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Acute idiopathic multifocal hemorrhagic retinal vasculitis in a child: aggressive early treatment to successfully prevent bilateralization

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ABSTRACT

Acute multifocal hemorrhagic retinal vasculitis (AMHRV) is a rare condition that is typically described usually in adults. Its aetiology remains elusive, and a diagnosis should only be made after ruling out other possible causes that are consistent with the clinical presentation. Due to the limited literature available, most of which is based on single case reports, the optimal therapeutic regimen for AMHRV remains unclear. An appropriate, aggressive early treatment is crucial in order to prevent bilateralization of the disease or long-term complications. In this report, we present the second case of AMHRV in pediatric population.

KEY WORDS: vasculitis; acute multifocal hemorrhagic retinal vasculitis; pediatric

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INTRODUCTION

Acute multifocal hemorrhagic retinal vasculitis (AMHRV) is a rare condition with an unknown etiology. It predominantly affects healthy immunocompetent adults, mostly men, and is characterized by bilateral but asymmetric involvement. The condition presents with sudden visual loss accompanied by characteristic fundoscopic findings, all without concomitant systemic symptoms [1, 2].

The clinical hallmark of AMHRV resides in its distinctive fundoscopic manifestations, characterized by vitreous inflammation, retinal vasculitis, retinal hemorrhage predominantly in the posterior segment, and papillitis. Diagnosis of AMHRV is fundamentally clinical and can only be confirmed after discarding systemic etiologies or other entities with similar features [1–3]. Among these entities, Behçet's disease, sarcoidosis, ocular syphilis, toxoplasmosis, viral infection and Eales' disease should be excluded [3]. Long-term complications of AMHRV include epiretinal membrane, vitreous hemorrhage, neovascularization, iris rubeosis, or neovascular glaucoma [4].

Due to the low incidence of AMHRV and the limited literature available, no standardized treatment exists. Noteworthy reported therapeutic approaches include oral, intravenous, or intravitreal anti-viral treatments, systemic corticosteroids, vitrectomy, photocoagulation for retinal ischemia, and immunomodulators such as rituximab, methotrexate or cyclophosphamide [1–4].

To the best of our knowledge, this is the second reported case of AMHRV in the pediatric demographic [5].

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CASE REPORT

An 11-year-old Caucasian boy, devoid of any noteworthy medical history, presented to the emergency department of our hospital with a chief complaint of sudden painless visual impairment in the right eye (RE) over the course of the preceding 24 hours. Notably, he reported the absence of any accompanying symptoms.

On examination, his best corrected visual acuity (BCVA) was hand motion (HM) in the RE and 20/25 (0.10 LogMar) in the left eye (LE). Dyschromatopsia and relative afferent pupillary defect were present in the RE. Fundoscopy showed perivasculitis with intraretinal hemorrhages following the course of the vessels and severe optic disc edema in the RE (Fig. 1A). Mild optic disc edema, without retinal nor vascular abnormalities, was present in the LE (Fig. 1B). Slit-lamp examination was normal, as there was no anterior chamber or vitreous cell inflammation. Intraocular pressure was within normal limits in both eyes.

Macular optical coherence tomography (OCT) revealed retinal edema in the RE (Fig. 2A) and normal findings in LE (Fig. 2D). Optic nerve head OCT confirmed the bilateral optic disc edema. Fluorescein angiography showed vasculitis with blocked fluorescence in areas of intraretinal hemorrhages ret,inal ischemia in the RE, and leakage from the optic nerve head in both eyes.

He was alert and oriented with no motor or sensory deficits. The clinical examination did not reveal the presence of concomitant symptoms or any indicative focal neurological deficits. In light of the presented clinical scenario, the differential diagnosis encompassed several potential etiologies, including but not limited to intracranial neoplasms, hematologic disorders, and the possibility of inflammatory papillophlebitis arising as a consequence of either an infectious process or an autoimmune alteration.

Subsequently, the patient was admitted for an extensive workup, encompassing a comprehensive battery of assessments. This workup included a series of

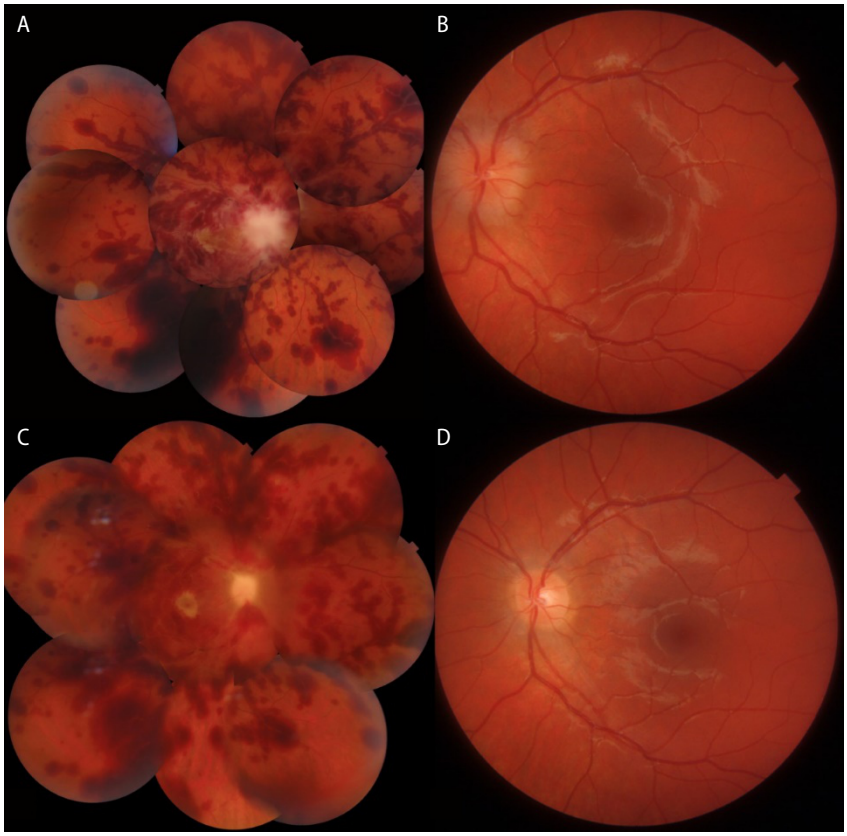

FIGURE 1. Fundoscopy findings. **A.** Right eye (RE) at presentation, fundoscopy showed periphlebitis with intraretinal hemorrhages following the course of the vessels and severe optic disc edema; **B.** Left eye (LE) at presentation, fundoscopy showed mild optic disc edema without retinal or vascular abnormalities; **C.** The first months after the onset of the disease, haemorrhages progressively decreased in RE; **D.** Optic disc edema progressively resolved in LE

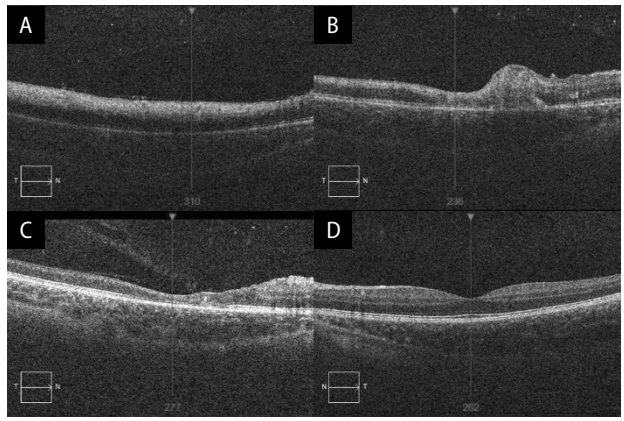

FIGURE 2. Macular optical coherence tomography (OCT) imaging. **A.** At presentation, macular OCT of the right eye (RE) showed diffuse retinal edema of the inner layer and loss of the fovea contour; **B.** Two weeks after, macular OCT showed retinal atrophy and traces of intraretinal haemorrhages in RE; **C.** One year after, macular OCT showed severe retinal atrophy in RE; **D.** Macular OCT showed normal findings during all the follow-up in LE

blood tests, imaging studies consisting of magnetic resonance imaging of the spine and brain, and a chest X-ray). A lumbar puncture (LP) and anterior chamber paracentesis were also undertaken as part of the diagnostic evaluation. The battery of laboratory investigations comprised a thorough assessment encompassing a complete blood count, flow cytometry, a metabolic profile, and exhaustive serological screenings for a spectrum of potential etiological factors, including syphilis, Bartonella, rickettsiae, human immunodeficiency virus, varicella-zoster virus, herpes simplex virus I/II, hepatitis, cytomegalovirus, Epstein-Barr virus, and toxoplasmosis. In parallel, specialized assessments were carried out, comprising evaluations for anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, anti-DNA antibodies, rheumatoid arthritis factor, human leukocyte antigen (HLA) B-27, HLA B-51, complement fractions, C-reactive protein, erythrocyte sedimentation rate, angiotensin-converting enzyme, calcium levels, serum protein electrophoresis, and cryoglobulins.

Empirical treatment with daily 30 mg/kg of methylprednisolone, 250 mg of acetazolamide, 1500 mg of bicarbonate, and Boi-K was initiated even before an established diagnosis. Within ten days, results from complementary tests were available, and results were unremarkable, ruling out any other underlying condition that could have caused the presenting features and symptoms. The patient was ultimately diagnosed with AMHRV. After the initial high dose of corticosteroid, he was discharged with ambulatory follow-up, which included monitoring with fundus photographs (Fig. 1C) and OCT (Fig. 2BC). During follow-up, optic disc edema of LE resolved (Fig. 1D), and its BCVA improved to 20/20 (0.0 LogMar). Retinal ischemia was detected in RE in a subsequent follow-up 3 months later (Fig. 3A). Hence, retinal laser photocoagulation was performed to prevent complications such as neovascularization or vitreous hemorrhage. However, despite photocoagulation, a 6 months later, vitreous hemorrhage occurred in RE (Fig. 3B). Retinal photocoagulation was repeated and supplemented.

As of two years later, he has not experienced any complications related to AMHRV. His visual acuity has improved to 20/100 in the RE and remains stable, 20/20 (0.0 LogMar) in the LE.

DISCUSSION

AMHRV is an uncommon condition, usually described in adults, whose etiology and treatment remain unclear. Early aggressive treatment is crucial to prevent bilateralization or long-term complications. In the case presented, corticosteroids were initiated even without a confirmed diagnosis.

Among all the different treatment options described in the literature, including steroids, antiviral drugs, and immunomodulatory drugs, in our case, only methylprednisolone was administered. While antiviral treatment has usually been ineffective, early aggressive immunosuppressive therapy has been the most effective treatment [1, 4].

Photocoagulation of areas with retinal ischemia is highly recommended to prevent potential complications [2]. Despite having undergone photocoagulation, our patient experienced vitreous hemorrhage, which required additional reinforcement.

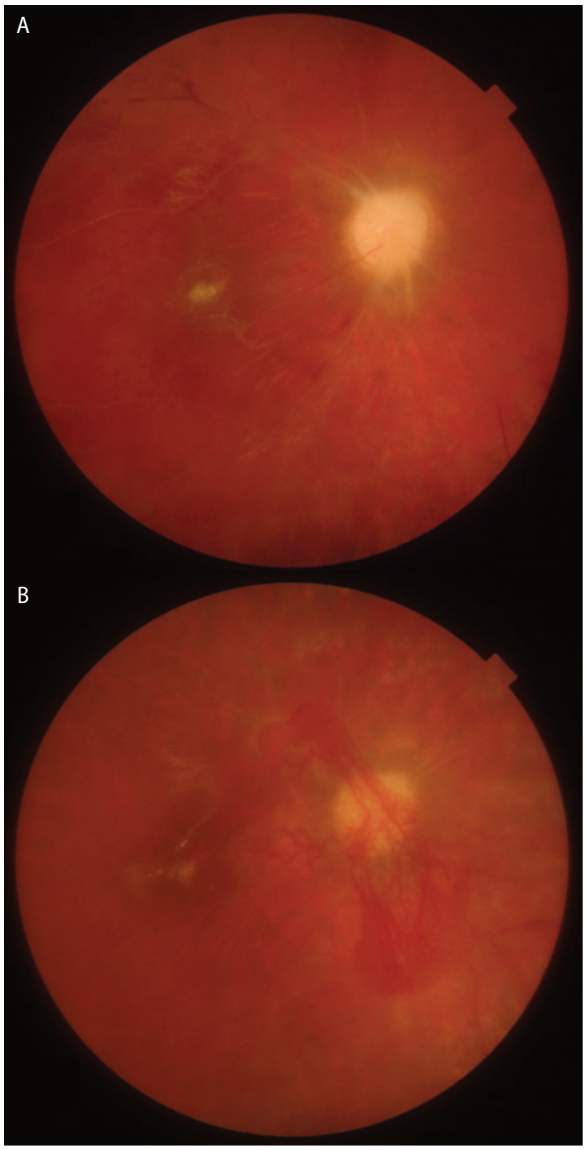

FIGURE 3. A. Fundoscopy with retinal ischemia was detected in right eye (RE) in subsequent follow-up; **B.** Vitreous hemorrhage occurred despite retinal laser photocoagulation in RE

In the previous case reported in a child [5], RE was followed by LE involvement 4 months later. Bilateral involvement was observed in our case at presentation. Notwithstanding, due to the asymmetric nature of the disease and early administration of a high dose of intravenous methylprednisolone, visual function in the left eye was preserved.

CONCLUSIONS

The presented case is the second reported case of AMHRV in a child in the literature. Multidisciplinary management was necessary for a prompt diagnosis and successful treatment. While there is no consensus on the most appropriate treatment regimen, early and aggressive immunosuppressive therapy, including megadose intravenous corticosteroids with or without other immunomodulators, appears to be the most effective approach.

LITERATURE SEARCH

To the best of our knowledge, this is the second case of AMHRV in the pediatric population available in the literature. The search term was "acute multifocal hemorrhagic retinal vasculitis," and the database used was PubMed, which was accessed on September 15th, 2023. The results included five articles, of which only one reported this entity in a child [5].

Ethics statement

Written informed consent was obtained from the patient included in this case report for the publication of their medical information, images, or any other relevant data. Every effort has been made to ensure the anonymity of the patient in this case report. Identifying details, including names, initials, or any other information that could reveal the identity of the patient, have been omitted.

Author contributions

The authors of this study have collectively contributed to all aspects of the research, and each author has participated significantly in the work. All authors have reviewed and approved the final version of the manuscript. They share responsibility for the study's integrity, accuracy, and adherence to ethical guidelines.

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Conflict of interest

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