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A Perplexing case of isolated abducens nerve palsy in a primigravida woman: A case report

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ABSTRACT

Isolated abducens nerve palsy is a rare presentation in women during pregnancy. When an abducens nerve palsy is elicited in a pregnant woman, work-up should start with labs and neuroimaging to rule out mechanical and organic causes such as tumors, preeclampsia, and multiple sclerosis. This case report highlights a 35-year-old woman, gravida 1, para 0, who was sent to the local medical center by her ophthalmologist at 37 weeks of gestation due to a left-sided headache and blurry vision. Upon admission, work-up was negative for pre-eclampsia. Tick-borne disease panel and lumbar puncture were unrevealing. No other mechanical or lab abnormalities were elicited. Magnetic resonance venography revealed a diminutive left transverse sinus, left sigmoid sinus, and left internal jugular vein in comparison with the right, indicating a possible congenital variant. Labor was induced to see if this would alleviate the patient's abducens nerve palsy. After induction of labor and initiation of dexamethasone, the patient's sixth cranial nerve palsy began to improve.

1. Introduction

Cranial nerve palsies are rare neurological complications that have various causes, including viral infection, vascular compromise, neoplasm, or trauma [1]. Cranial nerve palsies are reported to occur more frequently in pregnant women and have been associated with late pregnancy, gestational hypertension, and preeclampsia [2]. Abducens nerve palsy is extremely rare in pregnancy [2]. It may be caused by lesions anywhere along the course of the abducens nerve, which originates at its nucleus in the dorsal pons and terminates at the innervation of the lateral rectus within the orbit. Abducens nerve palsy can result in partial or complete paralysis of the ipsilateral lateral rectus muscle, resulting in the inability to move the eye laterally and medial strabismus of the eye. Additional symptoms can include diplopia, headaches, and visual changes. This case report describes a 35-year-old pregnant woman (G1P0) with a past medical history of migraines with acute onset of left abducens nerve palsy in the 37th week of pregnancy.

2. Case Presentation

A 35-year-old woman, gravida 1, para 0, presented to her local medical center at 37 weeks of gestation due to new-onset horizontal diplopia, blurry vision, and decreased peripheral vision for two days, preceded by a left-sided headache for one week. Earlier on the day of presentation, her ophthalmologist diagnosed a left abducens nerve palsy, without findings of papilledema. The patient's past medical history was significant for infrequent migraines with visual aura.

The patient's 14-point review of systems was negative, aside from her visual symptoms and headache. Her family history was remarkable for Parkinson's disease in her maternal uncle and multiple system atrophy in her mother. Her mother had also experienced visual disturbances of unclear etiology late into her second pregnancy, which regressed after delivery.

Upon admission, the patient's blood pressure was 105/61 mmHg and urine dipstick was negative for proteinuria. Laboratory values showed a normal complete blood count and comprehensive metabolic panel. Neurological examination revealed no sensory or motor deficits. Ocular examination showed a mild, left-sided medial strabismus, bilateral pupils that were reactive to light, and a normal fundoscopic exam. The rest of her physical exam was within normal limits.

Neurology was consulted and recommended initiation of dexamethasone, magnetic resonance imaging (MRI) of the head, magnetic resonance angiography (MRA) of the brain, magnetic resonance venography (MRV) of the brain, tick-borne disease panel, and lumbar

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puncture. MRI revealed no acute intracranial abnormalities and MRA displayed a normal circle of Willis. MRV found the patient's left transverse sinus, left sigmoid sinus, and left internal jugular vein (IJV) to be very diminutive compared with the right, indicative of a congenital variant. There was no evidence of dural sinus thrombosis.

Tick-borne disease panel of the patient's serum was negative for Lyme disease IgG and ehrlichiosis species DNA. The patient had a non-reactive syphilis screening and a normal level of HSV 1 and 2 IgM. Lumbar puncture revealed a normal opening pressure. CSF evaluation was negative for HSV and Lyme DNA, as well as oligoclonal bands. Her CSF glucose was 73 mg/dL. CSF protein, IgG, angiotensin converting enzyme and albumin were all within normal limits. CSF cultures were negative for growth and white blood cells at 5 days.

Induction of labor was initiated on the second day of hospital admission with delivery occurring via cesarean section due to arrest of descent on the third day of admission. The patient received a trial of dexamethasone consisting of 10 mg on the second day of admission, followed by 8 mg on the third day, and 4 mg for three subsequent days. She was discharged three days after delivery, with mild improvement in her abducens nerve palsy. She was encouraged to follow up with neurology and her primary care provider.

3. Discussion

The presentation of a headache ipsilateral to the ocular cranial nerve palsy indicates a possible etiology of recurrent painful ophthalmoplegic neuropathy (RPON). This is a poorly understood relapsing and remitting condition, with an incidence of 0.7 per million [3]. Risk factors include a family or personal history of migraines [4]. Three different mechanisms have been proposed, the first being a compressive mechanism where dilation or edema of the walls of the internal carotid artery during a migraine may compress the surrounding ocular cranial nerves [4]. The second proposed mechanism involves ischemia of the nerves due to vasospasm of the vasa nevorum, leading to an impaired blood brain barrier, vasogenic edema, and subsequent cranial nerve palsy [5]. The last proposed mechanism is demyelinating/inflammatory which is based on the MRI findings of thickening and enhancement of the ocular cranial nerve [6,7]. According to the International Classification of Headache Disorders (ICHD), in order to meet diagnostic criteria for RPON, two episodes of an ipsilateral headache followed by palsy of one or more ocular cranial nerves are needed [8]. Since this was the patient's first episode of ocular cranial nerve palsy, she did not meet this diagnostic criterion. Finally, the ICHD mentions that nerve enhancement can be seen on MRI with gadolinium; however, this patient could not receive MRI with gadolinium as she was pregnant. The lack of gadolinium may explain why nerve enhancement was not seen.

Tolosa–Hunt syndrome presents as unilateral orbital pain with paralysis of one or more ocular cranial nerves, with symptoms that relapse and remit. It occurs as a result of idiopathic granulomatous inflammation in the cavernous sinus, superior orbital fissure, or orbital apex. Diplopia is typical and the pain can present several days prior to the nerve palsy [9]. This is a rare syndrome with an incidence of one in one million. A case of Tolosa–Hunt syndrome has been reported in a pregnant woman at 37 weeks of gestation who presented with a unilateral headache and an isolated abducens nerve palsy, with MRI showing enhancement of the cavernous sinus. This patient was treated successfully with prednisolone [10]. The patient also improved following glucocorticoid administration, supporting Tolosa–Hunt syndrome as a possible etiology.

Hypoplasia and aplasia of the transverse sinus are considered normal variants, with approximately 20–39% of people having these findings on the left side [11]. However, transverse sinus hypoplasia has been associated with increased risk of thrombosis of the ipsilateral transverse sinus [12]. Research has reported hypoplasia or aplasia of the transverse sinus to be associated with prolonged cerebral circulation time and impaired cerebral blood flow autoregulation [11]. One case series of pediatric patients with cerebral venous thrombosis demonstrated that patients with contralateral hypoplastic venous sinuses were more likely to develop elevated intracranial pressure (ICP) and resultant abducens nerve palsy [13]. Hypoplastic venous sinuses resulted in elevated ICP and cranial nerve palsies only if thrombosis was also present. The patient had a normal ICP and no thrombus; therefore, it is not likely that a hypoplastic venous drainage system contributed to her palsy.

Multiple sclerosis (MS) is an autoimmune disease that targets the central nervous system and results in inflammation, demyelination, and neuronal loss [14]. Many visual symptoms accompany MS, including vision loss and diplopia due to inflammation of the optic nerve [14]. Although MS may be one of the differential diagnoses to explain the present patient's abducens nerve palsy, MRI without gadolinium contrast appeared to be non-revealing and her CSF was negative for oligoclonal bands.

Preeclampsia is a disorder that can affect pregnant women and visual symptoms occur in 25–40% of patients, with symptoms including decreased vision and diplopia [15]. Studies have estimated that 30–100% patients afflicted with preeclampsia have retinal arterial and structural abnormalities including retinal vascular anomalies such as arteriole narrowing, tortuosity and segmental retinal artery vasospasm [15]. However, work-up for preeclampsia in the patient proved to be unremarkable, including liver function tests, renal function, and complete blood count. She had no hypertensive episodes or significant proteinuria.

Miller Fisher syndrome (MFS) is a variant of Guillain Barre syndrome and is characterized by a triad of ophthalmoplegia, ataxia and areflexia [16]. Research has shown that IgG antibodies to GQ1b ganglioside are very sensitive for a diagnosis of MFS [16]. MFS seems to predominantly affect the III, IV and VI cranial nerves and it is believed that these specific cranial nerves are comprised of vast amounts of GQ1b ganglioside [16]. Visual symptoms associated with MFS may be caused by antibodies acting on the neuromuscular junction that is located between the cranial nerves and the ocular muscles involved [16]. A retrospective study analyzed 100 cases of unilateral abducens nerve palsy, with 25% of the patients being positive for IgG antibodies against GQ1b ganglioside [17]. All 100 patients were back to baseline after 6 months with treatment including steroids, intravenous immunoglobulin (IVIg), plasmapheresis, plasmapheresis followed by IVIg or no specific treatment [17]. Due to the partial autoimmune nature of this disease, it would be insightful to look at other autoimmune-related conditions in the patient, including antibodies against GQ1b.

4. Conclusion

In summary, abducens nerve palsy, particularly in pregnancy, is a rare finding. There are several mechanisms that have been linked to abducens nerve palsy, including recurrent painful ophthalmoplegic neuropathy, Tolosa–Hunt syndrome, hypoplasia or aplasia of the transverse sinus with resultant increased intracranial pressure, preeclampsia, multiple sclerosis and Miller Fisher syndrome. This case report details the history of a young, healthy primigravida woman with an isolated abducens nerve palsy in which the mechanism still remains unclear. Although lab results and imaging may aid in ruling out known causes of abducens nerve palsy, there are still unknown causes of this condition. This case report illustrates the complexity of abducens nerve palsy and encourages future research to be conducted regarding the cause behind isolated cases.

Contributors

Johnna M. Caputo contributed to patient care, conception of the case report, acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Marianna Catege contributed to patient care, conception of the case

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Ishani Dev contributed to patient care, conception of the case report, acquiring and interpreting the data, drafting the manuscript, undertaking the literature review and revising the article critically for important intellectual content.

Benjamin Souferi contributed to patient care, conception of the case report, acquiring and interpreting the data, undertaking the literature review and revising the article critically for important intellectual content.

Adele El Kareh contributed to patient care and revising the article critically for important intellectual content.

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Conflict of interest statement

The authors declare that they have no conflict of interest regarding the publication of this article.

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