Acute Case Study ERLogbook

OPHTHALMOLOGY
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Recurrent optic neuritis

25 year old female of Irish background presents with a 3rd episode within 10months of blurring of vision, pain on eye movement and heachache.

HxPC

July 2012 The initial episode of blurring of vision, headache and eye pain. The attack began with a headache and progressed over 4 weeks with worsening vision and pain. After 2 consultations she attended A&E, optic neuritis was diagnosed, IV hydrocortisone was administered followed by oral steroids tapering. The symptoms responded rapidly and premorbid visual function was regained within 4 weeks. Other signs elicited on examination at the time of diagnosis were an RAPD and colour desaturation. OCT studies were performed which showed a normal retinal thickness.

Jan 2013 Second attack occurred with the same symptoms of headache, blurring of vision and eye movement pain. The patient was unable to obtain treatment at the local hospital as she was not in Sydney at the time. Fortunately the attack was less severe and with no treatment recovered over 4 weeks.

May 2013 Current attack presented with the same symptoms as previous. Re-attended Sydney A&E after the symptoms progressed over 3 days. Treatment with IV hydrocortisone was initiated and improvement in vision overnight after the 1st dose occurred. The signs of RAPD, reduced colour saturation and reduced visual fields were present on presentation.

POHx-Nil

PMHx-Nil

FHx-Nil

Optic neuritis is an inflammation of the optic nerve most commonly due to an inflammatory attack and demyelination of the optic nerve. This can be T cell ((MS), B cell or complement mediated (NMO) and it has been found to be associated with HLA-DR15. It is more common in women (3:1 F:M) aged between 20-40yrs. There is a higher incidence in populations at higher latitudes and within the Caucasian population compared to afro-Caribbean.

Diagnosis of optic neuritis should consider and exclude other possible causes of optic neuritis. These should include exclusion of hereditary optic neuropathy, thyroid function tests, ESR, ANAC and anti-nuclear antibodies, exclusion of infection, pressure from space occupying lesions, tumours of the nerve sheath, gliomas of the optic nerve and infiltration from sarcoidosis.

Optic neuritis is most commonly monocular (bilateral optic neuritis more commonly occurs in children), with the most common clinical features eye pain and loss of vision (92.2% in the optic neuritis treatment trial (1991)) with the pain concurrent with the visual loss and worse on eye movement. In the Optic neuritis treatment trial which studied 457 patients, a variety of visual field defects where demonstrated however the most typical is a central scotoma. In this case the patient did not demonstrate a central scotoma (test was difficult due to the reduced visual acuity). The field tests in the optic neuritis trial did however in some cases demonstrate field defects in the other eye which improved suggesting a subclinical degree of involvement of the other eye in some cases.

Optic disc swelling from papillitis is only seen in approximately 30% of cases, and though a pale disc was seen no swelling was evident. However the optical coherence tomography(OCT) imaging demonstrated some swelling at the time of the first occurrence. The pale disc which was described during this attack is most likely demonstration of the atrophy from the previous attacks. The serial OCT

Optic neuritis is the presenting symptom of Multiple sclerosis in 15-20% of cases and occurs in 50% of cases at some stage in the course of the disease. The risk of developing MS is increased in suffers of optic neuritis and is higher (72% v 25%) in those who have white matter lesions demonstrated on MRI. Oligoclonal bands in the CSF is also an indicator of higher risk of developing MS.

Neuromyelitis optica (Devic's Disease) is a condition which is sometimes described as within the MS spectrum. It is a demyelinating disorder with a severe course in most cases. Diagnosis is based on the presence of optic neuritis, acute myelitis with a negative MRI brain and evidence of spinal cord lesions spanning 3 or more vertebral segments. Anti-aquaporin 4 antibodies are found to have a 70% sensitivity for neuromyelitis optica, and is a minor indicator in the Wingerchuck criteria. Optic neuritis can occur in isolation therefore this should also be considered.

In Older patients (>50yrs) ischaemic optic neuropathy should be considered as a likely diagnosis particularly in diabetics. Giant cell arteritis should also be ruled out, as early treatment can prevent progression and blindness from controlling contralateral eye involvement.

Treatment for typical optic neuritis is IV methylprednisolone, though this is found to hasten recovery but not to affect the end visual acuity at recovery. However recover occurs without treatment and therefore treatment is controversial. It may only be offered to patients with poor vision in the contralateral eye or severe pain. In this case the patient during her first episode, which she reported as more severe, received IV methylprednisolone however during her 2nd episode she did not receive any treatment and recovered spontaneously over 2 weeks.

There is some evidence to support early treatment with disease modifying agents such as β interferon, in cases at high risk of progressing to MS (MRI findings) with the outcome of lower risk of conversion to full MS. On this basis in the UK β interferon treatment is offered to those who have suffered 2 significant episodes within 2 yrs (CHAMPS trial and ETOMS study). In this case there are no findings to indicate a high risk of MS conversion; none of the MRI imaging studies has shown any CNS lesions.

As this case has no indications of the development of MS at this point the treatment with B interferon is contraindicated, as in cases of neuromyelitis optica the treatment has no effect and can in fact be detrimental. If it is neuromyelitis optica studies have shown in steroid refractive cases in the acute phase plasma exchange can be effective and in the longer term treatment and relapse prevention immunosuppression e.g. azathioprine, is a more effective method, during which a reduction in the number of anti-aquaporin 4antibodies can be demonstrated [Palace, *et al.*, 2010]. Future treatments using monoclonal antibodies directed against B cells (anti-CD20) and anti-complement are the subject of trials in treating neuromyelits optica, however due to the rarity of this disorder these trials are difficult to conduct but may lead to improved treatment of this aggressive disorder.