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FC Lam and CR Weir

Tennent Institute of Ophthalmology, Gartnavel General Hospital, 1053 Great Western Road, Glasgow G12 0YN, UK

Correspondence: FC Lam,
Tel: +44 141 211 3000;
Fax: +44 141 211 2054.
E-mail: fook_chang@hotmail.com

Eye (2007) **21,** 847–849; doi:10.1038/sj.eye.6702703; published online 30 March 2007

Sir,

Muckle–Wells syndrome: another cause of acute anterior uveitis

Muckle–Wells syndrome (MWS) is a rare genetic disorder characterised by recurrent urticaria, arthritis, sensorineural deafness, and general signs of inflammation and secondary amyloidosis. It affects the eyes in the form of conjunctivitis.[1] We present a case of a female patient with MWS who presented to us with recurrent attacks of severe acute anterior uveitis which has not been reported previously in association with this syndrome.

Case report

A 56-year-old female patient presented to our A + E department with 2 days history of photophobia and blurred vision of left eye. The patient was a known case of MWS and had always suffered from episodes of conjunctivitis in the past in both eyes. On examination, her acuities were 6/6 in the right eye and 6/18 in the left eye. Anterior segment examination of the left eye showed keratic precipitates with inflammatory cells and a hypopyon in the anterior chamber (Figure 1). Fundus examination was unremarkable. The anterior uveitis resolved with standard treatment and the acuity recovered to 6/6. She presented again 2 months later with severe anterior uveitis, again responding quickly to conventional treatment. The patient is currently symptom free and is on the tapering dose of steroid drops.

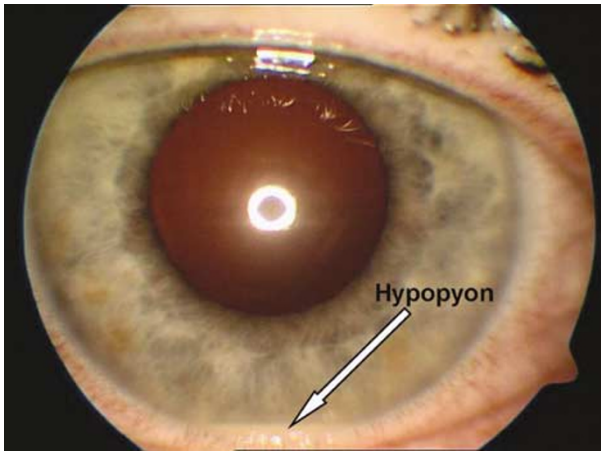

Figure 1 Left eye demonstrating hypopyon.

Comment

MWS (first described in 1962)[2] belongs to a group of hereditary periodic fever syndromes. These syndromes are characterised by intermittent attacks of fever. Four of these syndromes have been described. Familial Mediterranean fever and hyper IgD syndrome are transmitted as autosomal-recessive traits and MWS and tumour necrosis factor-receptor-associated periodic syndrome (TRAPS) are transmitted as autosomal-dominant trait.[3]

Patients with MWS suffer from acute febrile inflammatory episodes (denoted as 'aguey bouts' in Derbyshire, UK, where the first families with MWS were described). Each episode, which commonly manifests in childhood, include abdominal pain, arthritis, urticaria, and conjunctivitis. The disease may later be complicated by sensorineural deafness and secondary amyloidosis (type AA). The diagnosis is usually clinical and the CIAS1 gene (also called as NALP3 or PYPAF1) has been localised at chromosome 1q44 by linkage analysis.[2] Two other syndromes, familial cold autoinflammatory syndrome (FCAS) and chronic infantile neurological cutaneous and articular syndrome, are associated with mutations in the CIAS1 gene.[1]

The CIAS1 (cold-induced autoinflammatory syndrome 1) gene is expressed in peripheral blood leukocytes and encodes a protein 'cryopyrin' with the

same N-terminal domain as pyrin, a protein associated with familial Mediterranean fever. Cryopyrin appears to play a role in innate immune function by regulating the production of proinflammatory cytokines. Cryopyrin expression is also very similar in human and mouse. Significant expression of cryopryin occurs in mouse eye and skin tissue, which is consistent with symptoms observed in human cryopyrin-associated diseases.[4] Several different mutations of CIAS1 gene, all located in exon 3 have been described.[5] Our patient had an A439V mutation (genotyped in June 2004).

The clinical spectrum of diseases associated with CIAS1 mutations is very wide and includes other forms of familial urticaria which do not necessarily meet the clinical criteria of MWS and FCAS.[5] This case has highlighted a new association with MWS.

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A Shakeel and P Gouws

Department of Ophthalmology, Conquest Hospital, The Ridge, St Leonards on Sea, East Sussex, TN 37 7RD, UK

Correspondence: A Shakeel
Tel: +1424 755470 ext 8749;
Fax: +1424 758146.
E-mail: ayisha.shakeel@esht.nhs.uk

Eye (2007) **21,** 849–850; doi:10.1038/sj.eye.6702704; published online 2 February 2007

Sir,

Papillitis, Lyme disease, and cats

We present a case of papillitis caused by Lyme disease in the UK. We also highlight the increased risk of Lyme disease in cat owners.

Case report

A 50-year-old man presented with a 10-day history of central blurring of vision in his left eye. He had noted a rash on his right forearm 6 months previously. There was no definite history of a tick bite or of visiting high-risk areas, but his cat had ticks. Unaided vision was 6/6-3 right and 6/24 left improving to 6/12 with pinhole. There was a mild left relative afferent pupillary defect, a paracentral scotoma inferior to fixation, mild impairment of colour vision, and a pink swollen optic disc on this side.

Lyme disease antibody titres were positive by ELISA and Western blot methods, confirmed by a reference laboratory. The following investigations were normal or negative: full blood count, ESR, auto-antibody screen, coagulation screen, angiotensin converting enzyme, anti-neutrophil cytoplasmic antibody, syphilis serology, and lupus anticoagulant. The patient was treated with a 2-week course of intravenous ceftriaxone. The visual acuity however did not change with treatment.

Comment

Lyme disease can mimic many ophthalmic conditions. It has been reported to cause eyelid oedema, conjunctivitis, keratitis, episcleritis, anterior uveitis, vitritis, choroiditis, endophthalmitis, neuroretinitis, exudative retinal detachment, retinal vasculitis, optic neuritis, optic atrophy, pseudotumour cerebri, paresis of cranial nerves, and orbital myositis.[1,2] Optic neuritis in Lyme disease may or may not respond to treatment with antibiotics.[2]

In the mid-1970s, Dr Allen Steere and colleagues noted that a significant number of patients with Lyme disease had cats and had noted ticks on their pets, compared with their unaffected neighbours.[3] Lyme disease is now known to occur in cats, and *Borrelia burgdorferi* has been isolated from the tissue of adult and nymphal ticks removed from domestic cats.[4,5] The increased risk of Lyme disease in cat owners is not widely known. This case report may represent Lyme disease causing papillitis transmitted from the patient's cat.

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