SUCCESSFUL MANAGEMENT OF ISOLATED OPTIC NEURITIS IN ACUTE DISEMINATED ENCEPHALOMYELITIS: A CASE REPORT

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BACKGROUND

Acute Disseminated Encephalomyelitis (ADEM) is an autoimmune disease that causes multifocal inflammatory demyelination. The prevalence, clinical manifestations, and morbidity of ADEM vary. Although ADEM is a rare disease, the incidence rate is estimated at 1 in 125,000 - 250,000 population per year. The dominant population is children under 10 years of age and adolescents aged less than 20 years. The prevalence of women is more or less the same as that of men and is more common during cold weather. Risk factors include genetics, exposure to infectious organisms, and immunization [1].

ADEM is an autoimmune reaction to myelin proteins and neuronal cells. It can cross the blood brain barrier causing neurological signs and symptoms. The course of ADEM is preceded by fever due to viral infection or immunization. Infectious diseases in children that most often cause ADEM are viral infections, such as measles, mumps and chickenpox. The causes of post immunization are mainly inactive vaccines or live attenuated viruses such as influenza vaccines and measles rubella (MR) [2]

ADEM clinical manifestations include headache, weakness, nausea, and vomiting. Signs of encephalopathy such as irritability, psychosis, somnolence, and coma. Other signs are sensory and motor deficits, paraparesis and tetraparesis, brainstem deficits, dysarthria and oculomotor dysfunction, and signs of neurological abnormalities, such as seizures, meningismus, ataxia, aphasia, nystagmus, and optic neuritis [2].

Optic neuritis due to ADEM causes visual morbidity. Clinically, it can appear as an isolated condition or associated with various systemic autoimmune disorders. The incidence is 1 - 5 per 100,000 per year, especially in the Caucasian population. The pathophysiology of optic neuritis is inflammation, causing peripheral activation of T cells that can cross the blood brain barrier causing a type IV hypersensitivity reaction. This process results in the destruction of myelin with involvement of the nerve axons. Diagnosis and early management of optic neuritis due to ADEM has resulted in a favorable response.

Case presentation

A girl, age seven, presented to the emergency ward with the chief complaint of sudden blurred vision in both eyes. Complaints accompanied by pain in eye movements since 1 week ago, gradually getting worse. One week ago she got MR vaccination then had a fever. The patient did not have a history of wearing glasses, a history of trauma, and a history of taking medication.

Physical examination, general status within normal limits, good consciousness. Best visual acuity right and left eye hand movement, there was no relative afferent pupillary defect in both eyes, the other anterior segments were within normal limits. Examination of the posterior segment revealed hyperemia of both optic nerve papules. Color sensitivity examination using Ishihara color plate showed deficiencies in both eyes. Movement of the eyeballs is good in all directions, but pain is present. Optical coherence tomography (OCT) examination of both eyes revealed edema of the optic nerve head. MRI showed multiple hyperintense lesions in the right and left subcortical temporo-parietal suspicion of ADEM. Laboratory tests showed an increase in white blood cells (WBC), namely 19,430 / µl and C-Reactive Protein (CRP) 23,81mg / L.

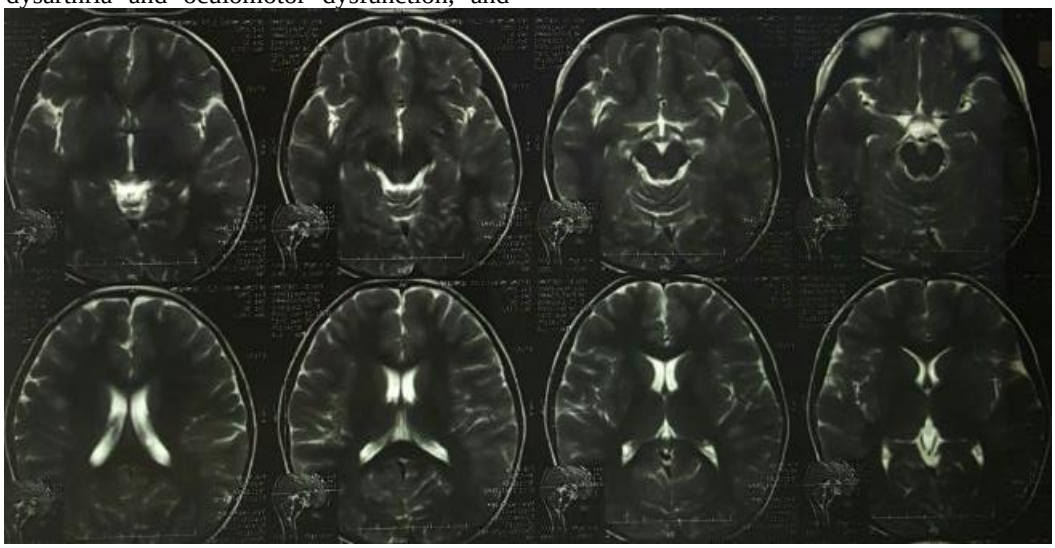

Figure 1. Multiple hyperintense lesions in the right and left subcortical temporo-parietal MRI examination.

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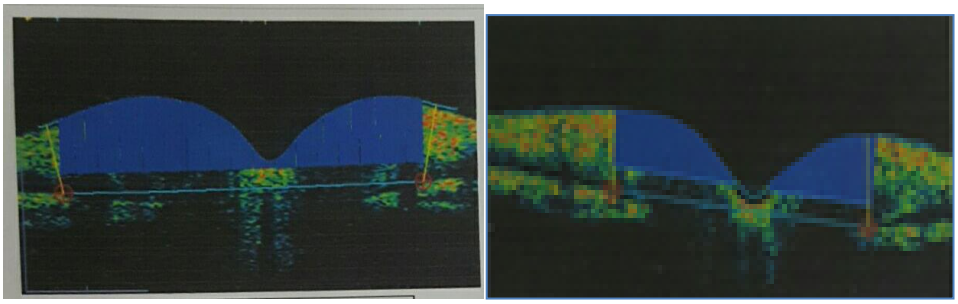

Figure 2. Optic nerve head edema on OCT examination.

Patients were given management according to the Optic Neuritis Treatment Trial (ONTT) guidelines, 125 mg of methylprednisolone intravenous injection every 6 hours for 3 days, followed by oral prednisone 1 mg / kgBW / day for 11 days, and added citicoline syrup 500 mg orally every 24 hours. On the 3rd day, visual acuity showed improvement to 2 meters in the right eye and 5/30 in the left eye, the pain in eye movement began to decrease, impaired color sensitivity and optic nerve disc edema were still present. On day 5, visual acuity improved to 5/5 in both eyes, no pain in eye movement, optic nerve papillary examination and color sensitivity also showed significant improvement.

Discussion

This case occurred in a child aged seven years with the main complaint of sudden blurred vision, seven days after the MR vaccine, accompanied by pain in eye movements and high fever that recurred. This is consistent with the clinical signs of ADEM with the manifestation of optic neuritis, characterized by sudden loss of visual acuity, occurring unilaterally, with visual acuity ranging from 0.8 to light perception. Ninety percent of sufferers complain of periocular pain of varying intensity associated with eye movement. Other clinical signs are Uhthoff's phenomenon, which is a sharp decrease in vision at high body temperature or during physical exercise, and the Pullfrich phenomenon in which objects moving in a straight line appear to have a curved path, this is the result of asymmetric conduction between the optic nerves. In optic neuritis there is also impaired color sensitivity, contrast, there is a central, paracentral or altitudinal scotoma, and a relative afferent defect in the pupillary reflex (RAPD). In 36 - 58% of patients, optic neuritis is accompanied by papilledema or swelling of the optic nerve head, but retrobulbar optic neuritis can also occur so that the papillary examination is normal [3] .

ADEM post vaccine was investigated as a result of reactions to vaccine adjuvants and molecular mimicry of viral proteins used for vaccination and body components. The most widely used vaccine adjuvant is aluminum. Aluminum interferes with lysosome function and stimulates the production and secretion of cytokines such as IL-1b, IL-18, and IL-33. Adjuvant vaccines function also provide protection against antigens, support the immune system's exposure to antigens for longer, and increase a stronger response to B cells and T cells. Molecular mimicry on vaccine components that resemble body components such as myelin in the central nervous system is one of the major immunopathogenic mechanisms in the demyelination process of the central nervous system post vaccination. Both of these mechanisms trigger immunological reactions that vary in several individuals including ADEM, myelitis, neuromyelitis, isolated optic neuritis, to multiple sclerosis [4].

The management strategy according to the ONTT guidelines is a study of the clinical profile of patients with optic neuritis, including assessing the effectiveness of steroid treatment, and examining the risk of developing multiple sclerosis (MS). The findings of the ONTT over the past 20 years have many implications. The ONTT recommends steroid therapy by intravenous injection of methylprednisolone 250 mg 4 times a day for 3 days, followed by tap off using oral prednisone 1 mg / kgBW / day for 11 days. The administration of megadose corticosteroid intravenously shows the effectiveness and safety of its use [5].

The prognosis for ADEM is generally good, depending on age, involvement of the central nervous system, and the time lag between symptom onset and initiation of therapy. Patients with older age and complaints that persist after administration of therapy show a worse prognosis of neurological sequelae. In cases of relapse and poor response to therapy, immunoglobulin and plasmapheresis therapy can be considered [6].

Conclusion

The manifestation of optic neuritis in ADEM is rare, but can occur at all stages of the disease. Early diagnosis and close monitoring of ADEM manifestations in the optic nerve are important to control the course of the disease, especially the progression of visual impairment.

Successful Management of Isolated Optic Neuritis in Acute Diseminated Encephalomyelitis: A Case Report

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Background: Optic neuritis is an inflammatory disease that results in demyelination of the optic nerve. Isolated Optic Neuritis in Acute Disseminated Encephalomyelitis is a rare case, this disease attacks the central nervous system. It is monophasic, and make various clinical manifestations. Early diagnosis of Isolated Optic Neuritis due to Acute Disseminated Encephalomyelitis is based on clinical manifestations accompanied by laboratory and radiological

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investigations. In Acute Disseminated Encephalomyelitis patient, radiological examination can shows normal result in the acute stage. This is a challenge in establishing the diagnosis of the disease. **Materials and Methods:** This is a case study in post-vaccine patient. Seven-year-old girl with complaints and typical clinical manifestations includes high fever, sudden decrease in vision accompanied by pain in the movement of the eyeball. Optic nerve papillary examination shows hyperemic color with firm borders, according to the results of optical coherence tomography examination. Laboratory examination showed an increase in white blood cell count and C-reactive protein value, and multiple hyperintense lesions in the subcortical temporo-parietal characteristic of Acute Disseminated Encephalomyelitis on Magnetic Resonance Imaging. **Results&Discussion :** Management guided by the Optic Neuritis Treatment Trial protocol accompanied by close observation showed significant clinical improvement. **Conclusion:** Optic neuritis in Acute Disseminated Encephalomyelitis is an acute and monophasic disease. The most common cause is inflammatory reaction to viral and molecular mimicry reactions of the vaccine component. Early diagnosis and proper management give good results and can prevent recurrence.
Keywords: Isolated Bilateral Optic Neuritis, Acute Disseminated Encephalomyelitis, central nervous system, vaccine.

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