Int Ophthalmol (2019) 39:451–453
https://doi.org/10.1007/s10792-017-0807-9

CASE REPORT

Retinal vasculitis associated with cutaneous leukocytoclastic vasculitis

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Received: 28 July 2017 / Accepted: 11 December 2017 / Published online: 18 December 2017

Abstract
Introduction To report a case of retinal vasculitis associated with cutaneous leukocytoclastic vasculitis.
Methods Retrospective chart review.
Results A 28-year-old man, who initially presented with occlusive retinal vasculitis and vitreous hemorrhage in right eye that resolved with sectoral photocoagulation. Laboratory investigations for tuberculosis, sarcoidosis, syphilis and sickle cell disease were negative. Past history included recent diagnosis of *Enterobacter* epididymo-orchitis and multiple red nodules on skin of forearm. Fourteen months later, he developed active retinal vasculitis in right eye and recurrent nodules on forearm. Skin biopsy revealed neutrophilic infiltrates in and around dermal vessels with destruction of vessel walls leading to scattered neutrophils, lymphocytes and histiocytes between collagen bundles, suggestive of leukocytoclastic vasculitis. Both skin and ocular lesions resolved with oral corticosteroid and methotrexate therapy and did not recur over a six-year period.
Conclusion We have reported the first case of clinically manifest retinal vasculitis, associated with a common form of cutaneous vasculitis.

Keywords Retinal vasculitis · Leukocytoclastic · Cutaneous · Epididymo-orchitis

Leukocytoclastic vasculitis (LCV) is a common form of cutaneous vasculitis [1]. Ophthalmic manifestations of LCV have rarely been described [2, 3]. To our knowledge, clinically manifest retinal vasculitis associated with LCV has never been reported.

A 28-year-old male reported with floaters in right eye for 2 days. His best-corrected visual acuity (BCVA) and intra-ocular pressure (IOP) in both eyes were 20/20 and 12 mmHg, respectively. Right eye showed retinal neovascularization in supero-nasal quadrant associated with pre-retinal and vitreous hemorrhage. Sclerosed vessels were noted in infero-temporal fundus. There was no sign of active inflammation. Left eye was clinically normal. Past medical history was significant for red nodules on his right forearm 4 months ago and an episode of hematospermia 2 months ago that was diagnosed as epididymo-orchitis and revealed *Enterobacter* on semen culture. The skin lesions resolved spontaneously, while epididymo-orchitis resolved with specific antibiotic therapy. Our investigations for etiological diagnosis showed elevated erythrocyte

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sedimentation rate (ESR, 30 mm at 1 h) but normal blood picture, tuberculin test, Quantiferon TB Gold Test, chest X-ray, serum angiotensin converting enzyme, sickling test, syphilis serology and serum anti-nuclear antibodies. We diagnosed healed retinal vasculitis with vitreous hemorrhage and treated him with sectoral photocoagulation, following which the retinal neovascularization and vitreous hemorrhage resolved. He was followed up regularly.

Fourteen months later, he again reported vitreous floaters in right eye and red nodules on right forearm for 2 weeks. We noted BCVA of 20/20 and IOP 16 mmHg in right eye. Right fundus showed multiple foci of perivenular infiltrates along supero-temporal arcade and its branches (Fig. 1a, b). Few retinal hemorrhages were seen. Dermatologic consultation for skin lesions (Fig. 1c) led to diagnosis of LCV. This was based on histopathologic demonstration of neutrophilic infiltrates in and around dermal vessels with destruction of vessel walls leading to scattered neutrophils, lymphocytes and histiocytes between collagen bundles (Fig. 1d, e). Scattered fragmented nuclei and debris material were noted. There was no history of oral aphthous or genital ulcers. Subsequent tests revealed negative results for anti-neutrophil cytoplasmic antibodies (ANCA), renal function tests including urinalysis, rheumatoid factor, hepatitis B and C antigens, and anti-phospholipid antibodies. He was treated with oral deflazacort 48 mg per day. His skin lesions and retinal vasculitis resolved over a period of 4 weeks. Deflazacort was tapered over a period of 5 months. Concurrently, the patient was also started

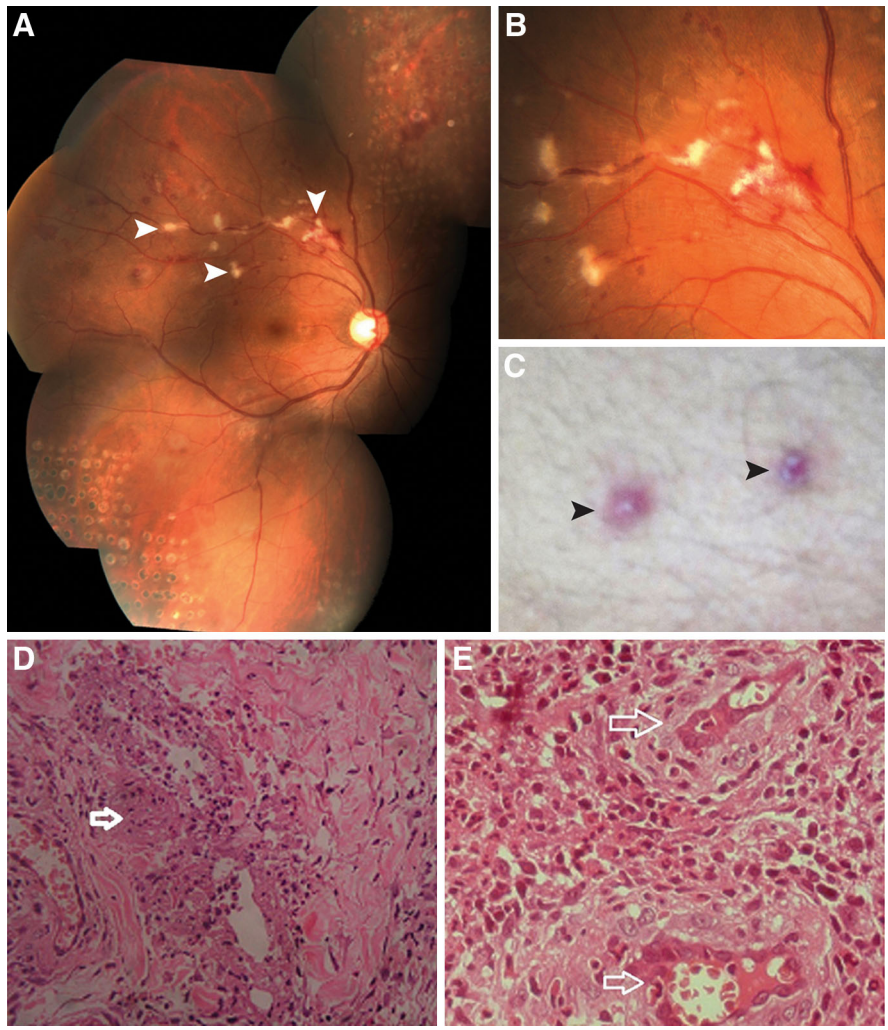

Fig. 1 **a**, **b** Right fundus photograph showing focal perivenular infiltrates (arrowheads) and retinal hemorrhages along the supero-temporal arcade and its tributaries. **c** Nodular lesions (arrowheads) on right forearm representing leukocytoclastic vasculitis. **d** Histological section of skin nodule showing small vessel (arrow) with fibrin in its lumen, fibrinoid changes of its wall along with neutrophils and debris in the wall and (E) vessels with plump endothelial cells and neutrophilic infiltration of the vessel wall and lumen (arrows). Fibrin deposits, polymorphs, lymphocytes and histiocytes are also seen between and within the collagen bundles

on oral methotrexate, reaching a maximum dose of 20 mg/week that was continued for a period of 18 months. There was no recurrence during 6 years of follow-up.

The term leukocytoclastic vasculitis is based on the histological characteristic of leukocytoclasia or apoptotic degeneration of granulocytic cell nucleus, seen in this condition. It is caused by deposition of large circulating immune complexes on vessel walls that attracts granulocytes that damage the vascular endothelium leading to extravasation of erythrocytes [1]. Common etiological factors for LCV include drugs, infections, collagenoses and malignant tumors. Besides the skin, several other organs like kidneys, joints, lungs, brain and gastrointestinal tract may be involved. Milder disease needs only supportive therapy, while more severe forms are treated with corticosteroids and occasionally colchicine, azathioprine, cyclophosphamide and even plasmapheresis [1].

Ophthalmic manifestations of LCV have been rarely reported. These include panuveitis, multifocal retinitis, marginal ulceration of cornea, chemosis, subconjunctival hemorrhage, episcleritis, pseudotumor cerebri and optic atrophy [2–5]. A PubMed search (keywords: leukocytoclastic vasculitis, ocular, retina) revealed that this is the first report of clinically evident retinal vasculitis associated with LCV. An earlier report described fluorescein angiographic features of peri-arteriolar leakage in association with multifocal retinitis and LCV though there was no clinical evidence of retinal vasculitis in that case [2]. Interestingly, the involvement of post-capillary venules in the retina in our patient matches the histological characteristics of skin lesions in LCV.

The temporal association of skin and ocular lesions with *Enterobacter* epididymo-orchitis suggests an etiological link between these conditions. This could have been substantiated if we had used direct immunofluorescence to identify specific antigen–antibody complexes in the skin biopsy. Hence, the role of drugs or an unidentified immunological condition could not be ruled out.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Author contributions SB contributed to collection and interpretation of data, drafting, revision and final approval of manuscript. RM helped in interpretation of data, revision and final approval of manuscript.

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