EJR European Journal of Rheumatology

DOI:10.5152/eurjrheum.2022.22018

Image in Rheumatology

Frosted Branch Angiitis in a Patient with Familial Mediterranean Fever Diagnosis

Reşit Yıldırım[1], Mustafa Dinler[1], Ahmet Özer[2], Nazife Şule Yaşar Bilge[1], Timuçin Kaşifoğlu[1]

A 41-year-old female with a previous diagnosis of familial Mediterranean fever (FMF) was admitted due to a sudden-onset vision loss in the right eye during the third day of FMF attack. Past medical records revealed that she was non-compliant with her colchicine medication and therefore experienced 4 serositis attacks in the last 6 months. The systemic rheumatologic questionnaire was otherwise non-contributory. Ophthalmologic examination of the left eye demonstrated no pathology, whereas fundus evaluation of the right eye showed severe, white vascular sheaths extending from the optic disc to the periphery and pre- and intra-retinal hemorrhages in all retinal quadrants (Figure 1). Dilated and tortuous veins were detected in fluorescent angiography imaging (Figure 2). Based on these findings, a diagnosis of frosted branch angiitis (FBA) was made. Laboratory investigations were unremarkable except for mild elevations of acute phase parameters. Serologic studies for antinuclear antibody, anticardiolipin antibodies, and antineutrophil cytoplasmic antibodies (ANCA) were negative. Possible infectious agents were also excluded, and extensive radiographic evaluation of the central nervous system was non-revealing. After excluding all other possibilities, a diagnosis of FBA possibly related to FMF was considered and a combination of methylprednisolone (1 mg/kg/day) with tapering doses, azathioprine (2.5 mg/kg/day) and colchicine (0.5 mg, 3 times daily), was initiated. The visual complaints in the right eye completely resolved at the end of the third month. In the fourth month of follow-up, she manifested with sudden-onset vision loss in the same eye during a serositis attack. Colchicine dosage was escalated to 4 times a day with concomitant moderate steroid administration. She is currently on colchicine and azathioprine without any FMF attack and relapses of FBA in the last 6 months

Familial Mediterranean fever is the prototype of periodic monogenic autoinflammatory diseases typically manifested by recurrent attacks of peritonitis, fever, erysipelas-like rash, and arthritis.[1] Ocular involvement is a well-defined clinical feature of systemic autoimmune diseases, especially in Behçet's syndrome (BS), spondyloarthropathy, ANCA-associated vasculitis, systemic lupus erythematosus, and rheumatoid arthritis, whereas eye disease is quite uncommon in FMF.[2]

Frosted branch angiitis is a type of retinal vasculitis characterized by the sheathing of vascular structures which is a unique diagnostic appearance.[3] The disease is generally considered idiopathic, but its association

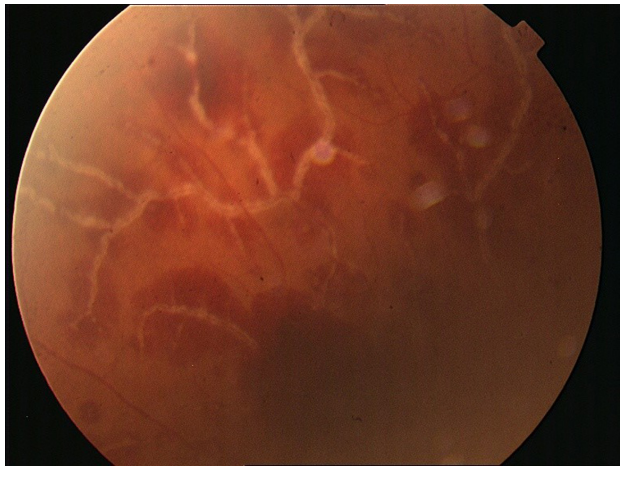

Figure 1. Fundus evaluation of the right eye revealed severe and white vascular sheaths extending from the optic disc to the periphery and pre- and intra-retinal hemorrhages in all retinal quadrants.

ORCID iDs of the authors:
R.Y. 0000-0003-4040-0212;
M.D. 0000-0002-8133-8278;
A.Ö. 0000-0002-8509-6982;
N.Ş.Y.B. 0000-0002-0783-1072;
T.K. 0000-0003-2544-8648.

Cite this article as: Yıldırım R, Dinler M, Özer A, Yaşar Bilge NŞ, Kaşifoğlu T. Frosted branch angiitis in a patient with familial mediterranean fever diagnosis. *Eur J Rheumatol*. 2023;10(1): 47-48.

[1] Division of Rheumatology, Department of Internal Medicine, Osmangazi University School of Medicine, Eskişehir, Turkey
[2] Division of Ophthalmology, Osmangazi University School of Medicine, Eskişehir, Turkey

Corresponding author:
Reşit Yıldırım
E-mail: celeng18@gmail.com

Received: March 18, 2022
Accepted: June 8, 2022
Publication Date: July 29, 2022

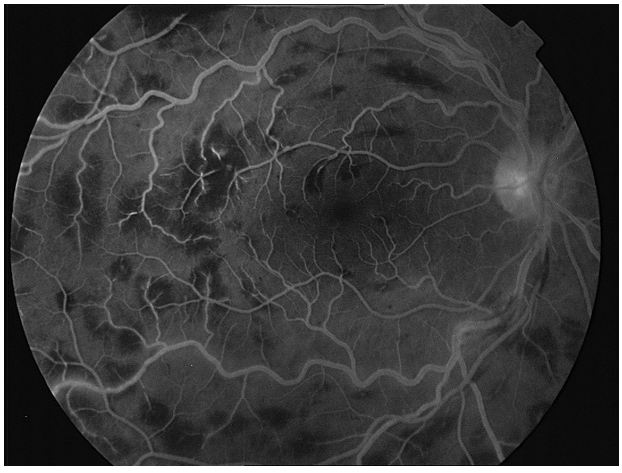

Figure 2. Dilated and tortuous veins were detected in fluorescent angiography.

with autoimmune diseases has been increasingly reported in the literature, particularly with granulomatosis with polyangiitis, FMF, and BS.[4] It is well described that some vasculitis disorders including immunoglobulin A vasculitis, BS, Takayasu arteritis, and polyarteritis nodosa might accompany with FMF, and all these clinical conditions may involve the eye.[5] Therefore, exclusion of all these possibilities is crucial before establishing a relationship between FMF and FBA. The association between FBA and FMF has been recently illustrated in a case series by Mansour et al.[4] In this report, a majority of FBA cases are seen in those with insufficient disease control. The authors also observed that effective control of inflammation in FMF with colchicine, especially in terms of normalizing C-reactive proteinP, is of great importance to control and prevent retinal vasculitis.[4]

Informed Consent: The informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - R.Y., M.D.; Design - R.Y., M.D.; Supervision - R.T., A.O., N.Ş.Y.B., T.K; Materials - R.Y., M.D., N.Ş.Y.B.; Data Collection and/or Processing - R.Y., M.D., N.Ş.Y.B.; Analysis and/or Interpretation - R.Y., A.O., N.Ş.Y.B., T.K.; Literature Review - R.Y., T.K.; Writing - R.Y., N.Ş.Y.B., T.K.; Critical Review - R.Y., A.O., N.Ş.Y.B., T.K.

Declaration of Interests: The authors have no conflicts of interest to declare.

Funding: The authors declared that this study has received no financial support.

References

1. Korkmaz C, Cansu DU, Cansu GB. Familial Mediterranean fever: the molecular pathways from stress exposure to attacks. *Rheumatol (Oxf Engl)*. 2020;59(12):3611-3621. [CrossRef]
2. Generali E, Cantarini L, Selmi C. Ocular involvement in systemic autoimmune diseases. *Clin Rev Allergy Immunol*. 2015;49(3):263-270. [CrossRef]
3. Walker S, Iguchi A, Jones NP. Frosted branch angiitis: a review. *Eye (Lond)*. 2004;18(5):527-533. [CrossRef]
4. Mansour HA, Ozdal PÇ, Kadayifcilar S, et al. Familial Mediterranean fever associated frosted branch angiitis, retinal vasculitis and vascular occlusion. *Eye (Lond)*. 2021. [CrossRef]
5. Abbara S, Grateau G, Ducharme-Bénard S, Saadoun D, Georgin-Lavialle S. Association of vasculitis and familial mediterranean fever. *Front Immunol*. 2019;10:763. [CrossRef]