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Fatal Neonatal Presentation of Congenital Varicella Syndrome Following First-Trimester Maternal Infection in a Resource Limited Setting: A Case Report

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Abstract

Background: Congenital varicella syndrome (CVS) is a rare but severe fetal consequence of primary maternal varicella-zoster virus (VZV) infection during early pregnancy. Characterized by cutaneous, musculoskeletal, ocular, neurologic, and visceral abnormalities, CVS is associated with substantial morbidity and mortality. Although uncommon, the syndrome remains clinically relevant in regions where varicella vaccination and post-exposure prophylaxis are not routinely available. **Case Presentation:** We report the case of a term male neonate born to a 35-year-old gravida 3 para 3 mother with documented clinical varicella infection at approximately 12 weeks' gestation. The maternal illness was characterized by a generalized vesicular rash and fever and was managed conservatively without antiviral therapy or varicella-zoster immunoglobulin (VZIG). The infant was delivered by elective cesarean section at term and presented shortly after birth with poor cry, respiratory distress, extensive cicatricial skin lesions, limb contractures, bilateral talipes equinovarus, microphthalmia, bilateral corneal opacities, micropenis, rudimentary scrotum, and bilateral cryptorchidism. The diagnosis of congenital varicella syndrome was made clinically based on the characteristic phenotype and documented maternal history, as virologic confirmation was not available. Despite supportive management, the infant developed recurrent apneic episodes and died within six hours of admission. **Discussion:** This case illustrates the classical phenotypic spectrum of CVS and the severe consequences of maternal varicella infection during the critical period of fetal organogenesis. The coexistence of extensive cutaneous, ophthalmologic, musculoskeletal, and genital abnormalities, together with early neonatal death, represents a particularly severe presentation. Diagnostic and therapeutic challenges were made worse

by limited access to virologic testing, fetal surveillance, antiviral therapy, and VZIG prophylaxis. **Conclusion:** Congenital varicella syndrome is a potentially fatal yet preventable condition. Improved screening, varicella vaccination strategies, timely post-exposure prophylaxis, and enhanced awareness among healthcare providers are critical to reducing the burden of CVS in low- and middle-income countries.

Keywords

Congenital Varicella Syndrome, Varicella-Zoster Virus, Maternal Infection, Congenital Anomalies, Neonatal Mortality, Resource-Limited Settings

1. Introduction

Congenital varicella syndrome (CVS) results from transplacental transmission of varicella-zoster virus (VZV) following primary maternal infection during early pregnancy [1] [2]. The syndrome is most commonly associated with maternal infection occurring between 8 and 20 weeks' gestation, a period corresponding to critical stages of fetal organogenesis and neural development [1]. Although the overall risk of CVS following maternal varicella infection is relatively low, estimated at approximately 0.4% - 2.0%, the resulting fetal consequences may be severe and irreversible [1].

The teratogenic effects of VZV are believed to result from viral replication within fetal tissues, particularly neuroectodermal and mesenchymal structures, leading to tissue necrosis, vascular injury, growth restriction, and segmental developmental abnormalities [3] [4]. The classical phenotype includes cicatricial skin scarring in a dermatomal distribution, limb hypoplasia or contractures, ocular abnormalities, central nervous system involvement, and growth impairment [1] [3] [5]. Less frequently reported manifestations include gastrointestinal, cardiovascular, and genitourinary anomalies [3] [5].

The introduction of routine varicella vaccination in many high-income countries has led to a marked decline in maternal varicella infection and CVS [6]. Unfortunately, the condition remains clinically important in low-and middle-income countries where vaccination is not universally implemented and access to post-exposure prophylaxis is limited [6]. Published reports from sub-Saharan Africa remain scarce, and fatal neonatal presentations are infrequently documented [3] [5].

We report a severe case of clinically diagnosed congenital varicella syndrome in a term neonate born following documented maternal varicella infection during the first trimester of pregnancy [1] [4]. The case was characterized by extensive multisystem involvement, including cutaneous, musculoskeletal, ophthalmologic, and genitourinary abnormalities, and resulted in early neonatal death [1] [5]. This report emphasizes the importance of preventive maternal health strategies in resource-constrained settings [2] [6].

2. Case Presentation

A male neonate was delivered at 38 weeks' gestation by elective cesarean section to a 35-year-old gravida 3 para 3 mother at a private health facility in Rivers State, Nigeria. The pregnancy was notable for a documented maternal illness consistent with primary varicella infection at approximately 12 weeks' gestation (first trimester). The mother reported the sudden onset of fever (38.5˚C), weakness, headache, and a generalized pruritic vesicular rash involving the face, trunk, and extremities in a centripetal distribution. The rash progressed from macules to papules to vesicles to crusts over 5 - 7 days. She was evaluated at a healthcare facility where a clinical diagnosis of chickenpox was made based on the characteristic vesicular rash pattern and centripetal distribution, typical of primary varicella-zoster virus (VZV) infection.

However, laboratory confirmation with varicella-zoster virus (VZV) serology (IgM/IgG) or polymerase chain reaction (PCR) testing of vesicular fluid was not performed due to limited diagnostic capacity at the presenting facility. The maternal illness was managed conservatively with symptomatic treatment (paracetamol for fever and calamine lotion for pruritus) at home following reassurance at the initial health facility that the condition would resolve spontaneously. Neither antiviral therapy (acyclovir) nor varicella-zoster immunoglobulin (VZIG) was administered.

The mother had no documented history of prior varicella infection or varicella vaccination. No other significant antenatal complications were reported. An early obstetric ultrasound scan at 10 weeks was reportedly normal, while a late third-trimester scan at 34 weeks noted unspecified fetal structural abnormalities without specific characterization.

The infant was born with a birth weight of 2.2 kg (10th percentile, consistent with fetal growth restriction). Apgar scores were 5 and 7 at one and five minutes, respectively. Detailed delivery room resuscitation records were unavailable. According to maternal report, the infant required resuscitation with bag-mask ventilation due to poor cry and respiratory effort at birth.

The infant was referred to the Special Care Baby Unit of the University of Port Harcourt Teaching Hospital at approximately nine hours of life because of poor cry, multiple congenital abnormalities, respiratory distress, and extensive skin lesions.

On admission, the infant appeared acutely ill and dysmorphic. He was hypothermic with an axillary temperature of 34.8˚C and exhibited signs of respiratory distress, including intercostal and subcostal recessions. Respiratory rate was 40 breaths per minute, and oxygen saturation was 93% while receiving supplemental oxygen. Bilateral lower lung zone crepitations were present on auscultation. Heart rate was 120 beats per minute, and no cardiac murmur was detected.

Dermatologic examination demonstrated multiple hypopigmented cicatricial scars interspersed with fresh ulcerative lesions involving the face, trunk, upper limbs, and lower limbs (**Figure 1**, **Figure 2**). The lesions displayed a segmental

distribution and were suggestive of healed intrauterine skin injury. Cutaneous lesions are among the most recognizable manifestations of CVS and are reported in approximately 70% of affected infants [1] [5].

Musculoskeletal abnormalities included fixed adduction deformity of the left hip, right knee flexion contracture, and bilateral talipes equinovarus (**Figure 1**). Limb abnormalities may include hypoplasia, contractures, muscular atrophy, syndactyly, or talipes deformities [1] [3].

Ophthalmologic examination revealed bilateral microphthalmia, dense corneal opacification, and non-reactive opaque pupils. Ophthalmologic examination was performed using a pen torch. Findings included bilateral microphthalmia (globes appeared small on external inspection), bilateral dense corneal opacification with pupils completely opaque and non-reactive to light bilaterally. Ocular manifestations include microphthalmia, cataracts, chorioretinitis, optic nerve atrophy, and corneal abnormalities [1] [3] [5].

Genital examination demonstrated micropenis with a stretched penile length of 1.6 cm, rudimentary scrotal development, and bilateral cryptorchidism (**Figure 1**). Genitourinary abnormalities are less frequently described in the literature than dermatologic or musculoskeletal manifestations but have been reported in association with severe fetal VZV infection [3] [5]. Cryptorchidism, genital hypoplasia, neurogenic bladder dysfunction, and abnormalities of external genital development have been documented in isolated case reports and small case series [3] [5].

Primitive neonatal reflexes were globally depressed. The depressed primitive reflexes and moderate birth asphyxia may represent the cumulative effects of systemic compromise at birth [1] [5].

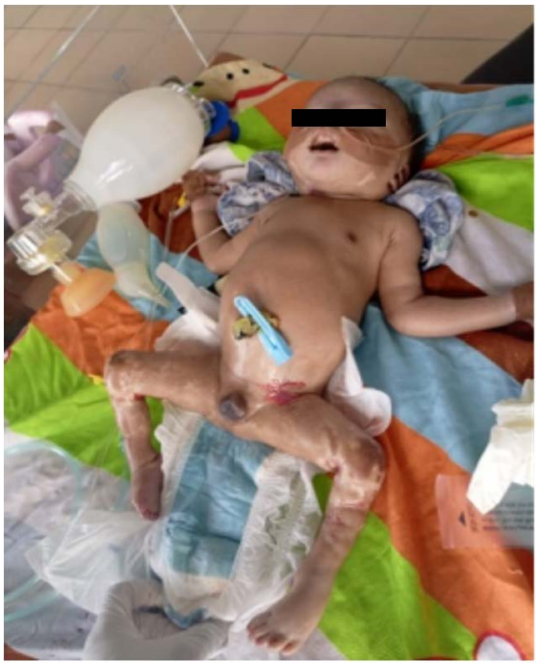

Figure 1. Image showing micropenis, rudimentary scrotum and limb abnormalities.

Figure 2. Image showing cicatricial skin lesions.

2.1. Initial Laboratory and Diagnostic Evaluation

Initial laboratory evaluation was limited due to the infant's rapidly deteriorating clinical condition and early death. The parents were also unable to afford the other requested investigations. Random blood glucose done was 4.8 mmol/L (within normal range for neonate).

2.2. Diagnosis

A clinical diagnosis of congenital varicella syndrome (CVS) was made based on: 1) documented maternal history of primary varicella infection at 12 weeks' gestation (first trimester, within the critical 8 - 20 week window for CVS teratogenesis); and 2) the characteristic constellation of neonatal findings including cicatricial skin lesions in dermatomal distribution, limb deformities (bilateral talipes equinovarus, knee contracture, hip adduction), ocular abnormalities (bilateral microphthalmia, corneal opacities), and genitourinary anomalies (micropenis, rudimentary scrotum, bilateral cryptorchidism) [1] [3] [4].

Virologic confirmation was not available. Specifically:

- VZV PCR testing of neonatal blood, cerebrospinal fluid, or skin lesion swabs
- Neonatal VZV IgM/IgG serology
- Placental histopathology with VZV immunohistochemistry
- Postmortem examination with tissue VZV PCR
- The diagnosis is therefore classified as ***PROBABLE/ CLINICAL congenital varicella syndrome*** according to established CDC case definitions for congenital varicella syndrome, which recognize clinical diagnosis when characteristic phenotype is present with compatible maternal history in settings where viro-

logic testing is unavailable [3] [7] [8].

Supportive management included thermal regulation, intravenous fluid administration, oxygen therapy, empiric broad-spectrum antibiotics, and continuous cardiorespiratory monitoring. Despite intensive supportive measures, the infant developed recurrent apneic episodes with progressive cardiorespiratory compromise and died approximately six hours after admission.

2.3. Differential Diagnosis

Several conditions were considered in the differential diagnosis of this neonate with cicatricial skin lesions, limb deformities, ocular abnormalities, and multisystem anomalies:

Congenital Cytomegalovirus (CMV) Infection:

Congenital CMV can present with intrauterine growth restriction, microcephaly, chorioretinitis, hearing loss, hepatosplenomegaly, and periventricular calcifications [3] [5]. However, characteristic cicatricial dermatomal skin scarring and limb contractures (often asymmetric) are uncommon in congenital CMV. Additionally, the documented maternal varicella infection at 12 weeks strongly favors CVS [1] [5].

Congenital Herpes Simplex Virus (HSV) Infection:

Congenital HSV is extremely rare and typically presents with skin scarring, microcephaly, chorioretinitis, and microphthalmia [3] [5]. However, HSV infection more commonly produces vesiculobullous lesions at birth rather than healed cicatricial scars, oftentimes without limb hypoplasia/contractures. The maternal history of typical chickenpox (not genital herpes) further favors CVS [1] [3].

The combination of 1) documented maternal first-trimester varicella infection, 2) cicatricial skin lesions in dermatomal distribution, 3) limb hypoplasia/contractures, 4) ocular abnormalities, and 5) genitourinary anomalies is strongly in keeping with CVS as described in systematic reviews and case series [1] [2] [5].

3. Discussion

Congenital varicella syndrome (CVS) is a rare but severe consequence of transplacental transmission of varicella-zoster virus (VZV) following primary maternal infection during pregnancy [1] [4]. The syndrome was first described in 1947 and remains one of the classical viral embryopathies associated with early intra-uterine infection [1] [3]. Although widespread childhood immunization programs have markedly reduced the incidence of maternal varicella infection in many high-income countries, CVS continues to occur sporadically, particularly in regions where routine varicella vaccination is not universally implemented or where access to preventive maternal healthcare remains limited [1] [4] [6].

3.1. Epidemiology and Risk Factors

The risk of CVS is closely related to gestational age at maternal infection [1] [5]. The highest fetal risk occurs when primary maternal varicella develops between 8 and 20

weeks of gestation, corresponding to critical periods of organogenesis and fetal neural development [1] [5]. Systematic reviews estimate the overall risk of CVS following maternal infection during the first 20 weeks of pregnancy to be approximately 0.4% - 2.0%, although reported estimates vary according to study design and population characteristics [1] [5]. Cases occurring after 20 weeks are uncommon, and fetal infection beyond 28 weeks is considered exceptionally rare [3] [5].

3.2. Pathogenesis and Clinical Spectrum

The pathogenesis of CVS is not completely understood but is believed to involve direct viral replication within fetal tissues, particularly neuroectodermal and mesenchymal structures [3] [4]. Viral-induced tissue necrosis, vascular compromise, inflammatory injury, and impaired cellular differentiation contribute to the characteristic pattern of developmental abnormalities observed in affected infants [3] [4]. The resulting embryopathy is often segmental, reflecting viral involvement of specific dermatomes and developing neural pathways [1] [4]. This mechanism explains the distinctive coexistence of cutaneous scarring, limb hypoplasia, ocular abnormalities, and neurologic dysfunction that characterizes the syndrome [1] [4].

Cutaneous lesions are among the most recognizable manifestations and are reported in approximately 70% of affected infants [1] [3]. These lesions reflect healed intrauterine viral injury and frequently appear as depressed, hypopigmented scars arranged along dermatomal lines [1] [3]. Limb abnormalities may include hypoplasia, contractures, muscular atrophy, syndactyly, or talipes deformities [1] [3] [5]. Ocular manifestations include microphthalmia, cataracts, chorioretinitis, optic nerve atrophy, and corneal abnormalities [1] [3] [5]. Neurologic complications may include cortical atrophy, seizures, developmental delay, autonomic dysfunction, and intellectual disability among survivors [1] [3]. The combination of these findings following documented maternal first-trimester varicella infection strongly supports the diagnosis of congenital varicella syndrome.

Genitourinary abnormalities are less frequently described in the literature than dermatologic or musculoskeletal manifestations but have been reported in association with severe fetal VZV infection [3] [5]. Cryptorchidism, genital hypoplasia, neurogenic bladder dysfunction, and abnormalities of external genital development have been documented in isolated case reports and small case series [3] [5]. The coexistence of micropenis, rudimentary scrotum, and bilateral cryptorchidism in our patient therefore represents an unusual and severe manifestation of extensive fetal developmental disruption [3] [5].

3.3. Prognosis and Outcomes

The prognosis of CVS remains poor, particularly among infants with extensive multisystem disease [1] [5]. Studies suggest that approximately 30% of affected infants die during infancy, with mortality often resulting from respiratory complications, severe neurologic dysfunction, recurrent aspiration, or multisystem organ involvement [1] [3]. Survivors frequently experience long-term neurodevel-

opmental impairment, visual disability, orthopedic complications, and chronic neurologic sequelae [1] [3]. The rapid deterioration observed in our patient, resulting in death within hours of admission, underscores the potentially devastating nature of severe congenital infection [1] [5].

4. Global Burden and Low-Resource Settings

The present case highlights critical public health challenges in low- and middle-income countries (LMICs) [6]. Varicella vaccination is not routinely included in national immunization schedules in most African countries, including Nigeria, leaving susceptible women vulnerable to primary infection during pregnancy [6]. Key barriers include limited access to preconception counseling and serologic screening, lack of VZIG availability, restricted access to antiviral medications (acyclovir) in rural facilities, inadequate awareness among healthcare providers regarding maternal varicella management, and limited diagnostic capacity for VZV PCR and serology [2]-[6].

Strengthening maternal infectious disease screening programs, introducing routine varicella vaccination into national immunization schedules, ensuring timely access to VZIG and antiviral therapy, and enhancing awareness among healthcare providers are essential strategies for preventing future cases and reducing associated morbidity and mortality [6].

5. Economic and Policy Implications

A 2022 global burden analysis estimated that varicella causes approximately 3.6 million cases and 6300 deaths annually worldwide, with the highest burden in low-income countries lacking vaccination programs [6]. Introduction of routine varicella vaccination in LMICs would require integration into existing childhood immunization programs (measles-rubella vaccination visits), surveillance systems to monitor vaccine impact and CVS incidence and health system strengthening for cold chain management and vaccine delivery [6].

The present case emphasizes the urgent need for policy intervention to prevent avoidable congenital infections in resource-constrained settings [6].

Diagnostic and Resource Limitations

This report has several limitations. Virologic confirmation of maternal or neonatal VZV infection was unavailable because molecular testing and specialized serologic investigations were not accessible at the time of presentation. Furthermore, advanced diagnostic investigations including neuroimaging, echocardiography, skeletal radiography and ophthalmologic imaging could not be completed because of the infant's rapid clinical deterioration and early death. Consequently, the diagnosis was based primarily on the characteristic clinical phenotype and documented maternal history.

6. Conclusion

Congenital varicella syndrome remains a rare but potentially fatal consequence of

maternal varicella infection during early pregnancy [1] [5]. This case demonstrates an unusually severe neonatal presentation characterized by extensive cutaneous, ophthalmologic, musculoskeletal, and genitourinary involvement with early neonatal death. In resource-constrained settings, diagnosis may depend largely on clinical recognition because access to virologic confirmation is often limited [3] [5]. Strengthening preconception screening, improving vaccine coverage, ensuring timely access to VZIG and antiviral therapy, and enhancing awareness among healthcare providers are essential strategies for preventing future cases and reducing associated morbidity and mortality [2] [6] [8].

Consent for Publication

Informed consent for publication of the clinical details, clinical images (limb abnormalities, cicatricial skin lesions, micropenis, rudimentary scrotum), and any accompanying figures was obtained from the infant's parents. No personal identifying information (name, initials, medical record number, dates of birth/admission) is included in this manuscript.

Availability of Data and Materials

The de-identified clinical data supporting the findings of this case report are available from the corresponding author upon reasonable request, subject to approval by the University of Port Harcourt Teaching Hospital Ethics Committee and in accordance with Nigerian data protection regulations.

Author Contributions

AAE conceptualized the report, managed the patient, reviewed the literature, and drafted the manuscript. CAN, NI, and OWU contributed to clinical management, data collection, manuscript revision, and critical intellectual review. All authors approved the final version of the manuscript and agree to be accountable for all aspects of the work.

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Conflicts of Interest

The authors declare no competing interests.

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