

Central retinal vein occlusion as an uncommon complication of sarcoidosis

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ABSTRACT

A 23-year-old black woman with acute blurred vision of the right eye was referred for ophthalmological examination. Fundus examination and fluorescence angiography showed a non-ischaemic central retinal vein occlusion (papillophlebitis). The diagnosis of sarcoidosis, suggested by the presence of bilateral hilar and mediastinal lymphadenopathy, was confirmed by transbronchial biopsy of the lymphadenopathy demonstrating noncaseating granulomas. The eye problems were successfully treated with systemic corticosteroids. Central retinal vein occlusion is a rare complication of sarcoidosis.

KEYWORDS

Sarcoidosis, central retinal vein occlusion, complication

INTRODUCTION

Sarcoidosis is a multisystem disorder of unknown aetiology that is characterised by tissue infiltration of noncaseating granulomas. Tissue biopsy is needed for a definitive diagnosis. Although the lungs are the most common site of inflammation, sarcoidosis can also involve other organs such as the skin, nervous system, heart, spleen and eyes.¹ Ocular involvement occurs in 25 to 50% of patients with sarcoidosis.²⁻⁵ The most common ocular manifestations are uveitis and conjunctival nodules.⁵ Central retinal vein occlusion, however, is an uncommon complication and only reported in a few case reports.⁶⁻¹¹ We describe a patient whose presenting clinical manifestation of sarcoidosis was a unilateral central retinal vein occlusion.

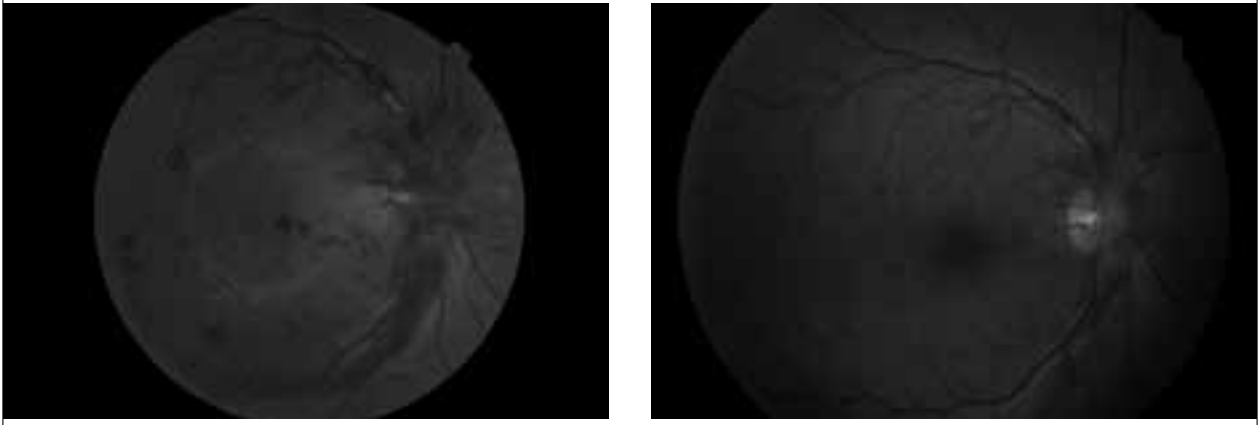
CASE REPORT

A 23-year-old black woman was referred to our hospital because of acute blurred vision in the right eye. Her medical history was unremarkable and she was not on any medication. There was no history of eye trauma. She had been consulting her general practitioner for four months because of dyspnoea and coughing; treatment with different antibiotics and inhalation therapy with corticosteroids and sympathicomimetics had no effect. Furthermore she complained of peaks of fever, fatigue, arthralgias of her hips, knees and ankles and she had lost 10 kg of weight.

Physical examination showed a moderately dyspnoeic young woman with a saturation of 97%, temperature of 36.0°C, blood pressure 110/65 mmHg, and pulse 90 beats/min. No pathological lymph nodes were found. Heart and breath sounds were normal. There was no hepatosplenomegaly, and no skin changes or signs of arthritis. Neurological examination was normal. Ocular examination showed a visual acuity of 20/30 in the right eye and 20/20 in the left eye and normal intraocular pressures. Slit lamp examination showed fine keratic precipitates and cells in the anterior chamber of both eyes. There were no clear signs of vitritis. Fundus examination of the right eye showed multiple flame-shaped haemorrhages surrounding the optic disc, tortuous retinal veins and dot-blot haemorrhages in all quadrants of the retina (*figure 1A*). The left eye showed some small retinal haemorrhages.

Laboratory tests showed an increased erythrocyte sedimentation rate, a mild anaemia and leucocytosis and an increased level of angiotensin-converting enzyme (ACE). Blood cultures remained negative. Chest X-ray demonstrated bilateral mediastinal and hilar lymphadenopathy. A high resolution computed tomography

Figure 1. A) Fundus examination of the right eye: multiple flame-shaped haemorrhages surrounding the optic disc, tortuous retinal veins and dot-blot haemorrhages in all quadrants of the retina; B) Fundus examination of the right eye ten days after starting systemic prednisolone: the fundus lesions have almost entirely disappeared



(HRCT) scan of the chest showed extensive bilateral symmetrical hilar and mediastinal lymphadenopathy and multiple nodular lesions in both lungs. Lung function testing was performed and showed static and dynamic values within the normal range and a normal diffusion. Transbronchial biopsy of the mediastinal lymph nodes was performed without complications and the specimens showed noncaseating granulomas confirming the diagnosis of systemic sarcoidosis. Because of the eye involvement high-dose systemic and local prednisolone was instituted and the eye lesions diminished within days (figure 1B).

DISCUSSION

Sarcoidosis is a multisystem, granulomatous disease of unknown aetiology which is characterised by noncaseating epithelioid granulomas in affected organs. The disease can affect every organ but the most common organs are the lungs.¹ Involvement of the eye is reported in up to 50% of the patients and is the presenting symptom in 5%.²⁻⁵ Ocular findings that support the diagnosis of sarcoidosis are the following: mutton-fat keratic precipitates, iris nodules, trabecular meshwork nodules, peripheral anterior synechia, vitreous opacities (snowball/string or pearls-like appearance), multiple chorioretinal peripheral lesions, retinal perivasculitis (periphlebitis), and/or candlewax retinochoroidal exudates and/or laser photocoagulation spot-like retinochoroidal atrophy, optic disc nodule(s)/granuloma(s) and/or solitary choroidal nodule and bilaterality.^{12,13} Ocular involvement in sarcoidosis encompasses a wide range of clinical manifestations. The most frequently seen are uveitis and conjunctival nodules. In addition, secondary glaucoma,

cataract formation and blindness are late complications in untreated patients.^{4,5,14,15}

A central retinal venous occlusion (CRVO) is an important cause of visual loss among older adults throughout the world and is mostly associated with arteriosclerotic ischaemic changes of the veins. However, the occurrence of a non-ischaemic CRVO in a young patient is rare. Moreover, when retinal venous occlusion occurs, it usually involves the smaller peripheral vessels, and has been attributed to retinal phlebitis, choroidal granuloma as well as impaired ocular circulation.^{7,8,11,16,17} Only a few case reports have described a central venous occlusion as a symptom of sarcoidosis.^{6-11,15} The exact pathogenesis of CRVO in sarcoidosis is unknown. Possibly, it is a clinical manifestation of microangiopathy that is also found in other organs affected with sarcoidosis.¹⁸ It can also be caused by infiltration of the retinal vessels by sarcoid granulomata manifesting in a perivascular mantling on the vessels with lymphocytes and epithelioid cells and perivascular proliferative changes compressing the vessels leading to luminal occlusion.¹⁹

Treatment of ocular sarcoidosis is indicated even when symptoms are slight, because of the risk of severe loss of vision. Corticosteroids are the mainstay of treatment. Mild cases of uveitis can usually be treated adequately with topical corticosteroids alone. Systemic corticosteroids are indicated in uveitis not responding to topical corticosteroids or in the presence of bilateral posterior involvement, especially with macular oedema and occlusive vasculitis. In 5 to 20% of the patients who are corticosteroid resistant or require an unacceptable dose to maintain remission, various cytotoxic steroid-sparing drugs are used. Methotrexate is the most commonly used steroid-sparing agent in the treatment of sarcoidosis, but

azathioprine, mycophenolate mofetil and leflunomide have also been shown to be useful.^{2,20-23} Anti-TNF biological therapies such as infliximab and adalimumab may be helpful for those patients experiencing persistent disease or an intolerance to cytotoxic immunosuppressive therapy. However, these agents themselves are associated with uveitis as well.²⁴

The main cause of visual loss is cystoid macular oedema.²⁵ Poor visual prognosis is associated with advanced age of the patient, black race, female sex, chronic systemic disease, and also with posterior segment involvement, peripheral punched out lesions, and the presence of cystoids macular oedema and glaucoma. However, the visual prognosis of sarcoidosis is usually good.²⁵

CONCLUSION

Our patient presented with a central retinal venous occlusion of the right eye as the presenting symptom of sarcoidosis. This is an uncommon complication of sarcoidosis.

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