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Case Study

Post viral acute cerebellar ataxia following chickenpox (Varicella) in a toddler.

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Abstract

Background: Varicella-zoster virus (VZV) is one of the most common infections of childhood causing chickenpox, which in a majority of cases is self-limiting without any complicating consequences. However, in a meagre 0.01-0.03% cases, chickenpox may lead to serious neurological complications. Among them is acute cerebellar ataxia, which presents with the characteristic sign of broad-based gait abnormality, progressing gradually over the course of days, the mechanism of which is still debated upon. As a treatment option, the use of antiviral remains controversial, and some recommend it for better prognosis while others do not base on its autoimmune pathogenesis. This case is being communicated due to its rarity.

Methodology: A case of a two-year-old female with a recent history of chickenpox eruption presented with the complaint of progressive loss of normal gait and difficulty in talking followed by generalized to-and-fro movements of her body leading to a diagnosis of post-viral acute cerebellar ataxia. There was no significant history of any past illnesses. Bacille Calmette Guerin (BCG) vaccine was given at birth, later followed by a single dose of pentavalent (Penta) vaccine.

Results: Central nervous system (CNS) examination showed ataxic gait, pupils bilaterally equally reflective to light (BERL), nystagmus, dysmetria, and decreased muscle tone. Laboratory investigations were within the normal range. Imaging modalities that were all normal excluded other causes, including metabolic, tumors, and toxins. The patient was empirically treated with IV acyclovir BD for one week.

Conclusion: As the rash of chickenpox resolves spontaneously in most cases, it is important that children infected with VZV should be closely observed for any neurological symptoms for timely and effective intervention.

Keywords

Cerebellar Ataxia, Toddler, Varicella-Zoster, Chickenpox.

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Introduction

VZV infection-causing chickenpox is characterized by vesicular dermal exanthemas on the entire body surface, usually occurring in preschool and school-going children, which resolves spontaneously in most cases. Neurological manifestations, although which are rare, can probably occur in the 0.01% to 0.03% of patients, including encephalitis, demyelinating polyradiculoneuropathy, meningitis, Reye's syndrome, peripheral motor neuropathy, transverse myelitis, and optic neuritis[1]. Acute cerebellitis is the second most frequent complication reported after encephalitis among the uncommon complexities of chickenpox[1, 2]. We report a case of an unvaccinated chickenpox infection in which the patient did not fully develop generalized itchy vesicular lesions presented with cerebellar ataxia, which occurs in 1 in 4000 of VZV cases and discusses the controversial management and strategies of the patient briefly. The clinicians, physicians, and providers need to be reminded and explained about the pathophysiology of this acute prognostic complication of the said condition, which encompasses autoimmunity[3].

Post-varicella acute cerebellar ataxia (PVACA) is a common neurological complication of varicella virus, occurring once in 4000 varicella cases among children who are younger than 15 and especially those under 14 years of age, even in the post-vaccine era[3]. The typical clinical presentations of varicella and herpes zoster are readily recognized by many experienced clinicians, specialists, and physicians. However, atypical clinical presentations, symptoms and uncommon or rare complications of these diseases can pose a significant diagnostic and therapeutic challenges. The issues that are related to the management of varicella become especially complex and complicated when varicella involves pregnant women, immunocompromised, chronic infectious and conditions or susceptible neonates[4]. Many children, including toddlers and infants with acute ataxia, are seen within several days of symptom onset, usually because of refusal to walk or the sudden development of a wide-based, "drunken" gait. Caregivers and providers less commonly remark on the unsteadiness of arm and leg movements, truncal titubation, and dysarthria[5]. The deep tendon reflexes of the upper and lower extremities can be pendular, with slowed contraction and relaxation phases, although this is quite often difficult to appreciate and comprehend.

Childhood ataxia can be diagnostically and therapeutically approached by consideration of the temporal course and presence or absence of associated neurologic abnormalities. In all forms of childhood ataxia, the outcome is largely determined by etiology[5]. In the clinical practice, a thorough history and physical examination are far more likely to identify the etiology of acute ataxia than is a series of "screening" investigations, including laboratory workup and radiological imaging modalities[5]. PVCA is usually associated with a favourable prognosis; however, neurological complications can occur. The real utility and significance of acyclovir or valacyclovir treatment and the central nervous system imaging studies in these children remain controversial[6]. Acute ataxia in childhood has a wide range of causes. The differential diagnosis for acute ataxia in children is broad and includes toxicological, infectious, structural, and metabolic causes[7]. Acute cerebellar ataxia (sudden onset of truncal ataxia and gait disturbances) usually follows a benign mode of illness (25% varicella). It is also described after the vaccination, like measles, mumps, rubella (MMR), and VZV[7].

Case Presentation

A four-year-old female child, weighing 15.8 kg, presented to the pediatric department of Dr. Ruth KM Pfau, Civil Hospital Karachi (CHK) in January 2019 with the complaint of progressive loss of normal gait and difficulty in talking followed by generalized to-and-fro movements of her body for the past one day. She had a previous history of chickenpox (varicella) for the past four days. According to her mother, the child was alright four days back when she developed vesicles over the abdomen, which later involved her face and extremities. The rash was associated with high-

grade fever, which was relieved by antipyretics. Eventually, the mother noticed that the child became unable to walk and was falling while walking due to progressive loss of balance. There was no significant history of any past illnesses, hospital admissions, or blood transfusions. The child was the third product of non-consanguineous marriage with no history of any chronic illnesses in the family. The child was delivered at term gestation via cesarean section. She was not completely vaccinated except for a dose of the BCG vaccine given at birth later, followed by a single dose of pentavalent (Penta) vaccine. Developmental history was found appropriate for her age. The nutritional status of the child was also reported to be satisfying.

Management & Results

On examination (O/E), the child was found conscious but clumsy. Initial vitals included blood pressure (BP) 100/65 mmHg, a regular pulse of 126 beats/min and a respiratory rate of 24 breaths/min with no febrility. Oxygen saturation was 99% with bilaterally clear chest, and normal heart sounds on auscultation. The abdomen was soft and non-distended with scabs of resolving chickenpox vesicles, which were also present on her face and extremities. Upon CNS examination, the child was found to have ataxic gait, pupils BERL, nystagmus, dysmetria, and decreased muscle tone with normal reflexes in all limbs. Generalized tremors, especially in arms when lifted against the gravity and in the head and trunk in an upright position, were noticed. All other systems had no significant findings. Laboratory investigations came up with a hemoglobin (Hb) of 9.5 gm/dl, mean corpuscular volume (MCV) of 62.5 fl [Normal (N) = 76-96], total leukocyte count (TLC) of 9.69 x 103/µL (N = 4-11), neutrophils of 26.7% (N = 50-75%), lymphocytes of 54.1 (N = 20-40%) and a platelet count (PLT) of 524 x 103/µL (N = 150-400). The level of C-reactive protein (CRP) was found to be 3.6 mg/L (N = <5). This is shown in Table 1 below.

Table 1: Laboratory investigation and results

Laboratory Test	Result
Hemoglobin	9.5 mg/dl
Mean Corpuscular Volume	62.5 fl
Total Leucocyte Count	9.69 x 103/µl
Neutrophils	26.7%
Lymphocyte	54.1%
Platelet count	524 x 103/µl
C-Reactive Proteins	3.6 mg/L
International Normalized Ratio	0.93
Prothrombin Time	9.8 s
Activated Partial Thromboplastin Time	23.5 s

A computed tomography (CT) scan of the brain was done to eliminate the presence of any tumor in the posterior fossa or acute hemorrhage. Other imaging modalities included electroencephalograph (EEG) and magnetic resonance imaging (MRI), which were merely performed to exclude more severe diseases, such as neuroblastoma, acute labyrinthitis, drug intoxication, or any metabolic disease. A more complicated test, lumbar puncture, was not preferred. Finally, based on the absence of any significant finding in the CT scan, EEG, or MRI, along with the typical clinical picture, a diagnosis of post-viral acute cerebellar ataxia (ACA) was made.

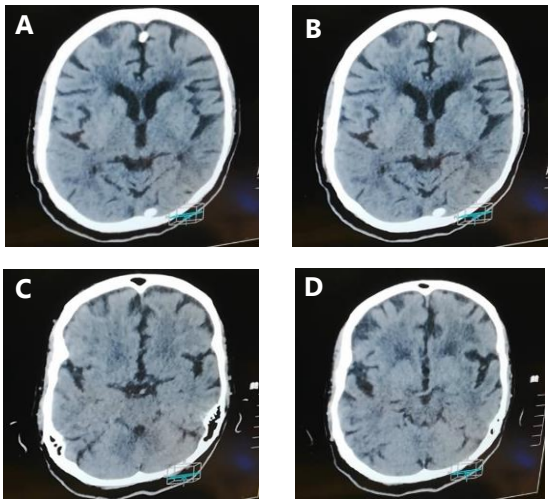

Figure 1: Computed tomography scans without bleed, lesions or masses

Figure 2: Magnetic resonance imaging depicted regions of the brain without any obvious bleed, lesion or masses

The patient was empirically treated with IV acyclovir BD for one week. After an established diagnosis, she was discharged from the hospital and needed no follow-ups.

Discussion

Varicella is an alpha herpes virus that only infects humans with no animal reservoir and typically affects T lymphocytes, epithelial cells, and ganglion[8]. Acute cerebellar ataxia (ACT) commonly occurs in the age group of two to seven-year-old children. Clinically it can present with a prodromal illness period accompanied with or without eruptive xanthomas, severe acute ataxic gait, dissymmetry, dysdiadochokinesia, slurred speech, and horizontal nystagmus in 50% of cases, usually developing within 72-hours in previously well child[3]. However, the most characteristic presentation is broad-based gait abnormality, which develops gradually over days, as seen in our case. The pathogenesis behind the development of neurological symptoms is still debated upon. Usually, the child has infection one or two weeks

before the onset of symptoms, the evidence shows inflammatory cells in cerebrospinal fluid but no organisms, indicating an active infection. In some cases, direct viral invasion in the cerebellum or immunological process elicited by the infection itself or vaccination can cause a cross-reaction between antibodies and cerebellar antigens contributing to the cerebellar demyelination[1, 3, 9].

Routine blood tests, including complete blood count, PT, INR, were normal in our study also as evidenced by Rudrajit et al[3]. Cerebellar signs found on examination makes imaging of head mandatory in which MRI is preferred over CT to clearly visualize posterior fossa. Post varicella acute cerebellar ataxia was established in our case when imaging modalities excluded all other neurological causes such as brain tumors, cerebellar contusion, subdural hematoma, cerebellar infarction, or hemorrhage as the study done by Monique et al. & Elizabeth et al. found out CT and MRI of the brain is normal in most children with post-infectious ataxia[3, 6]. Lumbar puncture was not recommended in our case because the presence of a CSF abnormality rarely affects prognosis or treatment plan[7]. Due to financial restraints, serological testing of varicella-zoster IgG and IgM was not performed. As recommended, no further workup is needed if acute cerebellitis develops following the rash to establish a diagnosis[8]. According to Camacho-Badilla et al., the use of antiviral remains controversial[9]; some recommend it for better prognosis while others do not base on its autoimmune pathogenesis[8]. Symptoms spontaneously resolve within a week from the onset[8]. The same study suggested otherwise against the utility of brain imaging. However, we started acyclovir, keeping in mind disease severity and to prevent its progression until the symptoms were improved. There is no inclusion of universal vaccination against varicella in the immunization schedule in Pakistan. Routine two-dose vaccination scheme, one to prevent varicella and one to prevent postherpetic neuralgia, should be introduced in the pediatric vaccination scheme to reduce the rate of incidence of disease. However, it could also result in severe acute cerebellar ataxia by inducing severe immune reactions[9, 10]. In the reviewed literature, we didn't find a case in which very few vesicular eruptions on the child were noted before the onset of symptoms, as presented by our case.

Conclusion

Although the rash of chickenpox resolves spontaneously in most cases, it is crucial for parents and guardians to be aware of the possible serious complications of this common infection. A child infected with VZV should be closely observed for any neurological symptoms for timely and effective intervention. Vaccination scheme against VZV should be revised and implemented to reduce the rate of incidences, particularly in third world countries like Pakistan.

Conflicts of Interest

None.

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Consent

Written consent was obtained from the subject.

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References

1. Girija AS, Rafeeque M, Abdurehman KP. Neurological complications of chickenpox. Ann Indian Acad Neur. 2007;10(4):240-224.
2. Paul R, Singhania P, Hashmi MA, Bandyopadhyay R, Banerjee AK. Post chicken pox neurological sequelae: Three distinct presentations. J Neurosci Rural Pract. 2010; 1(2):92.
3. Salas AA, Nava A. Acute cerebellar ataxia in childhood: initial approach in the emergency department. Emerg. Med J. 2010; 27(12):956-957.
4. Gnann Jr JW. Varicella-zoster virus: atypical presentations and unusual complications. J. Infect. Dis. 2002; 186(Supplement_1):S91-98.

5. Ryan MM, Engle EC. Topical review: acute ataxia in childhood. J Child Neurol. 2003; 18(5):309-316.
6. Camacho-Badilla K, Méndez I, Soriano-Fallas A, Ulloa-Gutiérrez R, Avila-Aguero ML. Postvaricella cerebellar ataxia in children in Costa Rica. In Anales de pediatria (Barcelona, Spain: 2003) 2008 (Vol. 68, No. 1, p. 49).
7. van der Maas NA, Vermeer-de Bondt PE, De Melker H, Kemmeren JM. Acute cerebellar ataxia in the Netherlands: a study on the association with vaccinations and varicella zoster infection. Vaccine. 2009;27(13):1970-1973.
8. Bozzola E, Bozzola M, Tozzi AE, Calcaterra V, Longo D, Krzystofiak A, Villani A. Acute cerebellitis in varicella: a ten year case series and systematic review of the literature. Ital. J. Pediatr. 2014; 40(1):57.
9. Koyuncu OO, Hogue IB, Enquist LW. Virus infections in the nervous system. Cell Host Microbe. 2013;13(4):379-393.
10. Connolly AM, Dodson WE, Prensky AL, Rust RS. Course and outcome of acute cerebellar ataxia. Ann. Neurol. 1994; 35(6):673-679.