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Siri Kavita Nalathamby, Jeyasakthy Saniasiaya, Jeyanthi Kulasegarah

Podwójne widzenie o nagłym początku u dziecka

Sudden onset of double vision in a child

Department of Otorhinolaryngology, Faculty of Medicine, Universiti Malaya, 50603 Kuala Lumpur, WP Kuala Lumpur, Malaysia
Adres do korespondencji: Jeyasakthy Saniasiaya, Department of Otorhinolaryngology, Faculty of Medicine, Universiti Malaya, Jalan Universiti, 50603 Kuala Lumpur, WP Kuala Lumpur, Malaysia, tel.: +6012-6841223, e-mail: shakthy_18@yahoo.com

ORCID iDs

1. Siri Kavita Nalathamby https://orcid.org/0000-0003-2735-547X
2. Jeyasakthy Saniasiaya https://orcid.org/0000-0003-1974-4379
3. Jeyanthi Kulasegarah https://orcid.org/0000-0001-8021-7864

Streszczenie

Wstęp: Nerw odwodzący, inaczej VI nerw czaszkowy (*cranial nerve six*, CN VI), jest najczęściej zajętym nerwem w przebiegu porażenia nerwu czaszkowego. Choć zidentyfikowano różne czynniki etiologiczne, nie ustalono jak dotąd powiązania z gammapatią monoklonalną. Celem pracy jest przedstawienie przypadku izolowanego porażenia nerwu czaszkowego u dziecka z gammapatią monoklonalną. **Opis przypadku:** W artykule przedstawiono przypadek zdrowej 6-letniej dziewczynki, u której nagle wystąpiły podwójne widzenie obuoczne, ból głowy i chwiejność chodu. Dalsze badania wykazały izolowane zapalenie lewego nerwu VI czaszkowego z towarzyszącą gammapatią monoklonalną, które leczono sterydami. **Omówienie:** Porażenie nerwu czaszkowego jest niepokojącym objawem, zwłaszcza u dzieci, i prawie zawsze wiąże się z patologiami wewnątrzczaszkowymi, takimi jak guz, wodogłowie i zapalenie opon mózgowo-rdzeniowych. **Wniosek:** Bezzwłoczne przeprowadzenie badań oraz wdrożenie odpowiedniego leczenia wiążą się z dobrym rokowaniem.

Słowa kluczowe: zapalenie nerwu wzrokowego, podwójne widzenie, porażenie nerwu odwodzącego, gammapatia monoklonalna

Abstract

Introduction: The abducens nerve, or cranial nerve six (CN VI), is the most common nerve involved in cranial nerve palsy. Various causes have been identified as the aetiology, but no association has been made with monoclonal gammopathy. We aim to describe isolated cranial nerve palsy in a child with monoclonal gammopathy. **Case report:** We report a case of a healthy 6-year-old girl who presented with sudden onset binocular diplopia, headache, and unsteady gait. Further investigation revealed isolated left sixth cranial nerve neuritis with monoclonal gammopathy, which was treated with steroids. **Discussion:** Cranial nerve palsy is an alarming sign, especially in children, and is almost always related to intracranial pathologies such as a tumour, hydrocephalus, and meningitis. **Conclusion:** Prompt investigations alongside relevant management result in a good prognosis.

Keywords: optic neuritis, diplopia, abducens nerve palsy, monoclonal gammopathy

INTRODUCTION

Sudden onset of diplopia in children is a paediatric emergency which may be an early sign of an acute vision loss or a life-threatening neurological disease[1]. Diplopia or double vision results from either extraocular muscle weakness or lesions within the cranial nerves, tracts or nuclei supplying these muscles. Traumatic, neoplastic, vascular, infectious, inflammatory, demyelination and idiopathic factors may be involved in diplopia secondary to cranial nerve palsy[2–4]. Isolated sixth nerve palsy due to microvascular anomaly is the most common aetiologic factor of diplopia in adults, especially in those with vasculopathy risk factors, such as diabetes and hypertension[5]. However, intracranial neoplasms have been advocated as the main culprit of sixth nerve involvement in the paediatric population[2,3,6,7]. Nonetheless, it is imperative to look for another rare cause of sudden onset diplopia in children.

AIM OF THE STUDY

To describe a case of isolated cranial nerve palsy in a child with monoclonal gammopathy.

CASE STUDY

A previously healthy 6-year-old girl presented with sudden-onset diplopia. Her mother claimed that the child complained of double vision after waking up, and she noticed asymmetry between her eyes. The child further claimed that her diplopia worsened upon lateral gaze and resolved when one eye was closed. Before diplopia, the child complained of a non-radiating headache the night before with no signs of meningism. Further history revealed that the child had an upper respiratory tract infection one week prior, which resolved with oral antipyretic. The mother also denied any seizures, abnormal behaviour, altered mental state or weakness of upper or lower limbs. There was also no sick contact or history of recent travelling.

Upon examination, the child appeared comfortable under room air. Her left eye extraocular movements were limited upon lateral gaze. She also had diplopia upon supero- and infero- temporal gaze, supero- and infero-nasal gaze, and superior gaze. The child was referred to an ophthalmology team for eye assessment. Her visual acuity was normal, and the relative afferent pupillary defect test was negative. Ear, nose and throat examination was normal. Flexible nasopharyngolaryngoscopy showed normal nasopharynx, oropharynx and larynx. Cranial nerve examination was unremarkable except for the left sixth cranial nerve. Cerebellar examination was also unremarkable.

RESULTS

A series of blood investigations were conducted, notably blood count, renal profile, liver function test, electrolytes, thyroid function test and viral screening, all within normal range. Chest radiograph showed no obvious abnormality as well. Contrast computed tomography of the brain showed normal brain parenchyma and ventricles with no space-occupying lesions. Brain magnetic resonance imaging (MRI) showed a slightly bulky left cavernous sinus with no focal enhancing parenchymal lesion or leptomeningeal enhancement (Fig. 1). Lumbar puncture and autoimmune investigations including immunoglobulin G index, albumin quotient, anti-nuclear factor, rheumatoid factor and anti-dsDNA were done and were within normal range. However, serum and cerebrospinal fluid electrophoresis showed monoclonal gammopathy. The child was diagnosed with abducens nerve neuritis with monoclonal gammopathy. A multidisciplinary team discussion was held and the child was started on a tapering dose of prednisolone. The parents were informed on treatment side effects such as nausea, vomiting, heartburn, dizziness, acne, mood changes, puffiness of the face, increased appetite and weight gain, increased blood sugar, and difficulty in sleeping, which they understood and agreed. Upon the third day of oral steroids, diplopia improved. Oral steroids were tapered over a period of 6 weeks. At her last clinic appointment, her symptoms resolved completely, and the child was kept under follow-up. She has been asymptomatic ever since.

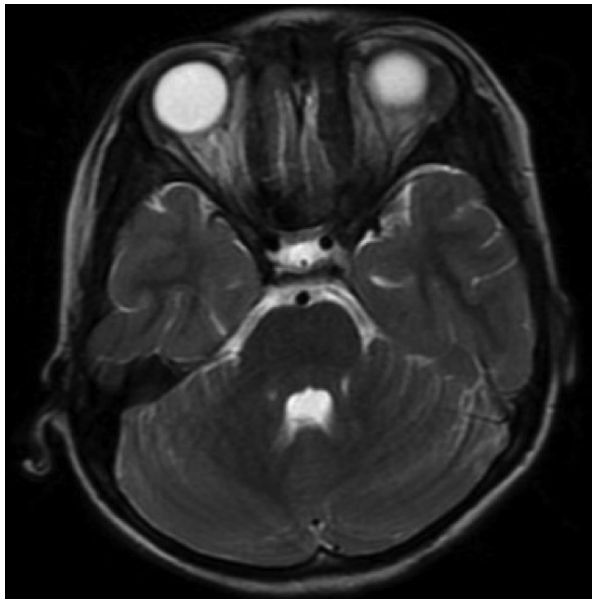

Fig. 1. Brain MRI showing no leptomeningeal enhancement

DISCUSSION

The abducens nerve, or the sixth cranial nerve, is the most common cranial nerve to be affected due to its long intracranial course. Its nucleus is located in the lower pons, just below the fourth ventricles on each sides. The course of this nerve begins intracranially as it arises from the cranial nerve nuclei and goes around the facial nerve nuclei to exit at the ventral surface of the pons on each side. Then it

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passes from the posterior cranial fossa to the middle cranial fossa, where it ascends the clivus to pass under the petroclinoid ligament in the Dorello's canal and enters the cavernous sinus. Next, it passes through the superior orbital fissure and into the orbit through the annular ligament of Zinn, and finally supplies the lateral rectus. The abducens nerve, unlike its fellow cranial nerves, is unsupported by the dural wall of the cavernous sinus. Although it is shorter in length compared to the trochlear nerve, its intracranial course makes it vulnerable to a variety of insults[8,9]. Knox et al. were the first to report cases of benign sixth cranial nerve palsies in children in 1967. In their study, they found that 12 of their patients had developed benign abducens palsy[9]. Benign abducens nerve palsy is defined as palsy that results not from an underlying sinister cause, such as an intracranial space-occupying lesion. Some authors also reported high female preponderance and left-sided predominance compared to the right side[10,11]. Nonetheless, the abducens nerve palsies are considered the commonest extraocular muscle palsy with a spontaneous recovery rate of 78.4%, as reported by King et al. in 1995[12].

Sixth nerve palsy may be a manifestation of other serious neurologic diseases besides an intracranial space-occupying lesion, such as idiopathic intracranial hypertension, multiple sclerosis, or anti-GQ1b syndrome. It is of paramount importance to identify the underlying pathology to initiate prompt treatment. A delay in treatment may cause further deterioration resulting in permanent disability.

A thorough history and physical examination are crucial to determine the nature of the disease. Therefore, early neurological and ophthalmological examination plays a pivotal role in establishing the diagnosis and line of management. In earlier literature, many authors do not recommend neuroimaging in cases of absence of other neurological findings as sixth nerve palsy is attributed to vasculopathy origin[13,14]. However, the vast advances in imaging have placed neuroimaging among early investigations to be carried out in children with sixth nerve palsy[15]. Additionally, routine blood investigations such as fasting blood glucose, complete cell count, serological studies along with blood pressure evaluation and lumbar puncture need to be performed hand-in-hand with other investigations[3,10,16].

Despite the advances in diagnostic neuroimaging and serological studies, the incidence of idiopathic sixth cranial nerve palsy has not changed significantly. This may be attributed to the lack of antibody testing in patients with anti-GQ1b syndrome or absence of an infectious trigger in post-infectious cases[17]. Detection of anti-GQ1b IgG antibody in a patient is a strong indication of an autoimmune disorder. It identifies a cluster of closely related syndromes with a common presentation of external ophthalmoplegia or cerebellar-like ataxia[18]. On the other hand, post-viral cranial nerve palsy is defined as the onset of cranial nerve palsy within two weeks of a documented upper respiratory tract illness[14]. These cranial nerve palsies are under-reported as they tend to resolve spontaneously. Nerve palsy which presents within one-month post-vaccination is known as abducens nerve palsy following vaccination. Although it is a known aetiology for abducens nerve palsy, its pathophysiology is still unknown, but could be related to autoimmune mechanisms or local demyelination[3,10]. It is also noteworthy that post-vaccination cranial nerve palsies tend to recur[7,17]. Our patient was believed to have post-viral abducens nerve neuritis with monoclonal gammopathy. However, there have been no reported cases of monoclonal gammopathy resulting in isolated abducens nerve palsy to date.

Our patient's investigations showed monoclonal gammopathies with no other clinical features besides isolated sixth cranial nerve palsy. Monoclonal gammopathy of undetermined significance (MGUS) indicates the presence of a monoclonal protein (M protein) in persons without evidence of amyloidosis, Waldenström's macroglobulinemia, multiple myeloma, or other related diseases[19]. Neuropathy is an uncommon presentation for patients with MGUS and is known to occur only in a small percentage of these patients, questioning the linkage of monoclonal proteins with neuropathy[20], which probably could indicate an incidental finding in our patient.

The recovery rate of isolated abducens nerve palsy has been reported to be around 2.3 months in children[13]. Having said that, the role of steroids in treating post-viral neuritis has shown promising results, with earlier recovery rates shown in our patients.

CONCLUSIONS

Sixth nerve palsy in the paediatric population poses a conundrum for the treating paediatrician. Although uncommon, abducens nerve palsy may be a manifestation of a serious underlying ailment. The highly variable aetiology requires prompt assessment and establishment of diagnosis so that suitable treatment can be rendered in a timely manner. Overall, the prognosis for benign recurrent sixth nerve palsy is excellent, with most patients recovering with complete resolution of symptoms.

Conflict of interest

All authors have no conflict of interest.

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