

Intermediate uveitis associated with familial Mediterranean fever

Sirs,

Familial Mediterranean fever (FMF) is a recessively inherited disorder characterised by recurrent attacks of fever, peritonitis, pleuritis, arthritis and skin rash. Rarely, it can cause inflammatory eye disease. Here we describe a patient with known FMF presenting with unilateral intermediate uveitis that responded well to an increase in colchicine therapy.

A 27-year-old female of Sephardic ethnicity presented with sudden onset of floaters in the right eye. She had no past ophthalmic history but was diagnosed with FMF 13 years previously on a background of repeated episodes of hip pain, fever and pleuritis, which responded promptly to colchicine. There were no other family members affected by the disease. At the time of presentation she was systemically well and on oral colchicine 0.5 mg/day. Her visual acuity was 6/5 in both eyes with quiet anterior chambers and a normal intraocular pressure. On examination, she was found to have retinal vascular sheathing, mild vitritis and snowballs in her right eye. Fundoscopy of her left eye was normal and she had no evidence of subretinal fluid in either eye. Her fundus fluorescein angiogram showed wide-spread periphlebitis in the right eye with optic disc leakage and no cystoid macula oedema. The inflammatory changes were focused on the retinal veins with no arterial involvement and were entirely unilateral (Fig. 1).

Her clinical appearance was characteristic of intermediate uveitis. An infectious screen was carried out as well as well as a full blood count, urea and electrolytes, liver function tests, serum angiotensin converting enzyme, inflammatory markers and a chest x-ray. All investigations were within normal limits. She was not treated with any topical or systemic steroids and was reviewed 2 weeks later. The fundus appearance and visual acuity with the right eye remained stable and her colchicine was increased to 1mg/day. Over the course of 12 months, her vitritis and sheathing improved. At no point did she require any other treatment for intraocular inflammation or raised intraocular pressure and her vision remained 6/5 BE.

Ocular involvement is rarely associated with FMF. In 1959 Michaelson *et al.* reported a series of 23 cases between 6 and 39 years. They found that over 50% had sub-retinal colloid-bodies present with no other ophthalmic abnormalities (1). This finding has not been reproduced in the literature over the last 60 years. However, other

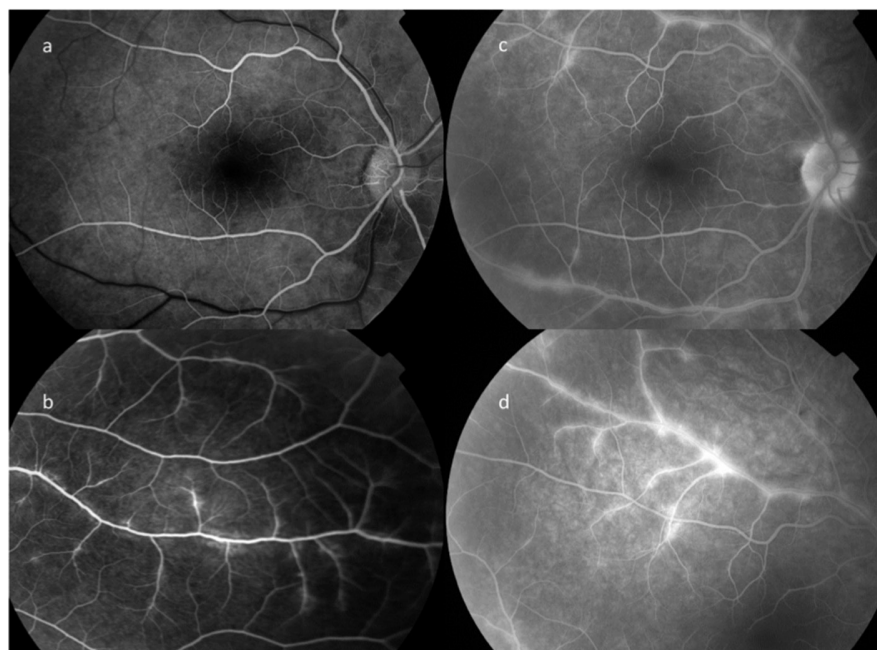


Fig. 1. Fundus fluorescein angiogram of the right eye in the arterial phase (a); venous phase – showing leakage from the retinal veins (b); and late phase – showing widespread leakage from the retinal veins and slight disk hyperfluorescence, with no cystoid macula oedema (c-d). There were no abnormalities in the left eye (not shown).

authors have described associations with anterior uveitis, episcleritis, intermediate uveitis and panuveitis (2, 3).

Patients who are prone to develop FMF are also more likely to develop Behçet's disease (BD) due to the geographic prevalence of the two diseases. In some instances, both diseases can be present (4). Patients with uveitis secondary to FMF may be misdiagnosed with BD due to their ethnicity and similar clinical phenotype. BD is a blinding, multisystemic inflammatory disease (5). It requires treatment with immunosuppressive agents and close follow-up. Whereas the uveitis found in FMF tends to run a milder course and often does not require treatment (6).

This case highlights the importance of observation in patients with uveitis who have a good visual prognosis and a self-limiting disease. As a result, this patient avoided unnecessary immunosuppressive treatment. Colchicine is the treatment of choice in FMF, the usual dose being 1–3 mg/day. In this case, the patient was taking 0.5 mg/day, which was insufficient to control the patient's disease. As such, she experienced a flare of her FMF in the form of intermediate uveitis. This case underlines the importance of establishing associated disease in patients with intermediate uveitis in order to guide management.

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