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A case of childhood multiple sclerosis

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Abstract

Aim: **To describe a case of multiple sclerosis which presented with sudden-onset esotropia.**
***Method*: A case is documented with history, ophthalmic and neurological findings and management. Multiple sclerosis is discussed in relation to paediatric onset.**
Results: **A previously healthy boy aged 2 years 10 months presented with malaise, fever and sudden-onset right esotropia, which followed a urinary tract infection 4 weeks earlier. Ophthalmic examination revealed bilateral papilloedema. Magnetic resonance imaging (MRI) showed multiple areas of high signal in the basal ganglia and subcortical white matter in the occipital and right frontal lobes. Steroid medication was commenced and resulted in a reduction in signs and symptoms. The provisional diagnosis was acute demyelinating encephalomyelitis (ADEM). Following two further similar episodes a diagnosis of multiple sclerosis was made based on the variation of neurological signs and resolving demyelination on MRI brain scans.**
***Conclusion*: Multiple sclerosis is very rare in childhood. This was not a typical case of esotropia in a 2-year-old child, as the presence of bilateral papilloedema was indicative of urgent investigation.**

Key words: Acute demyelinating encephalomyelitis, Esotropia, Multiple sclerosis, Optic atrophy, Papilloedema

Introduction

Multiple sclerosis (MS) is not easy to diagnose. There is no absolute specific diagnostic test and in making the diagnosis the clinical pattern and presentation are of prime importance. The condition is rare in the paediatric population and onset prior to age 10 years occurs in 0.2–2% of all cases.[1]

Since the introduction of magnetic resonance imaging (MRI), figures for paediatric MS have been increasing.[2] A clinical pattern for MS in children under 5 years old is evolving from reports in the literature.[3,4] Ruggieri *et al.*[3] and Cole and Stuart[4] have reported a greater incidence of MS in females in this age group compared with the juvenile and adult age groups. The first occurrence of MS appears to affect the child's general health more than in other age groups. Other symptoms of the disease appear to be similar to those reported in adults. Visual problems include diplopia, blurred vision, optic neuritis and papilloedema.[5] Sensory signs such as paraesthesia in one or more limbs and the face are common features. Dysarthria, vertigo and sphincter weakness are relatively uncommon in children.[1]

This report details a case of childhood MS. The features of the case are discussed in relation to the literature.

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Case report

At the age of 2 years 9 months the boy became unwell with a urinary tract infection. Four weeks later he developed general malaise and vomiting, followed 2 days later by sudden onset of a right esotropia. An urgent referral was made by the general practitioner to the local paediatric team with subsequent referral for ophthalmic examination.

Initial presentation

Orthoptic and ophthalmic examination

Orthoptic assessment confirmed a right esotropia of 30$^{\Delta}$. Ocular movements were full, with no evidence of lateral rectus weakness. There was a positive family history of squint on the paternal side. The child was uncooperative with vision assessment.

Cycloplegic refraction revealed a very small hypermetropic error. Examinations of the fundi and media revealed swollen and blurred disc margins R > L, and a diagnosis of bilateral papilloedema was made. The ophthalmic team requested an urgent MRI examination of the brain and brain stem.

Neurological assessment

The MRI examination was abnormal with multiple areas of high signal in the basal ganglia and in subcortical white matter in both occipital and right frontal lobes. The child was transferred to the regional children's hospital. A lumbar puncture was performed and the cerebrospinal fluid (CSF) showed a slightly raised white cell count. A diagnosis of acute demyelinating encephalomyelitis (ADEM) was made.

Under the supervision of the neurologist at the

regional children's hospital the child commenced steroid treatment and subsequently recovered from all symptoms including the right esotropia. However, optic discs remained pale and visual acuity was subnormal at 6/9 right and left.

Second presentation

Eight months later the child suddenly developed general malaise, unsteady gait and a recurrence of the right esotropia. He collapsed into unconsciousness and was immediately transferred to the regional children's hospital.

MRI revealed areas of inflammation that differed in location to the previous episode. Systemic steroids were administered with a resultant resolution of the inflammation and complete recovery of the right esotropia. The steroids were slowly reduced and eventually stopped.

Further presentations

Over the next 8 years the child suffered another three incidents of inflammation, during which he was unwell and required admission to the regional children's hospital and steroid treatment. Following the third episode, when the child was 5½ years old, the paediatric neurologist made an interim diagnosis of *probable* MS, which was responsive to steroids.

The fifth episode primarily affected the child's visual acuity. At this time the child was 10 years old. He presented with headaches and his parents reported visual difficulties at school. On assessment visual acuity was 6/36 right and left. Examination of the fundi and media revealed bilateral optic atrophy.

On admission to the regional children's hospital steroid medication was commenced. Over the next 3 months the visual acuity fluctuated greatly but deteriorated to R 4/60 L 4/60. The medication was changed to glatiramer acetate (Copaxone). Ten months later the vision had improved to R 6/18 L6/60.

Residual problems

Following the attacks of inflammation the child has residual problems, which include mild imbalance, right lower limb spasticity with abnormal posturing of the right ankle and an unstable bladder with occasional incontinence. He has one-to-one support in school and uses a CCTV for close work. The paediatrician and neurologist at the regional children's hospital continue to follow his care regularly.

Discussion

Childhood-onset MS is usually associated with systemic illness, malaise, fever, vomiting and sickness and follows a viral illness.[6,7] In this case, the presenting signs were right esotropia and bilateral papilloedema associated with general malaise and vomiting, which followed a urinary tract infection 4 weeks earlier. The child's condition responded to steroid treatment with a resolution of symptoms and the right esotropia, but optic discs were pale and visual acuity was subnormal.

MS may be difficult to diagnose because of variations in the pattern of onset, the course of the disease, and also its similarity to a number of other neurological conditions.[7] The assessment of CSF is not a specific test for MS. Viral and immune diseases show similar abnormalities and the presence of oligoclonal bands may be suggestive of MS. In this case there was a raised white cell count indicating the presence of inflammation.

A single episode of demyelination is not sufficient for a diagnosis.[5] Criteria are now being developed that relate to findings on MRI scans, although at present these have only been successfully applied to adults and not children. Using the diagnostic criteria of Poser,[8] which have been the gold standard in the past, diagnosis relies upon two separate episodes of white matter lesions in the central nervous system separated by place and time. In this case the diagnosis of MS was only made after a number of presentations with altering neurological signs and resolving demyelination on MRI brain scans.

In adults MS is described as being primary progressive, benign, relapsing remitting or secondary progressive. The child reported has a form of relapsing remitting MS, which responds to steroids. Relapsing remitting MS is reported to be common in childhood and the course of the disease is more favourable and less aggressive in young children.[4,13] In children less than 5 years of age there appears to be a shorter time interval between the first and second episodes of MS. Ruggieri *et al.*[3] report a 63% recurrence of inflammation within 1 year. In this child the first and second episodes of disease were 8 months apart.

There are no reports of clinical trials on the use of interferon beta1b or glatiramer acetate (Copaxone) in childhood. Glatiramer acetate has similar properties to interferon and it is expected to reduce the relapse rate and progression of the disease in relapsing remitting MS.[9,10] Details of the steroid treatment the child received are limited as the medication was under the supervision of paediatric neurologists at the regional children's hospital. Due to deterioration in the child's visual acuity the medication was changed from steroids to glatiramer acetate. Ten months later the child's vision had improved and stabilised. Adams *et al.*[2] reported on a child with relapsing remitting MS who, after using interferon beta-1b, demonstrated a dramatic clinical improvement and remission of some lesions. Ghezzi *et al.*[9] reported on a group of 65 children who were treated with glatiramer acetate or interferon drugs. All demonstrated a reduction in the relapse rate and progression of the disease.

The long-term prognosis of childhood MS is not well reported in the literature. The youngest documented case of MS was reported in 1911 and the child died at the age of 2 years 6 months.[12] The child reported here has had five episodes of MS since the age of 2 years 10 months, which have left him with residual physical and visual disabilities. Since commencing glatiramer acetate he has not experienced any further acute attacks.

Comment

Multiple sclerosis is very rare in childhood. This was not a typical case of esotropia in a 2-year-old child, as the presence of bilateral papilloedema was indicative of urgent investigation.

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There are no competing interests.

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