Sudden loss of vision caused by a vasculiticophthalamic artery occlusion in a patient with ankylosing spondylitis and Crohn’s disease

We report for the first time a vasculiticophthalamic artery in an HLA-B27 positive patient with ankylosing spondylitis and a new presentation of Crohn’s disease.

Case report

A 37-year-old woman presented with sudden painless loss of vision in the right eye to perception of light, the left having poor vision from chronic uveitis and glaucoma. Examination showed iritis, an intra-ocular pressure of 2 mm Hg, a cherry red spot at the macula, and a normal fundus. The left eye showed a pale disc from long standing glaucoma. General examination showed that sinus tachycardia and blood pressure was normal.

The patient had a history of bilateral uveitis and ankylosing spondylitis for 14 years and was HLA-B27 positive. She had undergone cataract surgery bilaterally, followed by the development of glaucoma, requiring bilateral trabeculectomies and subsequently Molteno implants for its control.

Some weeks before admission, she had reported general constitutional symptoms, abdominal pain and loose stools which were controlled with sulphasalazine.

Acute investigations showed an erythrocyte sedimentation rate of 67 mm/h, C reactive protein 142 IU, neutrophilia and macrocytic anaemia (Hb 11.4 g/dl). Initial management comprised methylprednisolone (1 g/day) followed by 80 mg of prednisolone daily with a rapid taper and subcutaneous heparin. Magnetic resonance imaging and magnetic resonance angiography showed no evidence of cerebral vasculitis and normal flow in the ophthalmic arteries. Echocardiograms showed no source of embolus. Carotid ultrasound was normal. She was then referred for further management.

At this stage, she was on 40 mg of prednisolone and her vision had improved to count fingers in the right eye. Goldman visual fields showed marked constriction to all targets with a central scotoma to the V4e in the right eye. The intra-ocular pressure was 11 mm Hg bilaterally. Right fundal examination showed attenuated blood vessels and a pale disc. Inflammatory markers had risen again with a C reactive protein 108 IU. Antineutrophil cytoplasmic antibodies, lupus anticoagulant and thrombophilia screen were normal. Coloscopy showed moderate to severe colitis but no vasculitis including the terminal ileum. Histopathology of the large-bowel mucosa showed chronic active colitis, cryptitis, crypt absceses and a diagnosis of active Crohn’s disease was made. TNFα blockade treatment using infliximab was instituted with improvement of bowel and joint symptoms but her vision remained unchanged.

Comment

Iritis is the most common extra intestinal feature of inflammatory bowel disease (2% in women; 1.1% in men). Common ocular features in Crohn’s disease include anterior uveitis, episcleritis and more rarely scleritis, keratitis, orbital pseudotumour and retinal vasculitis which may cause retinal artery occlusion. Branch retinal artery occlusion as a complication of retinal vasculitis causing subsequent retinal neovascularisation has also been reported and fluorescein angiography has confirmed evidence of subclinical retinal vasculitis in patients with inflammatory bowel disease. A case of Crohn’s disease with joint involvement has been reported after presentation with ophthalmic artery occlusion but this is the first case report of ophthalmic artery occlusion with oculus ischaemia associated with ankylosing spondylitis (HLA-B27 positive) and later diagnosis of Crohn’s disease.

Summary

The likely aetiology is an obliterative vasculitis caused by granulomas in the blood vessel wall. Crohn’s disease has also been reported in association with large vessel arteritis and arterial occlusion. Anti-TNFα agents have shown effectiveness in the treatment of spondylarthropathies and Crohn’s disease; amelioration of the extra-intestinal manifestations of the disease is variable. This case highlights the need to consider vasculitic causes of oculus ischaemia in patients with seronegative arthropathies who are HLA-B27 positive and should alert ophthalmologists that further investigation is necessary.

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References


Orbital brachytherapy for extrascleral extension of choroidal melanoma

Extracocular extension of choroidal melanoma increases the risk of metastasis. Anterior extracocular extension can be treated by local resection with enucleation or scleral reinforcement after local radiation therapy. Advanced cases are dealt with by enucleation with resection of all visible orbital melanoma followed by radiation. External beam radiation therapy (EBRT) is used for presumed residual microscopic orbital melanoma. Massive extracocular extension may require orbital exenteration (also followed by irradiation).

Treated similarly, extracocular extension can also occur after plaque radiation, local resection and trans-scleral thermodony (TTT) or radiation therapy is used to reduce the rates of orbital and systemic recurrence. We report on the first use of orbital brachytherapy as an alternative to EBRT for extrascleral extension of choroidal melanoma.

A 63-year-old man presented to The New York Eye Cancer Center, with an American Joint Committee on Cancer (AJCC) T4N0M0 choroidal melanoma. Ultrasound disclosed a 6.5-mm-high tumour, 25×23 mm base, with extrascleral extension.

A metastatic survey was negative. The eye and all visible extrascleral tumour was removed and a 20-mm polyethylene-terephthalate implant was inserted. Histopathology showed an epithelioid malignant melanoma,
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