Bilateral posterior uveitis in 3,5-year-old boy with presumed Behcet disease

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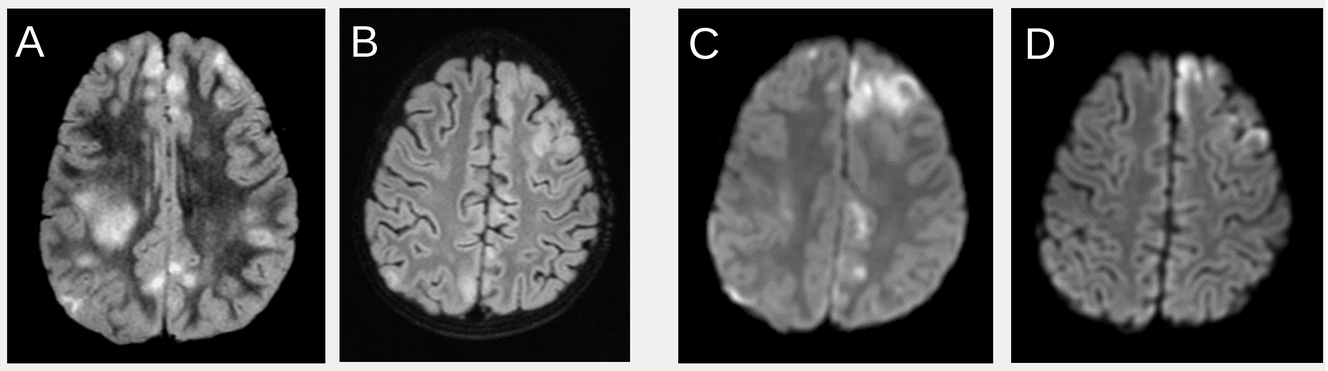
Introduction

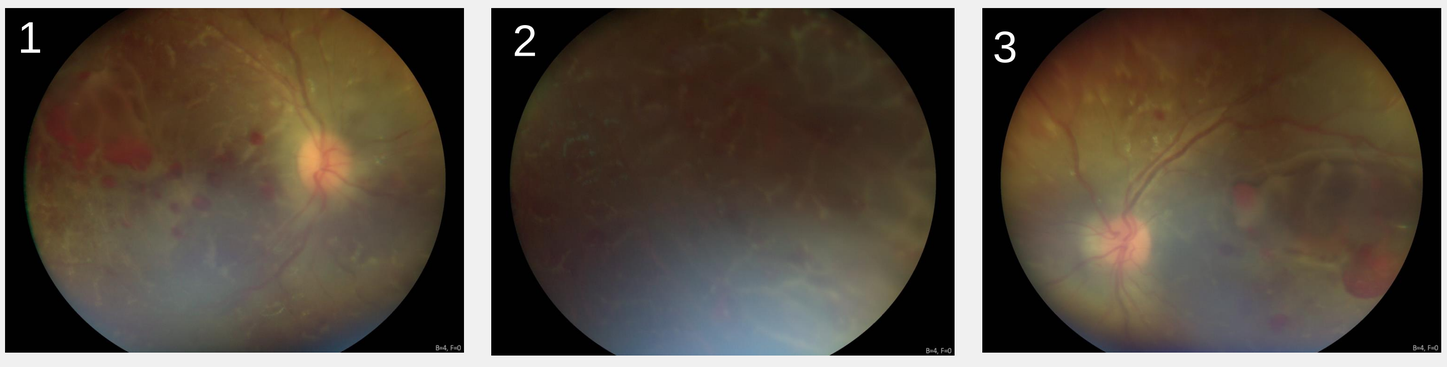
Behcet disease is a multisystemic vasculitis affecting vessels of all sizes but especially veins. Onset in childhood varies from 4-24% depending on the population's prevalence (territory and ethnicity). Mean age at onset varies from 4,9 to 12,3 years. We present a rare case of Bechet disease regarding not only severity and extend of retinal vasculitis but even age group. The ability to image the fundus with a non-contact handheld camera gave us the opportunity to visualize an impressive panvasculitis.

Our case

Previously healthy 3,5-year-old boy from the Middle East presents with fever, sore throat, abdominal pain and vomiting, first treated with antibiotics. Rapid deterioration of general status gives initially suspicion of Kawasaki disease therefore treated with aspirin, cortisone and immunoglobulin. He develops neurological symptoms with left side paresis, loss of speech and deteriorating consciousness and is intubated. Even heart valve dysfunction and pericarditis, oral ulcers are noted. Imaging reveals multiple ischemic and hemorrhagic CNS lesions as in vasculitis. High suspicion of systemic vasculitis is confirmed by fundus examination.



(A) T2 AX FLAIR: fresh ischemic lesions in cortex and white matter, less after one week (B)
(C) DWI: diffuse cytotoxic oedema, less after one week (D)



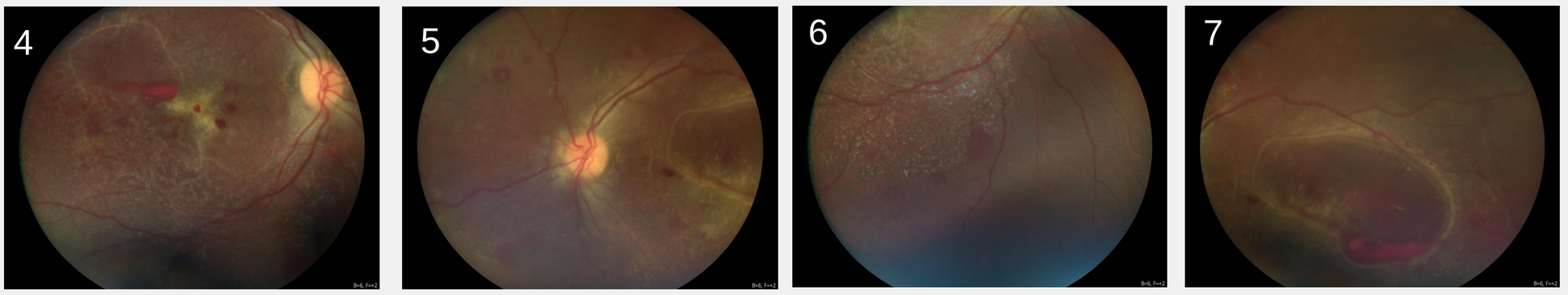
On presentation: 1 RE posterior pole, 2 RE periphery, 3 LE posterior pole

Eye examination in the ICU

facial palsy with low blinking frequency

esotropia due to bilateral abduction palsy

panuveitis: bilateral cells in anterior chamber and vitreous, diffuse chorioretinopathy with hemorrhages, "frozen branch" vasculitis and serous detachment



One week later: 4 RE posterior pole, 5 LE optic nerve, 6 RE inferior temporal, 7 LE macula and temporal

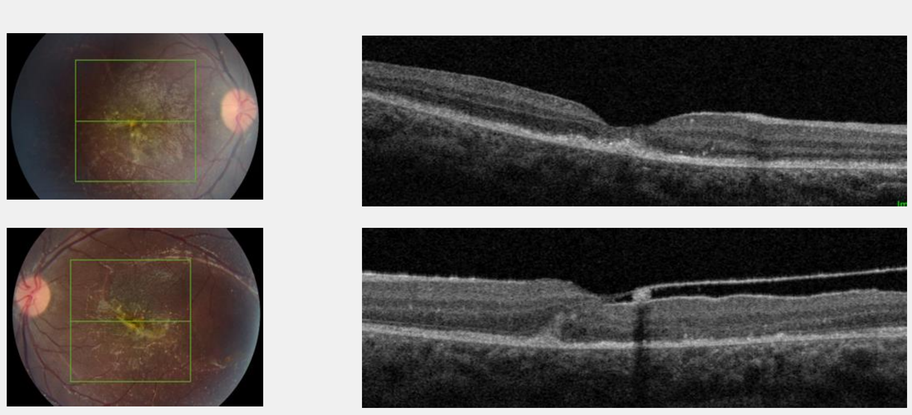
Fundus pictures were obtained with handheld fundus camera in the ICU

Results

Ocular findings were crucial in the diagnosis as the extensive bilateral retinal vasculitis confirmed aggressive systemic vasculitis due to Behcet disease. Treatment with systemic cortisone and anti-TNFa was successful and the retinal vasculitis regressed gradually.

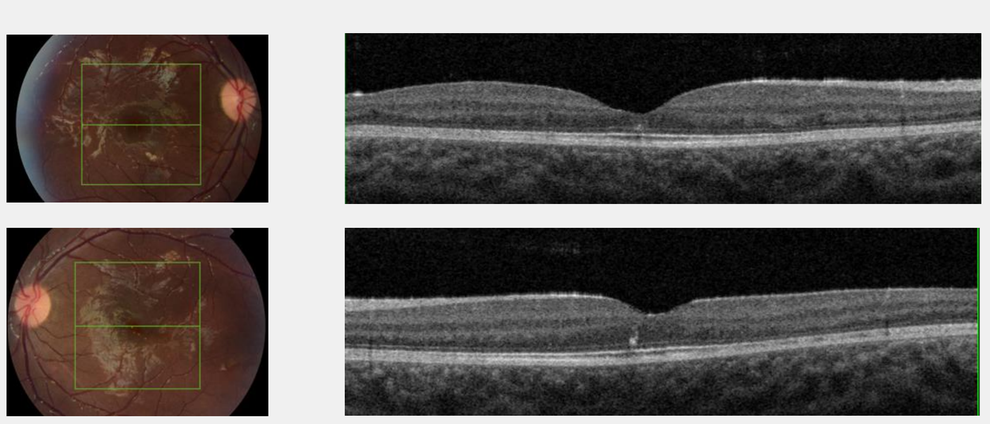
At 2,5 years of follow-up the patient is in remission, still treated with oral prednisolone, Methotrexate and Adalimumab.

The best corrected visual acuity is 0,5/0,7 with residual exudates and RPE changes seen on fundus examination and OCT.



Left: Fundus and OCT macula pictures shortly after discharge from the ICU

Right: Fundus and OCT macula pictures at 2,5 years follow-up



Take home message

Though Behcet disease is rare in Europe, especially in very young children as in our case, the clinical suspicion should be high. Ophthalmological examination should be considered early. We stress the importance of photo documentation with a non-contact handheld camera for both diagnosis and follow-up.

Literature

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