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Case Report Open Access

Unilateral Anterior Uveitis and Amaurosis Fugax in a Patient with Familial Mediterranean Fever

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Abstract

Purpose: To increase awareness and emphasize clinically relevant management issues for patients with familial Mediterranean fever (FMF) presenting with ocular inflammation and other ocular symptoms.

Method: A case of unilateral anterior uveitis and amaurosis fugax in a patient with FMF is described. The features of FMF including clinical signs and objective verification of a recurrence of an FMF attack are explained. Management of the described patient and patients with FMF presenting with ocular inflammation in general are discussed.

Conclusion: Ophthalmologists are encouraged to report more cases of eye symptoms in FMF patients to elucidate the panorama of ocular pathologies associated with FMF. Objective verification of a recurrence of FMF attacks at first visit is recommended and appropriate samples should be taken. It is also worth noting that eye manifestations in FMF patients may occur during systemic prophylactic treatment.

Keyword: Familial mediterranean fever; Uveitis; Amaurosis fugax

Introduction

Globally, familial Mediterranean fever (FMF) is the most common auto-inflammatory disease affecting 1 per 250 to 1 per 1.000 of the inhabitants in the Eastern parts of the Mediterranean area, where it is most prevalent [1,2].

FMF is an autosomal recessive disease caused by mutations in the *MEFV* (MEditerranean FeVer) gene, which encodes pyrin. The diagnosis is still based on characteristic clinical presentation. A gene analysis can confirm the diagnosis, but may fail to show the mutation [3]. Between attacks the patient is usually free of symptoms. Since 1972, colchicine is the main drug in the treatment of FMF [4].

Little is known about ocular involvement in FMF, although a few ocular manifestations have been described [5-11].

Case

A 28-year-old woman with FMF, pregnant in week 15, who had undergone uncomplicated bilateral refractive laser surgery (LASIK) four months earlier, presented with one day duration of redness, pain and photophobia in her right eye. The same day the patient had experienced episodes with transient greyish visual disturbances in the right eye lasting for a few seconds. One week earlier the patient had experienced five episodes of total visual loss in the same eye. No neurological deficits occurred. The patient experienced abdominal pain, typical for FMF attacks, coinciding with her ocular symptoms. Though no samples for objective verification were taken, the patient was convinced of having experienced a recurrence of FMF symptoms. At that time the patient was on treatment with colchicine 1 mg daily and she confirmed compliance to the treatment.

Uncorrected visual acuity at the first visit to our clinic was 20/40 in the right and 20/25 in the left eye. Intraocular pressure was 8 mmHg in the right and 9 mmHg in the left eye, as measured by rebound tonometry (ICare). The right eye showed fine keratic precipitates, mild flare and cells in the anterior chamber but no posterior synechiae. Posterior segment status was unremarkable. The left eye showed no signs of inflammation. Confrontation visual field testing showed no defects. Pupillary reactions, motility and colour vision were all normal. The patient was put on treatment with topical steroids (dexamethasone, 0.1 %) and cycloplegics (atropin, 1%). At follow-up one week later the symptoms had resolved and treatment was tapered. Uncorrected visual acuity was 20/20 in the right and 20/25 in the left eye. Doppler ultrasonography of the carotid arteries and echocardiography were normal. The patient's visual fields were examined with a Humphrey field analyzer (Carl Zeiss Meditec, SITA-fast program) at a later stage and the results were within normal limits.

Discussion

Ocular involvement in FMF of the anterior segment has been reported more frequently than affections of other parts of the eye. A few case reports also describe FMF patients with episcleritis and/or, as in this case, anterior uveitis [5,7,10,11]. Eye affection in the described patient occurred during a relapse of her FMF and indicates a possible association of the uveitis with her systemic disease. Scharf et al described two patients presenting with episcleritis during a free interval of FMF [10]. Episcleritis and later anterior uveitis in the same patient with FMF was reported by Yazici et al. [11]. Necrotizing blepharitis combined with nodular episcleritis in a patient with FMF on colchicine treatment was reported by Berestizschevsky et al. [7]. Three cases of bilateral uveitis, two of them pediatric, have been described [5,8,9]. One of these patients had a 7-year-old sibling with FMF presenting with episcleritis,

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suggesting a genetic familiar predisposition to ocular involvement [5]. Retinal detachment, presumably due to retinal tears secondary to posterior uveitis was also described in one of those patients [8]. Sub- and preretinal hemorrhage in a patient after secondary systemic amyloidosis due to FMF was reported by Altiparmak [6].

Knowledge of the common clinical signs of a recurrence of FMF is useful to determine whether ocular inflammation in a patient with FMF is related to the disease. FMF is characterized by recurrent episodes of fever and serositis involving mainly peritoneum, pleura and/or joints. This patient reported abdominal pain caused by serositis as a symptom typical for recurrences of FMF attacks. Pericarditis, erysipelas-like skin lesions, meningitis and testicular inflammation occur but are less common. These recurrent episodes usually last for up to three days. The disease is treated prophylactically with colchicines. The most severe complication in untreated individuals is systemic amyloidosis, leading to chronic renal failure [3].

To objectively verify a recurrence of FMF attack blood samples should be taken. There is a high inflammatory response during an attack. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), white blood cell count (WBC) and especially serumamyloid A protein (SAA) are elevated [12,13].

Acute attacks of FMF may be treated with non-steroidal anti-inflammatory drugs (NSAIDs). Early treatment with corticosteroids during an attack of FMF may also suppress the symptoms [14]. Based on this observation ocular inflammation should most likely also be treated with corticosteroids. Ophthalmologists are advised to liaise with the patient's physician when treating ocular inflammation in patients with FMF. Physicians who treat patients with FMF are advised to ask for ocular symptoms even if the patient is on treatment with colchicine.

It is now considered that patients with inflammatory diseases, such as systemic lupus erythematosus and rheumatoid arthritis, have an increased risk of atherosclerotic cardiovascular complications. Although it has been shown that ischemic heart disease prevalence was lower than expected in FMF patients, endothelial dysfunction and increased intima media thickness of carotid arteries have been demonstrated in patients with FMF [15,16]. The amaurosis fugax-like symptoms described by the patient in this report could thus, despite the young age of the patient, be linked to a thickening of the carotid artery wall. Doppler ultrasonography of the carotid arteries, however, was normal and the symptoms subsided after treatment of the anterior uveitis. The visual disturbances may have represented prodromals at the onset of her uveitis.

In conclusion, ophthalmologists are encouraged to report more cases of eye symptoms in FMF patients to elucidate the panorama of ocular pathologies associated with FMF. Objective verification of a recurrence of FMF attacks at first visit is recommended and appropriate samples should be taken. It is also worth noting that eye manifestations in FMF patients may occur during colchicine treatment.

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References

1. Ben-Chetrit E, Touitou I (2009) Familial mediterranean Fever in the world. Arthritis Rheum 61: 1447-1453.
2. Samuels J, Aksentijevich I, Torosyan Y, Centola M, Deng Z, et al. (1998) Familial Mediterranean fever at the millennium. Clinical spectrum, ancient mutations, and a survey of 100 American referrals to the National Institutes of Health. Medicine (Baltimore) 77: 268-297.
3. Ben-Chetrit E, Levy M (1998) Familial Mediterranean fever. Lancet 351: 659-64.
4. Goldfinger SE (1972) Colchicine for familial Mediterranean fever. N Engl J Med 287: 1302.
5. Akman A, Varan B, Akova YA, Aydin P (2001) Ocular involvement in siblings with familial mediterranean fever. J Pediatr Ophthalmol Strabismus 38: 114-116.
6. Altiparmak UE, Koklu B, Unlu,Yagci R, Bayrak R, et al. (2008) Macular involvement in secondary systemic amyloidosis. Eur J Ophthalmol 18: 459-461.
7. Berestizschevsky S, Weinberger D, Avisar I, Avisar R (2008) Episcleritis associated with familial Mediterranean fever. Isr Med Assoc J 10: 318-319.
8. Hirsh A, Huna R, Ashkenazi I, Bartov E, Blumenthal M (1990) Recurrent bilateral panuveitis and rhegmatogenous retinal detachment in a patient with familial Mediterranean fever. Am J Ophthalmol 110: 702-703.
9. Ozaltin F, Bakkaloglu A, Orhon M, Duzova A, Irkec M (2001) Bilateral uveitis in a 7-year-old patient with familial Mediterranean fever. An extremely rare complication. Clin Exp Rheumatol 19: S80-S81.
10. Scharf J, Meyer E, Zonis S (1985) Episcleritis associated with familial Mediterranean fever. Am J Ophthalmol 100: 337-339.
11. Yazici H, Pazarli H (1982) Eye involvement in a patient with familial Mediterranean fever. J Rheumatol 9: 644.
12. Gang N, Drenth JP, Langevitz P, Zemer D, Brezniak N et al. (1999) Activation of the cytokine network in familial Mediterranean fever. J Rheumatol 26: 890-897.
13. Knecht A, de Beer FC, Pras M (1985) Serum amyloid A protein in familial Mediterranean fever. Ann Intern Med 102: 71-72.
14. Erken E, Ozer HT, Bozkurt B, Gunesacar R, Erken EG, et al. (2008) Early suppression of familial Mediterranean fever attacks by single medium dose methyl-prednisolone infusion. Joint Bone Spine 75: 370-372.
15. Akdogan A, Calguneri M, Yavuz B, Arslan EB, Kalyoncu U, et al. (2006) Are familial Mediterranean fever (FMF) patients at increased risk for atherosclerosis? Impaired endothelial function and increased intima media thickness are found in FMF. J Am Coll Cardiol 48: 2351-2353.
16. Langevitz P, Livneh A, Neumann L, Buskila D, Shemer J, et al. (2001) Prevalence of ischemic heart disease in patients with familial Mediterranean fever. Isr Med Assoc J 3: 9-12.