was a large effusion but aspiration of fluid was difficult and the synovium had a wooden quality to it. The synovial fluid was bloodstained and no malignant cells were seen. The total white blood cell count was 1.3 x 10^9/l (polymorphonuclear cells 26%, monocytes 3%). Markers of inflammation were raised only modestly, erythrocyte sedimentation rate was 21 mm/h and C-reactive protein concentration 25 mg/l. The full blood count was normal and lactate dehydrogenase 361 U/l (normal range 211–423). An MRI scan demonstrated bony erosion with bone marrow oedema affecting the distal femur anteriorly and the medial femoral condyle and the tibia in the intercondylar region (Fig. 1). There was gross synovial hypertrophy affecting the suprapatellar pouch. A 3-cm mass was seen posterior to the femur; it had similar signal characteristics to synovium but no connection with the joint could be seen. This was thought to be an enlarged popliteal lymph node. Synovial involvement is less common and is usually due to direct extension from bone [1]. In this case the synovium was felt to be the primary site of the tumour due to the extent of the synovial hypertrophy compared with the amount of bony involvement. There have been few cases of primary synovial extranodal NHL reported in the literature and only two cases of lymphoma reported in association with ankylosing spondylitis, neither affecting the knee [2, 3]. The MRI appearances of synovial involvement from NHL have only been reported once in the literature [4].

The authors have declared no conflicts of interest.

### Table

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<td>Always bear in mind differential causes for a swollen joint even in an inflammatory joint disease, as important but rare causes may be missed.</td>
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Visual loss from optic neuropathy in dermatomyositis

Sir, A 58-yr-old woman with dermatomyositis was evaluated for painless bilateral visual loss of 2 weeks duration. Three months
prior to neuro-ophthalmic evaluation she developed multiple systemic symptoms including joint pain and stiffness, a rash on the arms, back and chest, fevers and weakness of the upper and lower extremities, which required hospitalization and treatment with intravenous corticosteroids. Laboratory evaluation showed an elevated creatine kinase of 201 U/l (normal range: 26–140). This prompted a thigh muscle biopsy which showed chronic inflammation consistent with dermatomyositis. The remainder of her laboratory evaluation was normal and did not reveal evidence of another collagen vascular disease. She had no prior history of hypertension, diabetes mellitus or cardiovascular disease.

She was discharged on 40 mg of oral prednisone which was gradually tapered over the next 3 months. The day after completing the prednisone taper she reported blurred vision in both eyes that gradually progressed over the next 4 days. During this time she also noted increasing weakness and pain in the extremities. She restarted 40 mg of prednisone per day and saw an ophthalmologist who noted bilateral optic disc oedema, without retinopathy, and referred her for neuro-ophthalmic evaluation.

Her visual acuity was 20/20 OD and 20/25 OS, and there was a left relative afferent pupillary defect. She identified 7 of 10 Ishihara pseudoisochromatic colour plates with the right eye and 2 of 10 with the left eye. Automated perimetry revealed superior and inferior arcuate visual field defects on the right and an inferior altitudinal defect on the left (Fig. 1). Dilated fundoscopy revealed resolving pallid optic disc oedema, with opacification of the superior and inferior retinal nerve fibre layer, on the left more than the right (Fig. 2). The remainder of the fundus examination was normal in each eye, without cotton wool spots or haemorrhages. Contrast-enhanced magnetic resonance imaging (MRI) of the brain and orbits was normal, without optic nerve enhancement. Her prednisone was increased to 60 mg per day and she reported no additional deterioration of vision. She returned 1 month later and her visual acuity and fields were stable. Her optic disc oedema had completely resolved, and the remainder of the fundus examination was normal.

Ocular involvement from dermatomyositis is an uncommon but well described phenomenon [1, 2]. Apart from the characteristic heliotropic rash which affects the eyelids, retinopathy, usually producing multiple cotton wool spots, is the most common finding, and may be asymptomatic [3]. Retinal involvement is believed to be due to vasculitis with endothelial disruption and platelet thrombi. In one report of 43 patients with previously diagnosed polymyositis or dermatomyositis referred for routine eye examination, six patients had retinopathy, all of whom were asymptomatic [3]. Some authors have suggested that retinopathy may be underestimated in patients with dermatomyositis and that routine ophthalmological evaluation may be warranted [3, 4].

Retinopathy is believed to be more common in children with dermatomyositis because accompanying vasculitis is more common at a younger age [2]. In the prior reports retinopathy has been clinically evident with fundoscopy, but one report suggested...
that intravenous fluorescein angiography (IVFA) may show more vascular involvement than is clinically obvious [5]. Despite this suggestion we were not able to find a patient who had evidence of involvement on IVFA without fundoscopically evident retinopathy.

The precise incidence of visual loss in dermatomyositis is not known, and in prior reports decreased vision has been attributed to retinopathy and infarction within the retinal nerve fibre layer [6]. Intraretinal haemorrhages and macular exudates have also been described as causing decreased vision in patients with dermatomyositis [6]. The retinopathy and visual loss appear to be reversible in most patients but persistent visual loss has also been noted despite treatment with corticosteroids and other immunosuppressive agents [6].

Optic neuropathy has been described in patients with dermatomyositis, but only in association with retinopathy [2]. We were unable to find a prior report of a patient with dermatomyositis who had optic nerve involvement without retinopathy using the following search terms: dermatomyositis, optic disc, vision, blindness, retinopathy (Pub Med, www.ncbi.nlm.nih.gov June 2003).

The timing of this patient’s visual loss, 1 day after discontinuing prednisone and in association with increasing myalgias and weakness, suggests that reactivation of the dermatomyositis was the cause of her visual loss. The relative afferent pupillary defect, dyschromatopsia, visual field defects and pallid optic disc oedema in the absence of retinopathy are evidence that the visual loss was from an optic neuropathy and not a retinopathy. Fortunately, reinitiation of corticosteroid therapy led to stabilization of her optic neuropathy with resolution of optic disc oedema and stabilization of visual function. She had no history of vascular disease, simultaneously swollen optic nerves and a relatively large cup-to-disc ratio in each eye, all of which would be extremely unusual for another cause of optic neuropathy such as non-arteritic ischaemic optic neuropathy [7]. We cannot totally exclude the presence of subclinical retinopathy and we did not perform IVFA, however there were no cotton wool spots or intraretinal haemorrhages characteristic of ocular involvement in patients with dermatomyositis.

It is not clear why retinopathy did not develop in this patient. Perhaps prior treatment with corticosteroids somehow protected the retinal vascular circulation. Spontaneous regression of retinal neovascularization has been noted in juvenile dermatomyositis, months after the initial fundus findings were documented [8]. The retinopathy in our patient may have been present prior to initial ophthalmic evaluation and disappeared by the time fundoscopy was performed, however her visual loss occurred just 2 weeks prior to neuro-ophthalmic assessment, during which time visual-impairing retinopathy would be expected to persist.

Optic neuropathy may occur in the absence of retinopathy in a patient with dermatomyositis. The absence of retinopathy does not preclude visual loss and should not delay the institution of corticosteroid therapy.

The author has declared no conflicts of interest.

### Key points

**Rheumatology**

- Optic neuropathy may occur without retinopathy in dermatomyositis.

### References


Vitamin D deficiency in a patient with systemic lupus erythematosus

Sir, You would not imagine that the patient illustrated opposite was vitamin D-deficient. However, she is a 21-yr-old Caucasian with systemic lupus erythematosus (SLE). Because of the nature of her condition she has to wear sunblock factor 30 all year round. Her tanned appearance is not derived from UVB exposure but from self-tanning agents.

She was diagnosed with SLE in 1997. Initially she presented with a widespread photosensitive rash. She was found to be positive for antinuclear antibodies and double-stranded DNA antibodies. A skin biopsy was consistent with cutaneous lupus. Photosensitivity tests demonstrated sensitivity to UVB light.

Soon after the diagnosis was established she was admitted to hospital with features of alveolitis, cerebral vasculitis and lupus nephritis. She was treated initially with steroids and intravenous cyclophosphamide. She is now maintained on prednisolone (11 mg once per day) and azathioprine (75 mg once per day).

The patient approached us concerned that she might be vitamin D-deficient because she was avoiding all sunlight exposure. Measurements revealed that she was vitamin D-deficient (25-OH vitamin D3 10.0 ng/ml, normal range 15–60 ng/ml). Other parameters were normal, including parathyroid hormone, alkaline phosphatase, calcium corrected for albumin and 25-OH vitamin D2 (which reflects preceding dietary intake). Her food diary confirmed a normal dietary intake of vitamin D. She was initiated on Calcichew D3 Forte (Shire, Basingstoke, UK) which contains 400 IU of cholecalciferol (vitamin D3). However, 25-OH vitamin D3 levels remained below the normal range despite normal hepatic function. The patient was felt to be compliant. Previous reports have suggested that high-dose steroids can lead to low serum concentrations of vitamin D3 [1]. This might explain the resistance in our patient. However, there is contradictory evidence, especially in patients taking low or moderate doses of prednisolone, such as our patient [2, 3]. It was felt more likely that the use of sunblock would necessitate doses greater than 400 IU, as reported in Moslem women [4]. She was started on alfalcaldiol 1 mg in order to increase the dose of vitamin D without increasing the calcium intake. Her renal function tests were normal at this time, though she did have persistent, stable proteinuria (0.25 g/l). Renal dysfunction would not explain her inability to convert vitamin D3 to 25-OH-vitamin.