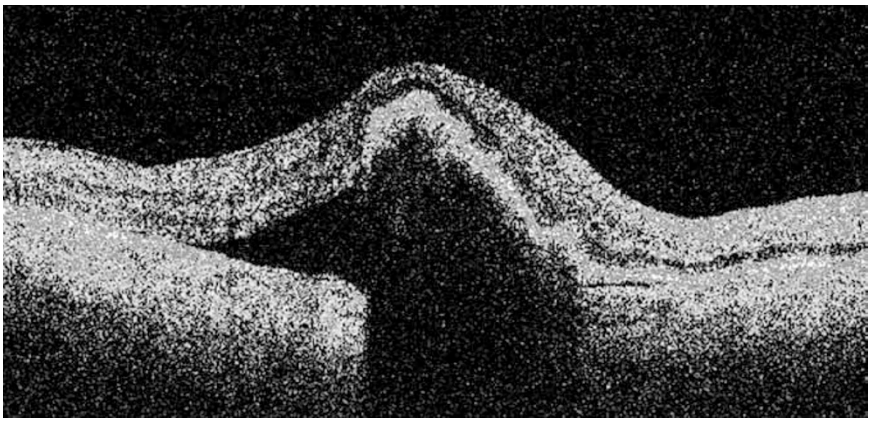

Figure 2 Optical coherence tomography after the intravitreal injection showing the classical retraction of the RPE.

Visual recovery after RPE tear is uncommon but possible in some instances, especially when the fovea is spared and conserves the RPE.

RPE tears can also follow intravitreal injections of ranibizumab presumably for similar reasons as with the other anti-VEGF agents.

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Sir,

Non-Hodgkin's lymphoma presenting with nystagmus: an unusual case

Malignant lymphomas are primary neoplasms of lymphoid tissue derived from lymphocytes and occur as solid tumours usually within lymph nodes and less often in extranodal lymphoid tissues such as the tonsil, gastrointestinal tract, and spleen. Most of the patients with non-Hodgkin's lymphomas present with superficial lymph node enlargement, with or without systemic symptoms. Presentation with purely central nervous system symptoms in the absence of other systemic signs is extremely rare and to our knowledge there are only three previous reports of patients with lymphoma presenting with nystagmus.[1–3] We therefore report a further case in a patient who initially presented with nystagmus and ataxia and was subsequently diagnosed with non-Hodgkin's lymphoma.

Case report

A 60-year-old male was referred to the ophthalmology department with a 6-month history of an intermittent vertical diplopia and oscillopsia. The diplopia usually occurred during close work and usually lasted several seconds. He had no history of vertigo, nausea, or vomiting, although he was noted to have an unsteady

gait. He was otherwise fit and well with no past medical or ophthalmic history of note. He was not taking any drug medication. His visual acuities were 6/6 in each eye and orthoptic examination revealed full extraocular movements and convergence to 10 cm. There was a small amplitude gaze evoked nystagmus of moderate frequency. The nystagmus was conjugated with a mixed horizontal and rotatory movement. The fast component of the horizontal nystagmus was in the direction of gaze. The rotatory component was clockwise on left gaze and counter clockwise on right gaze. The vertical and horizontal saccades were felt to be hypometric but smooth pursuits and convergence were thought to be normal by two independent and experienced observers. The prism cover test revealed an exophoria of 4 Δ (near) and an esophoria of 1–2 Δ (distance). Prism fusion range was 2 Δ base in to 2 Δ base out (near), and 2 Δ base in to 6 Δ base out (distance).

Other than a possible left-sided sensory-neural hearing loss, lower cranial nerve function was normal. Tone, power, and reflexes were normal in all four limbs. Coordination of the upper limbs was normal but ataxia was present on tandem walking and he had a positive Romberg's test. The remainder of his ophthalmic examination was normal. In view of his neurological signs, he was referred to the neurology department for further investigations.

MRI scan revealed thickening of the fifth, seventh, and eighth cranial nerves with enhancement after contrast, the appearance of which was thought to be consistent with an inflammatory disorder. The brain parenchyma, ventricular system, and cerebellopontine angles were clear. CSF examination showed a raised white count of 18 mm^3 with an associated raised protein of 1.62 g/l and a normal glucose of 3.4 mmol/l. In addition, a polyclonal increase in CSF gamma globulin was noted. Mycobacterial cultures were negative. Routine blood investigations were normal although the adjusted serum calcium was slightly high at 2.61 mmol/l. B_{12}, folate, and thyroid function tests were normal. Rheumatoid factor, antinuclear antibody, anti-neutrophil cytoplasmic antibody, VDRL, and treponemal enzyme immunoassay were all negative and his chest X-ray was normal. The diagnosis at this stage was unclear but neurosarcoidosis was thought to be the most likely possibility. He was therefore commenced on oral prednisolone, and although his symptoms improved gradually over a period of several months, he was still bothered by intermittent diplopia and oscillopsia.

Eleven months after his initial presentation, he developed myocardial type chest pain. He was found to be anaemic, thrombocytopaenic, neutropaenic, and the LDH was significantly elevated. A bone marrow biopsy was undertaken and this revealed a malignant infiltrate of lymphoid origin giving a diagnosis of a diffuse large B-cell lymphoma (stage IVB). A CT scan of the chest, abdomen, and pelvis revealed no evidence of bulk disease. A repeat MRI scan at this stage with gadolinum revealed no focal enhancement and no evidence of significant enhancement in relation to the seventh and eighth cranial nerves.

The patient was given a 3 weekly course of chemotherapy with R-CHOP (cyclophosphamide, hydroxodaunorubicin, vincristine(oncovin) and prednisolone) with CNS prophylaxis over a 6-month period. His oscillopsia, diplopia, gaze evoked nystagmus, and ataxia improved dramatically within 3 months. His prism fusion range improved to 8 Δ base in to 14 Δ base out (near) and 4 Δ base in to 10 Δ base out (distance). A repeat bone marrow trephine biopsy undertaken 6 months later revealed no evidence of residual lymphoma and he has remained asymptomatic.

Comment

Torsional nystagmus can result from peripheral vestibular imbalance or direct labyrinthine destruction; however, a pure torsional nystagmus almost never occurs with peripheral vestibular disease, as this would require the selective involvement of the vertical semicircular canals in both ears.[4,5] This imbalance in the vestibular tone can also cause a gait ataxia. It is extremely rare for lymphoma to present in this manner, and although this patient did have a prolonged period of investigation before a final diagnosis was reached, we believe that his initial presentation with nystagmus was due to lymphoma. We postulate that the infiltration of the eighth cranial nerves with lymphoma resulted in a disturbed vestibular tone from the semicircular canals, and consequently, the mixed horizontal and rotatory nystagmus, and the gait ataxia. The initial partial remission of his neuroophthalmological signs and the resolution of cranial nerve infiltration on the repeat MRI scan after steroid therapy are consistent with the diagnosis of lymphoma. This is further supported by the significant improvement following his chemotherapy. The intermittent diplopia was felt to be due to the patient's poor fusional range. The patient's fusional range and diplopia improved with the treatment of the lymphoma and orthoptic exercises. This case highlights the importance of appropriate referral and thorough neurological investigation in such patients particularly as appropriate management can lead to resolution of disabling visual symptoms.

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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.

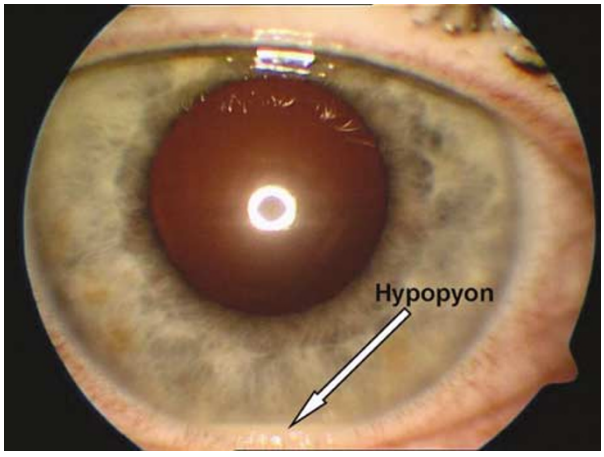

Figure 1 Left eye demonstrating hypopyon.

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