·Case Report·

Optic neuritis with multiple sclerosis in a 10-year-old Asian girl

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Abstract

• AIM: To report a case of unilateral optic neuritis with multiple sclerosis in an Asian child.

• METHODS: A case report.

• RESULTS: A 10-year-old Chinese girl presented with history of sudden loss of vision of the right eye for 3 days' duration. It was associated with pain in eye movement. She gave history of fever associated with ataxic gait a year ago. She had been diagnosed to have acute disseminated encephalomyelitis (ADEM). Visual acuity in the right eye was 'counting finger' with positive afferent pupillary defect. The optic disc was swollen and hyperemic. The colour vision was severely impaired. Visual field showed central scotoma and enlarged blind spot. Magnetic resonance imaging (MRI) of the brain revealed multiple intense lesions in the left occipital lobe, basal ganglia and periventricular regions suggesting multiple sclerosis. She was treated with intravenous methylprednisolone for 3 days, followed by oral prednisolone for 11 days. She had excellent recovery and her visual acuity improved to 6/9. She remained asymptomatic for 3 years.

• CONCLUSION: The simultaneous occurrence of optic neuritis and multiple sclerosis is less common in children and seldom reported. We presented this case to highlight the possibility of this disease occurring in Asian population in a younger age group.

• KEYWORDS: optic neuritis; multiple sclerosis; acute disseminated encephalomyelitis; childhood

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INTRODUCTION

Childhood multiple sclerosis (MS) is thought to be a rare phenomenon. We presented this case to highlight the possibility of this disease in a younger age group occurring in this part of the world. Childhood MS is difficult to diagnose since it has no specific criteria as compared to adult MS. Younger patient with MS usually presented with fits, brain stem and cerebella symptoms compared to adults[1]. Magnetic resonance Imaging (MRI) is one of the tools used to evaluate the evidence of MS in childhood. The features of MRI in patient with acute disseminated encephalomyelitis may be similar to those of MS patients.

CASE REPORT

A 10-year-old Chinese girl presented with history of sudden loss of vision of the right eye for 3 days' duration. It was associated with pain in eye movement. She had a history of fever associated with ataxic gait a year ago. She had been diagnosed to have acute disseminated encephalomyelitis (ADEM) at that time and had been treated with oral prednisolone. Her condition improved rapidly and had been well since then.

During the current admission, ocular examination revealed visual acuity of the right eye was counting finger with a positive afferent pupillary defect. Visual acuity of the left eye was 6/6. Anterior segments of both eyes showed normal ocular findings. Funduscopy showed swollen and hyperemic disc(Figure 1). The contrast sensitivity and colour vision test were severely impaired. The visual field showed central scotoma. The central nervous system examination was unremarkable. A diagnosis of unilateral optic neuritis was made. Blood investigations which included full blood picture, erythrocyte sedimentation rate and collagen screening were negative. Chest x-ray was normal. Magnetic resonance imaging (MRI) showed thickened right optic nerve and multiple hyperintense regions in cerebellar peduncles and cerebellar hemisphere (Figure 2,3). The visual evoke potential (VEP) showed a delayed latency at P100 with reduced amplitude in the right eye. Contrast sensitivity function (CSF) examination was deferred by the patient's parents.

She was treated with intravenous methylprednisolone 1mg/kg for 3 days, followed by oral prednisolone for 11 days. Her vision improved to 6/60 after 3 days. Upon discharge, her visual acuity was 6/9. She was followed up regularly for one

year. Her visual acuity was back to 6/6. Funduscopy showed normal appearance of optic disc. However, the central scotoma persisted but the size was smaller. The relative afferent pupillary defect was still present (grade 1). Colour vision test using Ishihara chart was normal. Subjectively, the red desaturation was reduced as compared to the other eye. There were no abnormal neurological signs elicited. She was completely recovered up by the end of 3 years' follow-up.

DISCUSSION

Multiple sclerosis is a disease of adolescent and middle-aged people. The incidence of MS in Asian population is about 5/100 000 and is the lowest prevalence rate in the world [2]. Malaysia has a low prevalence rate of MS that is about 2/100 000 and is more common in Chinese population[3]. The MS in young age group is rarely considered [4]. The percentage of multiple sclerosis in patients with childhood onset is between 2.7% and 4.4% among all MS patients, and the percentage of MS patients who had onset before the age of ten is between 0.2% and 1.6%. The diagnosis of MS is rare in children and it's more difficult because there are no specific criteria for diagnosis. Pediatric patients usually presented as sensory system disturbances, cerebellar and brain stem abnormalities, long tract dysfunction and optic neuritis[5]. Optic neuritis is one of the clinical manifestations that occur preceding MS. According to Kira[3], MS in Asian population is characterized by the selective and severe involvement of optic nerve. The cause of optic neuritis is not well established. Approximately 15% to 20% of MS patients present as optic neuritis [6]. Inflammation occurring at an isolated central nervous system site, for example, transverse myelitis, optic neuritis, brainstem dysfunction, can be monophasic or followed by a further neurological episode at another central nervous system site, qualified for conversion to MS [7]. Based on the study of Lucchinetti *et al* [7], about 13% of idiopathic isolated optic neuritis progresses to definite MS by 10 years, follow-up and the incidence increases with age.

However, in this Chinese girl, the development of MS was after one year of episode of acute disseminated encephalo-myelitis (ADEM) and simultaneously with optic neuritis, which is a rare presentation. Most childhood MS patients have a relapsing-remitting form of the disease, and less than 3% have a primary progressive form[5].

The possibility of multiphasic demyelinated encephalomyelitis (MDEM) also needs to be considered in children with clinical features suggesting MS. ADEM is a monophasic illness characterized by depressed sensorium, seizures and focal deficits. According to relevant studies, polysymptomatic presentation, encephalomyelitis, pyramidal signs, preceding illness such as chickenpox, measles, mumps, infectious mononucleosis and vaccination are more inclined to ADEM or MDEM. Recurrence of ADEM or MDEM is generally within 6 months after the initial presentation and tends to involve the same site [8-10].

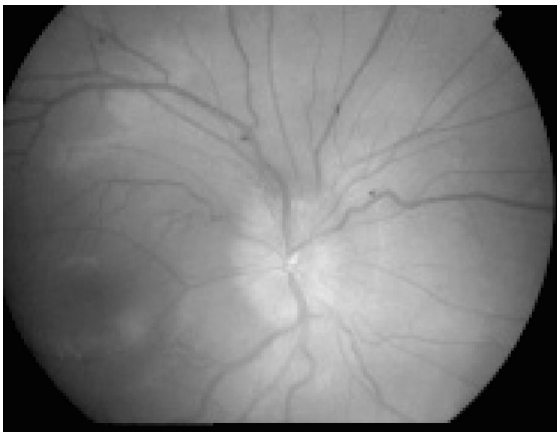

Figure 1 Right optic disc swelling and hyperemia at the onset of optic neuritis

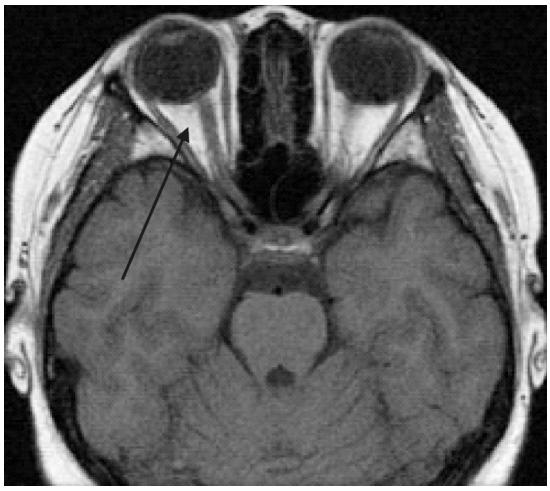

Figure 2 MRI of the brain and orbit showing a thickened right optic nerve

There are a few clinical characteristics of MS in children. They include acute onset of headache, nausea, vomiting, fever, seizures, altered state of consciousness, motor sensory hemi-syndromes, cerebellar and brain stem dysfunction [4]. However, bilateral involvement of optic neuritis is more common in ADEM or MDEM whereas the children with MS will usually present with unilateral optic neuritis [11].The ability to differentiate the diagnosis of ADEM form MS is important because it has a prognostic value. Children with ADEM usually will have a generally good outcome but those children with MS are thought to have a significant disability [10]. Therefore the diagnostic criteria in clinic and investigations including laboratory and imaging tools must be

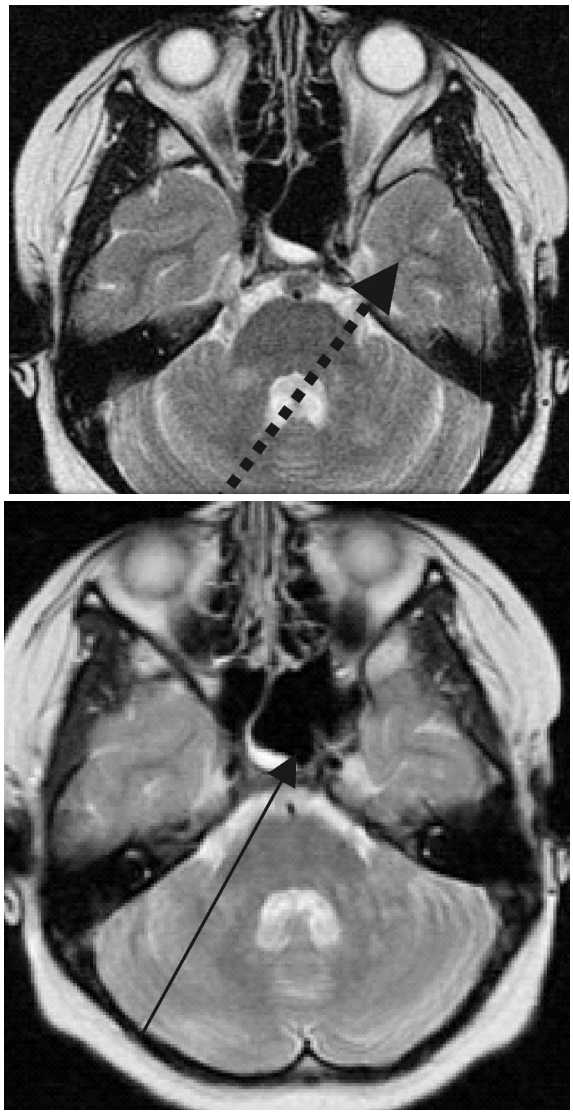

Figure 3 MRI of the brain and orbit demonstrates multiple well defined plaques and hyperintense areas in cerebral peduncles (thin arrows) and cerebellar hemisphere (dotted arrow)

further evaluated in order to come with accurate diagnosis.

Magnetic resonance imaging (MRI) is one of the recent modalities with features that may differentiate between ADEM and MS. Singh *et al* [11] and Dale *et al* [12] distinguish ADEM and MS based on MRI. They concluded that in ADEM, the lesion has a poorly defined margin, and tends to be deeper in white matter with periventricular sparing and grey matter of cortical and basal ganglia is frequently involved [11,12]. MS will give well defined "plaque" like margins, confined more at periaqueduct, corpus callosum and periventricular white matter [11,12]. However, the MRI findings in patients who presented clinically with ADEM may be difficult to be differentiated from those of the first attack of MS [13]. The changes are commonly resolved with clinical improvement of the patients.

In this patient, the diagnosis of MS is supported by history of cerebellar dysfunction which manifests as instability of gait, unilateral involvement of optic neuritis and multiple well defined plaques at cerebral peduncles and cerebellar hemisphere on MRI (Figure 3).

Corticosteroid is still the main treatment for optic neuritis. The treatment of optic neuritis has been investigated in several trials. The Optic Neuritis Treatment Trial (ONTT) was a landmark study which concluded that steroid hastens the visual recovery but not affects the final visual outcome [14,15]. However, it reduced the rate of clinical definite MS for 2 years [15]. Immunoglobulin infusion had been used as a treatment in MS. Spalice *et al* [15] concluded that the treatment with immunoglobulin alone at first, and then followed by interferon may improve visual acuity.

Interferon β-1α is one of the treatments available for optic neuritis. Study done by CHAMPS group concluded that combination of intravenous steroid and weekly intramuscular interferon β-1α reduced the rate of developing neurological event[16]. However, intravenous corticosteroid is still the mainstay for the treatment of optic neuritis. The combination of other treatment modalities should weigh the risks and benefits and not forget the cost factor. It is hopeful that the recent modalities of treatment will reduce the risks of developing MS and significant disability.

The prognosis of optic neuritis in children is relatively good. Brady *et al* [17] concluded that the normal MRI of the brain might be associated with a better outcome. Bilateral optic neuritis is more common in younger age group and about 22% end up with visual disability [17]. Our patient, presented with unilateral optic neuritis in the presence of brain lesion. The prognosis is good in this child but there is a reduction in the optic nerve function in the affected eye.

In conclusion, development of MS is rare in Asian population, particularly involving the younger population. Therefore, it should alert the pediatrician about this disease. The management of the patient with MS will involve multiple disciplines including ophthalmology, pediatrics and neurology. The ability to differentiate MS form other neurological events is important because this will give a different prognostic value. Explanation should be given to parents regarding the risks of developing MS in children who presented with optic neuritis.

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