

## Bilateral Optic Disc Edema and Retinal Vascular Tortuosity in Kawasaki Disease: Rare Manifestations with Good Outcome

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### Abstract

**Introduction:** Optic disc edema and/or vascular tortuosity associated with Kawasaki disease (KD) have been rarely described. We, therefore, reported four cases with optic disc swelling and retinal vascular tortuosity associated with KD identified in early fundoscopy exam in children with remarkable conjunctival injection.

**Case Reports:** The median age at KD diagnosis was 3.25 years (range 2.42 - 5.5). All patients fulfilled the EULAR/PreS criteria for KD, with bilateral bulbar nonexudative conjunctivitis. Ophthalmological examination (Slit-lamp biomicroscopy, ophthalmoscopy and visual acuity) revealed: four patients had bilateral optic disc swelling and two presented bilateral retinal vascular tortuosity. All KD patients received intravenous immunoglobulin and acetylsalicylic acid, with total improvement of ophthalmological abnormalities in up to two months.

**Discussion:** We observed herein posterior eye segment involvement resulting in bilateral optic disc swelling and retinal tortuosity in KD patients with marked conjunctivitis during acute febrile phase. Prompt KD treatment suggested good outcome for these ophthalmological abnormalities.

**Keywords:** Kawasaki Disease; Ocular; Eye; Optic Disc Edema and Children

### Introduction

Kawasaki disease (KD) is a primary vasculitis that occurs in early childhood, predominantly in children younger than 5 years-old [1-3].

KD diagnosis is essentially clinical, based on classification criteria including persisting fever for at least five days and four of the five subsequent finds: changes in the lips or oral cavity, changes in the peripheral extremities or perineal area, polymorphous exanthema, cervical lymphadenopathy and bilateral conjunctival injection [4,5].

Nonexudative conjunctivitis is common and has been reported in up to 90% of KD [6,7]. Other ocular findings such as anterior/posterior uveitis, keratitis, vitreous opacities, subconjunctival hemorrhage can be identified in the acute phase of the disease by ophthalmological

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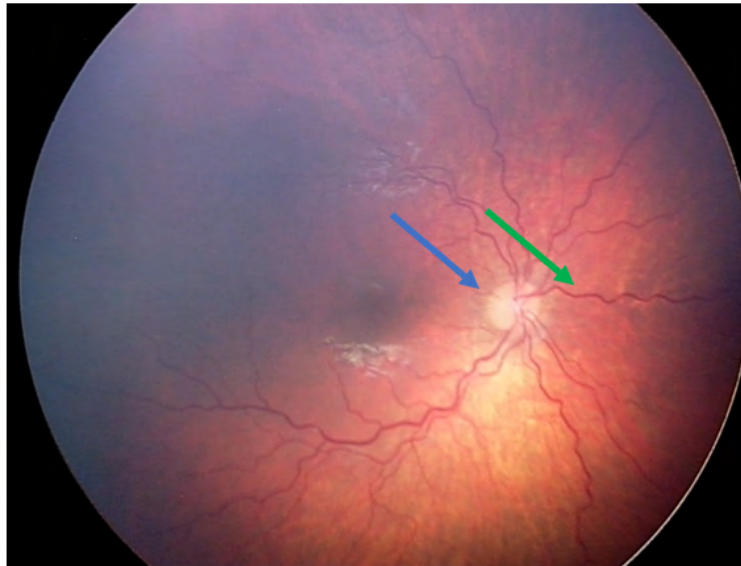
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evaluation [4,5,8-11]. However, they are not included in the endorsed consensus criteria of European League Against Rheumatism (EULAR)/Paediatric Rheumatology European Society (PReS) [5].

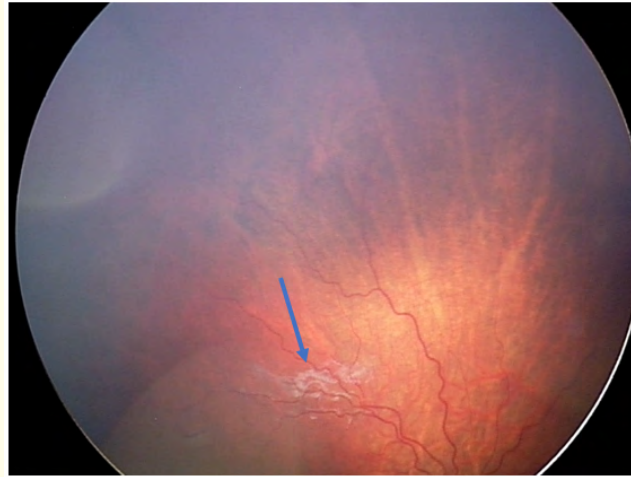
Of note, optic disc swelling and/or retinal vascular tortuosity associated with KD have been rarely described in case reports [10,12-14] or case series [6], particularly in Asian and European KD populations. We, therefore, reported four cases describing optic disc swelling and retinal vascular tortuosity associated with KD identified in early funduscopy exam in children with remarkable conjunctival injection.

### Case Reports

**Case 1:** A 2 years and 9 months old male had persistent fever for 5 days, prominent bilateral bulbar nonexudative conjunctivitis, lip erythema, cervical lymphadenopathy > 1.5 cm, edema in peripheral extremities and diffused exanthema, and fulfilled the EULAR/PReS criteria for KD [5]. In addition, he had concomitantly irritability, persistent vomiting, anorexia and weight loss. His laboratorial exams showed: hemoglobin 11.6 g/dL, hematocrit 33%, white blood cell count 14,300/mm<sup>3</sup> (67% neutrophils, 17% lymphocytes, 13% monocytes and 3% eosinophils), platelets 401,000/mm<sup>3</sup>, erythrocyte sedimentation rate (ESR) 89 mm/1<sup>st</sup> hour (reference value < 20 mm/1<sup>st</sup> hour), C-reactive protein (CRP) 4.7 mg/L (reference value < 1 mg/L), urea 7 mg/dL (reference value 10 - 50 mg/dL), creatinine 0.32 mg/dL (reference value 0.3 - 0.7 mg/dL), albumin 3.1 g/dL (reference value 3.5 - 5.2 g/dL), aspartate aminotransferase 19 U/L (reference value < 40 U/L) and alanine aminotransferase 14 U/L (reference value < 41 U/L). Urinalysis was normal. No dilatation of coronary arteries was evidenced in echocardiography exam. Abdominal ultrasound was normal. He received 2 g/kg/dose of intravenous immunoglobulin (IVIG) and aspirin 60 mg/kg/day. His symptoms promptly improved. Ophthalmological examination (Slit-lamp biomicroscopy, ophthalmoscopy and visual acuity) showed bilateral optic disc swelling with retinal vascular tortuosity (Figure 1), and exudate (Figure 2) in the sixth day, without visual acuity impairment. At that moment, neurological evaluation and brain magnetic resonance imaging were normal. Ophthalmological exam was repeated after 31 days, with resolution of ophthalmological abnormalities.



**Figure 1:** Ophthalmological examination showed bilateral optic disc swelling (blue arrow) with retinal vascular tortuosity (green arrow) in case 1.



**Figure 2:** Ophthalmological examination showed exudate (blue arrow) in case 1.

**Case 2:** A 5 years and 6 months old female with a clinical history of fever for 7 days, cervical lymphadenopathy > 1.5 cm, pruritic exanthema on trunk and extremities, bilateral bulbar injection, cracked lips and peripheral desquamation in hands, fulfilling the EULAR/PReS criteria for KD [5]. She also presented irritability, abdominal pain, vomiting and myalgia. At that moment, hemoglobin was 11.5 g/dL, hematocrit 39.6%, white blood cell count 14,300/mm<sup>3</sup> (62% neutrophils, 30% lymphocytes, 6% monocytes and 2% eosinophils), platelets 448,000/mm<sup>3</sup>, ESR 120 mm/1<sup>st</sup> hour, CRP 7 mg/L, urea 22 mg/dL, creatinine 0.4 mg/dL, albumin 3.1 g/dL, aspartate aminotransferase 28 U/L, alanine aminotransferase 59 U/L and a normal urinalysis. No coronary artery abnormalities were observed. She was treated with 2 g/kg/dose of IVIG and aspirin 60 mg/kg/day. She was afebrile after 48 hours. Ophthalmological examination (Slit-lamp biomicroscopy, ophthalmoscopy and visual acuity) showed bilateral optic disc swelling without visual impairment. Optic disc edema remitted after one month.

**Case 3:** A 3 years and 9 months old male with 7 days of fever, conjunctivitis, strawberry tongue and lip erythema, cervical pain (cervical ultrasound confirmed 2.5 cm right anterior lymphadenopathy), perineal erythema with penis edema and desquamation in hands, fulfilling the EULAR/PReS criteria for KD [5]. His laboratorial exams showed: hemoglobin 9.2 g/dL, hematocrit 26.3%, white blood cell count 13,300/mm<sup>3</sup> (87% neutrophils, 9% lymphocytes, 2% monocytes and 2% eosinophils), platelets 112,000/mm<sup>3</sup>, ESR 99 mm/1<sup>st</sup> hour, CRP 27.9 mg/L, urea 65 mg/dL, creatinine 0.5 mg/dL, albumin 3.4 g/dL, aspartate aminotransferase 40 U/L and alanine aminotransferase 17 U/L. Urinalysis demonstrated 13 white blood cells/high power field and proteinuria. Echocardiogram showed dilatations in anterior descending (Z score +4.76) and marginal branch of right coronary (Z score +3.57) arteries. He was treated with 2 g/kg/dose of IVIG, oral prednisone 2 mg/kg/day and aspirin 60 mg/kg/day. Fever and clinical manifestations improved after 48 hours. Ophthalmological examination (Slit-lamp biomicroscopy, ophthalmoscopy and visual acuity) evidenced bilateral optic disc swelling and vascular tortuosity, without visual impairment. Total remission of coronary abnormalities was observed after one month, as well as funduscopy exam.

**Case 4:** A 2 years and 5 months old male presenting 5 days of fever, injected pharyngitis, exanthema on the face and trunk, edema in extremities and prominent conjunctival hyperemia, fulfilling the EULAR/PReS criteria for KD [5]. He had bilateral limp, tenderness and pain on motion in hip joints. Hips ultrasound confirmed bilateral hip joint arthritis. His laboratorial exams showed: hemoglobin 11.7 g/dL, hematocrit 33.8%, white blood cell count 9,700/mm<sup>3</sup> (53% neutrophils, 37% lymphocytes, 8% monocytes and 2% eosinophils), platelets

269,000/mm<sup>3</sup>, ESR 64 mm/1<sup>st</sup> hour, CRP 85 mg/L, urea 12 mg/dL, creatinine 0.28 mg/dL, albumin 3.0 g/dL, aspartate aminotransferase 31 U/L and alanine aminotransferase 28 U/L and normal urinalysis. Coronary dilatations arteries were not detected. He received 2 g/kg/dose of IVIG, aspirin 60 mg/kg/day and improved within 48 hours. Ophthalmological examination (Slit-lamp biomicroscopy, ophthalmoscopy and visual acuity) revealed bilateral optic disc swelling and retinal vascular tortuosity without visual impairment. Remission of funduscopy abnormalities were observed two months later.

### Discussion

We evidenced herein posterior eye segment involvement resulting in bilateral optic disc edema and retinal tortuosity, of KD in patients with evident conjunctivitis during acute febrile phase. Prompt KD treatment suggested good outcome for these ophthalmological abnormalities.

Ocular abnormalities are a common manifestation at KD diagnosis, particularly bilateral bulbar nonexudative conjunctivitis [6,7]. The severity of this ocular manifestation in our KD patients indicated the need of ophthalmological examination. One limitation of the present study was the lack of tonometry analysis during ophthalmological examination, due to patients irritability at KD diagnosis.

Optic disc edema and retinal tortuosity were early manifestations of KD observed in the present study. Posterior segment involvement in KD was rarely reported and included macular and disc edema, retinal ischemia, vitritis and retinal vasculitis [6,14]. Moreover, acute iridocyclitis was not observed in any of our patients. These manifestations occur during the acute disease phase and were reported from 11% to 29% [9,11].

Papilledema is a cause of optic disc edema, precisely associated with intracranial hypertension [15]. This finding was excluded in the first case at KD diagnosis after neurological and brain magnetic resonance evaluations. The absence of neurological signs and symptoms during follow-up and improvement of ocular manifestations did not suggest this complication in the other three KD patients of the present study.

Regarding outcome, all patients improved the ophthalmological manifestations after prompt KD cornerstone treatment. This anti-inflammatory regimen may contribute to improve aforementioned eye manifestations. Therefore, we suggest routinely ophthalmologic evaluation during the acute phase in KD patients, particularly in those with remarkable conjunctivitis.

### Conclusion

In conclusion, bilateral optic disc edema and retinal tortuosity were rarely observed in KD patients with marked conjunctivitis during the acute febrile phase. Prompt KD treatment suggested good outcome for these ophthalmological abnormalities.

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### Conflicts of Interest Statement

The authors have no conflict of interest that is directly relevant to the content of this manuscript.

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