CASE REPORT

Bilateral multifocal posterior pole lesions in Reiter syndrome

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Professor Ahmad M Mansour,
ammansourmd@gmail.com**SUMMARY**

Reactive arthritis is associated with conjunctivitis or iritis. Rarely reactive arthritis is accompanied by permanent visual loss from macular infarction or foveal scarring. We present the case of a rheumatologist who had a sudden onset of skin lesions, arthritis of several joints and bilateral visual loss. Most of these manifestations resolved after a course of oral corticosteroids. However he was left with decreased vision in the left eye and multiple lesions in the fovea over a follow-up of 2 years.

BACKGROUND

Reiter syndrome identifies a triad of reactive arthritis of large joints, ocular inflammation in the form of conjunctivitis or iridocyclitis, and non-bacterial urethritis in men or cervicitis in women.^{1–18} Major ocular manifestations have included mild, symmetric and bilateral conjunctivitis with a mucopurulent discharge and a papillary or follicular reaction, and anterior mild non-granulomatous uveitis with fine-sized to medium-sized white keratic precipitates.^{1–10} Scleritis, episcleritis,³ keratitis,¹¹ disc oedema, retinal oedema, retinal vasculitis⁶ and retinal detachment^{1 2} have also been reported. We present the retinal findings in a rheumatologist with retinal manifestations from Reiter syndrome.

CASE PRESENTATION

This 50-year-old Iraqi rheumatologist noted sudden bilateral visual loss 2 years before presentation to our clinic. There was absence of signs of either conjunctivitis or uveitis. He was diagnosed as having multifocal choroiditis (figures 1–4). This coincided with painful swelling of shoulders, cervical spine, knees and ankles, as well as multiple reddish tiny skin lesions on the extremities along with an oral thrush. Chest radiograph was normal. Workup was negative for Behcet's disease, inflammatory bowel disease, ankylosing spondylitis, psoriatic arthritis and rheumatoid arthritis (negative RA latex and negative antinuclear antibodies (ANA) test). C reactive protein and erythrocyte sedimentation rate (65 mm/h) were elevated. Family history was positive for fibromyalgia in a brother. The clinical diagnosis was Reiter syndrome. Dermatological and rheumatological signs and symptoms disappeared after 2 weeks of oral corticosteroids.

Upon presentation 2 years later, best corrected visual acuity was 6/6 right eye and 6/15 left eye. The eyes were quiet with bilateral multiple posterior pole lesions at the level of the retinal pigment epithelium resembling drusen and sparing the fovea in the right eye (figures 5–10). Autofluorescence

Figure 1 Initial fundus photograph and intravenous fluorescein angiography transits of the right eye done in Iraq: hyperfluorescence of lesions in the posterior pole.

revealed more widespread disease of the retinal pigment epithelium in the left eye. Ocular coherence tomography revealed punched out lesions at the level of the retinal pigment epithelium-Bruch membrane (figure 9) and more diffuse atrophy of the outer retina in the left eye (figure 10).

Figure 2 Initial fundus photograph and intravenous fluorescein angiography transits of the left eye: hyperfluorescence of lesions in and around the fovea.

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Figure 3 Ocular coherence tomographic horizontal scan in the acute phase shows subretinal fluid in the right fovea.

INVESTIGATIONS

Purified protein derivative (PPD) skin test and repeat ANA blood test were negative. The patient tested positive for HLA-B51.

DIFFERENTIAL DIAGNOSIS

Systemic findings differential include: ankylosing spondylitis, inflammatory bowel disease, psoriatic arthritis, tuberculosis, sarcoidosis and syphilis. Ocular findings differential include acute multifocal pigment epitheliopathy, multifocal choroiditis, Stargardt disease, familial dominant drusen, age-related macular degeneration and pattern macular dystrophy.

DISCUSSION

Some infectious microorganisms appear to be the aetiological agents in reactive arthritis as large scale epidemics of dysentery have been linked to simultaneous onset of arthritis, urethritis and conjunctivitis.¹⁷ Three possible mechanisms have been

Figure 4 Ocular coherence tomographic horizontal scan in the acute phase shows subretinal fluid in the left fovea and left temporal peripapillary area.

Figure 5 Posterior pole of the right eye at the 2-year follow-up shows scattered tiny yellowish spots at the level of the retinal pigment epithelium.

proposed to explain the association of reactive arthritis with HLA-B27: (1) molecular mimicry; (2) HLA-B27 acts as a receptor for the inciting organism and (3) HLA-B27 is linked to a defective class I antigen-mediated cellular response.

The classic triad of the disease, namely urethritis, arthritis and conjunctivitis, is present in only one-third of the patients. Conjunctivitis appears in approximately 50% of patients and can be easily overlooked. Reactive arthritis is preferably viewed as a tetrad, with the addition of the mucocutaneous findings as well as psoriasis-like lesions such as of either circinate balanitis or keratoderma blennorrhagicum to the classic triad.¹⁴ Reactive arthritis has an important genetic component as it tends to cluster in certain families, and preferentially affects young white men, with HLA-B27 identified in around 75% of patients.¹⁶ Our patient was positive for HLA B-51 which is also associated with reactive arthritis.^{15 16}

Kim *et al*⁶ described a case of bilateral visual loss (6/60 right eye and 6/120 left eye) from macular infarct in a 53-year-old woman 3 days after presenting with fever, chills and bilateral knee pain. Conway *et al*⁷ reported a patient with uveitis that progressed to involve the posterior segment as two transient white retinal spots. After resolving, a retinal pigment epithelial defect persisted at the site of one of the lesions. Hovland *et al*¹⁹

Figure 6 Posterior pole of the left eye at the 2-year follow-up shows tiny yellowish spots inside the arcade and confluent large spots in the fovea.

Figure 7 Fundus autofluorescence mode of the right eye: multiple tiny hypofluorescent spots in the posterior pole (fovea, superior arcade and inferior arcade) with occasional hyperautofluorescent spots.

presented a patient in the third decade with juxtafoveal choroidal neovascularisation measuring 2.3 mm in diameter that was associated with Reiter's syndrome and pretreatment visual acuity of 6/60, received two sessions of photodynamic therapy 1 month apart, and was observed to have a stabilised lesion, and an improvement of visual acuity to 6/18 at the 6-month follow-up. Needham *et al*⁵ presented a patient with bilateral multifocal choroiditis. This 35-year-old man had bilaterally 6/4 visual acuity with hyperfluorescent scattered posterior pole lesions. The lesions were not visible by examination with stable vision at the 8-month follow-up. The lesions described in the current case could be the result of multifocal choroiditis yet they share also common features with lesions noted in punctate inner choroidopathy by the size of the lesions, presence of sub-retinal fluid and findings on both ocular coherence tomography²⁰ and autofluorescence patterns.²¹ Although visual acuity is usually preserved in most cases with Reiter syndrome, permanent visual loss rarely occurs as in the current case. Similarly,

Figure 8 Fundus autofluorescence mode of the left eye: confluent tiny hypofluorescent spots aggregating in the foveal region with occasional hyperautofluorescent spots.

Figure 9 High definition spectral domain ocular coherence tomography of the right eye shows the area of disrupted inner and outer segment junction around the fovea (between arrows).

Figure 10 High definition spectral domain ocular coherent tomography of the left eye shows hyper-reflectance throughout the outer nuclear layer and photoreceptor layer under the fovea.

a small number develop long-term severe dermatological or joint sequelae.^{14 18}

Learning points

- ▶ Reactive arthritis is associated with conjunctivitis or iritis. Rarely reactive arthritis is accompanied by permanent visual loss from macular infarction or foveal scarring.
- ▶ Reiter syndrome can be associated with permanent visual loss in the same way that it is associated with permanent dermatological or joint damage.
- ▶ Reiter syndrome needs to be added in the differential diagnosis of multifocal lesions in the posterior pole.
- ▶ Reiter syndrome is associated most often with HLA B27 and less so with HLA B51.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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