CASE REPORT

Unusual cause of brachial palsy with diaphragmatic palsy

Vishal Gupta,¹ Aakash Pandita,² Astha Panghal,² Neha Hassan³

¹Neonatology, Max Hospital, New Delhi, India

²Neonatology, Sanjay Gandhi Post Graduate Institute Of Medical Sciences, Lucknow, Uttar Pradesh, India

³Vidant Medical Center, East Carolina University, Greenville, North Carolina, USA

Correspondence to

Dr Aakash Pandita,
aakash.pandita@gmail.com

Accepted 2 April 2018

SUMMARY

We report a preterm neonate born with respiratory distress. The neonate was found to have diaphragmatic palsy and brachial palsy. The neonate was born by caesarean section and there was no history of birth trauma. On examination, there was bilateral congenital talipes equinovarus and a scar was present on the forearm. The mother had a history of chickenpox during the 16 weeks of pregnancy for which no treatment was sought. On investigation, PCR for varicella was found to be positive in the neonate.

BACKGROUND

Congenital varicella syndrome (CVS) is a rare disorder, which occurs in 1%–2% of fetuses of mothers suffering from primary varicella infection in the first two trimesters.^{1–3} The affected infants have distinctive abnormalities at birth including low birth weight, cutaneous lesions like cicatrix zig-zag scarring following dermatomal distribution (70%), papular lesions resembling connective tissue nevi, ocular abnormalities (66%) like chorioretinitis, cataracts, microphthalmia, Horner's syndrome, nystagmus, limb hypoplasia (50%), central nervous system abnormalities (46%) like seizures, mental retardation, hydrocephalus, cortical atrophy, cerebellar aplasia, encephalomyelitis and dorsal radiculitis and poor sphincter control due to improper development of sympathetic nervous system (32%).⁴ The diagnosis of varicella-zoster virus fetopathy is based primarily on the maternal history of chickenpox in early gestation combined with fetal stigmata.⁵ Antenatal sonograms assisted by fetal MR studies may pick the stigmata's in utero like hydrocephalus, clubfoot, a large bulbous skin lesion originally believed to be a meningocele, calcifications in the liver and other organs and a hypoplastic limb.^{6–8} Approximately 30% of affected babies, especially those having intracranial malformations die during the neonatal period.⁹ Although, many case reports have shown that CVS is associated with multiorgan involvement with permanent sequelae, but few authors propose that CVS may produce complete brachial plexus palsy also due to the involvement of neuroanatomical structures such as anterior horn motor neurons and the dorsal root ganglia neurons. Weakness in these patients may result from the intrinsic brachial plexus malformation or from a peripartum injury superimposed on a weakened brachial plexus.

CVS is a one of the unusual cause of diaphragmatic palsy, but none of the case reports mention diaphragmatic palsy in any of the cases. Here, we report a case of CVS with complete brachial plexus palsy and diaphragmatic palsy which has never been reported in past.^{10 11}

CASE PRESENTATION

A male baby weighing 880 g, product of a non-consanguineous marriage, was born to a 28-year-old primigravida mother who had been under regular follow-up. The anomaly scan done at 18–20 weeks was suggestive of right congenital talipes equinovarus (CTEV) but no other anomalies were evident. Mother suffered from chicken pox at 16 weeks of gestation and received no treatment. The baby was born by lower segment caesarean section at 32 weeks of gestation in view of intrauterine growth retardation and absent end-diastolic flow in umbilical artery. The baby cried at birth but developed secondary apnoea and received positive pressure ventilation for 30 s. The baby was admitted in neonatal intensive care unit (NICU) with respiratory distress syndrome. The baby received single dose of surfactant and required respiratory support in form of ventilation in view of poor respiratory efforts. On clinical examination, there was a bilateral CTEV and a scar on forearm (figure 1). There was hypoplasia of left upper limb with atrophy of left shoulder muscle and paucity of movements of left upper limb. Right foot CTEV was present with atrophy of calf muscle (figure 2). Eye examination showed scars on foveae region and pigmentary changes in fundus. No other major findings were present on general and systemic examination. The baby was gradually shifted to low flow oxygen devices and subsequently to room air. The baby was discharged after an NICU stay of 3 months.

INVESTIGATIONS

Chest X-ray showed features of hyaline membrane disease with elevated left dome of diaphragm (figure 3), which was due to diaphragmatic palsy confirmed by diminished left dome movement on ultrasonography. Echocardiography was normal. Investigation revealed haemoglobin of 19.8 g/dL, leucocyte count $15.5 \times 10^9/L$, platelet count $1.73 \times 10^9/L$. Cranial sonogram was normal. Varicella IgM was negative but IgG was positive. PCR for varicella-zoster virus was positive both in blood and cerebrospinal fluid.

To cite: Gupta V, Pandita A, Panghal A, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223624

Author response: Done as advised

Figure 1 Arrow showing scar on left forearm with brachial palsy of left side.

DIFFERENTIAL DIAGNOSIS

In addition to congenital varicella syndrome, other diagnoses which can be considered are:

- ▶ Shoulder dystocia.
- ▶ Neoplasm (neuromas, rhabdoid tumour).
- ▶ Intrauterine compression.
- ▶ Humeral osteomyelitis.
- ▶ Exocytosis of the first rib.

TREATMENT

The baby was given developmentally supportive care and rehabilitative measures for atrophied limbs. For CTEV, cast application by Ponsetti technique was applied. The baby was managed with antibiotics and injection acyclovir for 21 days.

OUTCOME AND FOLLOW-UP

The baby was discharged at 3 months of life and was thriving well on breast feeds. Ophthalmological follow-up was done and was reported to be normal.

Figure 2 CTEV right foot. CTEV, congenital talipes equinovarus .

Figure 3 Chest X-ray showing elevated left dome of diaphragm.

DISCUSSION

Fetal varicella syndrome is a rare complication of maternal varicella infection. The first case was reported by Laforet and Lynch in 1947.¹² Until now, more than 130 neonates born with signs of CVS have been described in the literature. A follow-up report in the literature demonstrated that in spite of initially poor prognosis, a good long-term outcome can occur in patients with CVS.¹³ Sauerbrei *et al*, reported a stillborn baby with CVS with limb hypoplasia, dermatomal skin lesions, microphthalmia, optic atrophy, chorioretinitis, coarctation of the aorta, Meckel's diverticulum, calcified necrosis in the lungs, liver and adrenal glands.¹⁴ Sharma *et al*, reported CVS in a newborn with maternal exposure at the 17th week of pregnancy. The baby had haemorrhagic bullous skin lesion with cicatricial limb scars.¹⁵ Chaudhary *et al*, reported scarring over right thigh and leg.¹⁶ Cicatricial lesions may not be present since birth in some cases, a case report mentioned a baby born with partially ruptured bulla over limb with haziness in the eye which gradually healed with cicatrization of limb and opacity in eye, respectively.¹⁷ The virus usually affects tissues in the rapid development phase, for example, limb buds, thus producing the characteristic manifestation 'the cicatrix'. Often, manifestations do not progress in postnatal phase and are supposed to be non-infective, hence, do not usually require antiviral or immunoglobulin treatment.⁵ But, once the stigmata of CVS occur they are usually permanent. So, during antenatal visits, mothers should be questioned regarding their immune status against varicella, and non-immune mothers should be advised to avoid contact with individuals with chickenpox. Universal immunisation may not be a cost-effective option for developing countries. Termination of pregnancy is not advisable as the risk of fetal damage is less, but attempts of early identification of stigmata of CVS should be made and mothers can be offered termination once CVS is suspected. Efforts should be made to prevent varicella-zoster virus infection during pregnancy by giving varicella-zoster immunoglobulin in non-immune expectant mother who has had a significant exposure to chickenpox at any gestation after informing her that it would not alter the risk of congenital varicella syndrome.

Acyclovir therapy of pregnant women also does not have an established role in preventing or modifying the severity of varicella-zoster fetopathy.¹⁸ For those babies born with the cicatricial lesion, plastic surgery may be considered. We report this case to increase awareness that CVS may present with complete brachial and diaphragmatic palsy apart from the well-known complication reported previously. A high index of suspicion and careful history is required for the diagnosis.

Learning points

- ▶ Antenatal history and high degree of suspicion are needed to diagnose congenital varicella syndrome (CVS).
- ▶ CVS is an unusual cause of complete brachial palsy.
- ▶ CVS may manifest as a diaphragmatic palsy.
- ▶ In the absence of effective treatment, prevention remains the best means to avoid CVS.
- ▶ IgM assays should not be used to confirm or rule out infection in a neonate.

Contributors VG, APandita and NH conceptualised and designed the manuscript, drafted the initial manuscript, reviewed and revised the manuscript. APanghal conceptualised, critically reviewed and revised the manuscript. All authors approved the final manuscript as submitted.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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