Rare disease

Volkmann ischemic contracture in a newborn

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Summary

Congenital Volkmann ischemic contracture is a very rare condition in which a neonate presents skin, muscular and nerve lesions due to increased intracompartment pressure and subsequent ischemia, probably due to extrinsic intrauterine compression. In this age group, there are only about 50 reported cases and a specific cause is unknown. The authors describe the case of a newborn who presented with bullous and ulcerated skin lesions and nerve palsy of his forearm at birth, evolving to subcutaneous and muscular necrosis and contracture. Two surgeries were performed and the baby began a daily physiotherapy program that resulted in aesthetical improvement and recovery of his hand and forearm mobility. Early recognition of this rare entity and subsequent emergency fasciotomy are the best ways to improve prognosis.

BACKGROUND

Volkmann ischemic contracture syndrome consists of ischemic neuromuscular and skin lesions due to increased intracompartment pressure. It is a very rare condition in the newborn and a specific cause in this age is unknown. The lesions are present at birth and characterised as bullae that quickly burst into deep ulcers evolving to necrotic areas. This diagnosis is rarely taken in consideration immediately leading to evolution with sequelae. Early recognition of this entity and subsequent emergency fasciotomy are the best ways to improve prognosis. The authors describe a case of a congenital Volkmann ischemic contracture to alert for the possibility of this diagnosis in a newborn presenting open wounds skin injuries at birth.

CASE PRESENTATION

The patient was a newborn male with extensive cutaneous lesions on the left forearm present since birth. He was the first offspring of young healthy parents with no history of consanguinity. Antenatal care was adequate and maternal routine serologic testing and viral markers were negative. Echography parameters were normal until the delivery date, when oligohydramnios was detected. Cephalic position and adequate foetal movement perception were constant throughout pregnancy. Hydroxyzine and oseltamivir were administered during the third trimester due to a flu syndrome. The mother gained 23 kg (51 lb) during gestation (prepregnancy overweight–68 kg). Delivery was induced at 38 weeks and 5 days due to oligohydramnios.

Figure 1 (A, B) Affected limb in the delivery room: disrupted bullous and ulcerated skin throughout the left forearm.

Figure 2 Forearm on second day: skin and muscular necrosis.

It was extremely difficult and vacuum extraction was necessary. Apgar scores were 7 and 8 at first and fifth min, respectively and birth weight was 3470 g. Physical examination revealed signs of cyanosis, hypotonia and slow reflex responses but the baby recovered without the need for resuscitation procedures. Upper left limb was prone, without spontaneous motion or palm prehension. The forearm was flattened, showing bullous and ulcerated skin throughout and the hand was cyanotic but not cold (figure 1A,B). Mild reduction of the lower limbs extension movements, small denuded skin areas on the inguinal pleats and antecubital zones as well as two small bullae on the right hand and foot were also observed. Rapid evolution to skin and muscular necrosis on the day after the birth was observed (figure 2).

INVESTIGATIONS

Laboratory tests requested, namely a complete blood count, C-reactive protein, liver enzymes, serum electrolytes, blood urea nitrogen, creatinine, routine blood coagulation tests, urinalysis, blood and urine cultures, were all normal except for a slight increase in creatine kinase and lactate dehydrogenase values. The mother's serologies for varicella zoster and herpes simplex were negative. There are no signs of fracture on x-ray. Cerebral ultrasound revealed a hyperechogenic focus, corresponding to a sub-cortical haemorrhage detected on brain MRI. Macroscopic and histological examination of the placenta was normal. Skin and muscular biopsy made on second day confirmed tissular necrosis.

DIFFERENTIAL DIAGNOSIS

The scenario of multiple skin lesions, mostly on the upper left forearm, associated with palsy of the limb, lead to the following differential diagnosis consideration: bullous epidermolysis, amniotic band syndrome, congenital aplasia cutis, thrombosis, bacterial or viral infection. The skin lesions at birth coupled with their evolution to necrosis formed the basis for a 4th day diagnosis of Volkmann ischemic contracture.

TREATMENT

The child began topical treatment with silver sulfadiazine on the 2nd day and was submitted to surgical debridement on the 13th day with posterior skin graft on the 24th day. A daily physiotherapy programme was started after the first week and is still maintained.

OUTCOME AND FOLLOW-UP

Evolution to retraction of fingers and hand, when flexed, and limited wrist extension were initially observed, with subsequent gradual mobility recovery. One year after diagnosis, some scarring and forearm atrophy were still present (figure 3A,B).

DISCUSSION

Volkmann ischemic contracture syndrome was first described by Richard von Volkmann in 1869¹ and consists of the presence of neuromuscular and skin lesions originating in a rapid increase of interstitial pressure in a closed anatomic space.¹⁻⁸ It is most commonly found in children or adults who have suffered intense trauma, prolonged external compression, excessive exercise, burns and animal bites.¹⁻² Congenital Volkmann ischemic contracture, also known as neonatal compartment syndrome, is a very rare condition. In this age group there are only about 50 documented cases, all affecting an upper limb, even though it may occur in the lower limbs.¹⁻¹⁶

The lesions are present at birth and characterised as bullae that quickly burst into deep ulcers evolving to necrotic areas.^{1-4 8-10} Later, muscular atrophy becomes evident with contraction and flexing of the fingers and claw-like hand position.^{2 4 9} This contraction is due to fibrosis and contracture of the flexing muscle, presenting at birth or developing during the first weeks.² The histological characteristics are unspecified and the necrosis severity is related to ischemia duration.⁴ In the cases previously described, there is no evidence of compression in other anatomical regions.¹⁻¹⁶ However, in the present case a few discreet skin lesions were detected in other regions that may be related to slight compression from conflict

Figure 3 (A, B) Affected limb 1 year after diagnosis: some scarring and forearm atrophy, recovery of forearm and hand mobility.

of intrauterine compartment. The haemorrhagic cerebral lesion observed in the magnetic resonance was probably due to traumatic delivery. There is only one reported case of a preterm neonate with concomitant cerebral lesions due to vascular obliteration of the sylvian artery, probably due to vascular emboli consecutive to the death of a co-twin during pregnancy.⁵

The specific cause for this syndrome is still unknown, but is consensual that the ischemic lesion is caused by an increase in intracompartmental pressure.^{2–8} The average diastolic pressure of the newborn is approximately 40 mm Hg or less.⁹ Therefore, even small increases in intracompartmental pressure may rapidly compromise muscular perfusion.^{3 9} At this age group, it seems to be the result of external intrauterine compression, for which several situations may be responsible such as instrumental deliveries, difficult extractions, foetopelvic incompatibility, inadequate limb positioning, oligohydramnios, amniotic band or umbilical cord constriction and twin pregnancy.^{2–9} Other factors that may lead to difficult extractions and initial intrauterine injury are mother's diabetes, prolonged membrane rupture, pre-eclampsia, premature birth, uterine anomaly and excessive weight gain on the mother.^{2–9} The administration of psychotropic medication during pregnancy, such as narcoleptics and benzodiazepines, has been considered as a risk towards foetal hypomotility and abnormal intrauterine positioning.³ Intrinsic factors to the

newborn include hypercoagulability states.⁹ However, some studies show that there is a predominant involvement of the extensor muscles, a superficial muscular group, which is suggestive that the initial injury is due to external direct compression instead of vascular alterations.^{2 10} Even though there may be some associated factors, there are reported cases in which both pregnancy and delivery presented with none of the above related factors.^{2 9 10} In the present case, both prenatal and perinatal factors were identified such as the mother's excessive weight gain, hydroxizine administration that may cause foetal hypomotility, oligohydramnios and a difficult instrumental delivery. Side effects of oseltamivir use during pregnancy are poorly documented, rendering it hard to evaluate its implication, but it seems not to be involved.^{17–21}

When the lesions are recent, ideally during the first 24 h, fasciotomy is the choice treatment as it allows a reduction in intracompartmental pressure and arterial reperfusion.^{2 6 9 11} However, this diagnosis is rarely taken in consideration immediately.^{2 6 10} In a study of 24 cases, fasciotomy during the first 24 h was limited to a single occurrence, the only one to evolve with no sequelae.⁹ At a later stage, other surgical treatments are needed including devitalised tissue debridement and skin grafting, nerve decompression, scar revision, contractures release and angular deformity correction.^{4 6 9} The timing of these procedures depends on age, limb size and surgical goals.⁹

In this case, as with most documented cases,^{1–16} sequelae are atrophy, scarring, mobility limitation and probable limitation of bone growth. The study by Ragland *et al* showed skeletal involvement in 18 cases and nerve lesions in 16 out of 24, with neurological recovery in four.⁹

Learning points

- ▶ Skin injuries with open wounds are rarely found in newborns. When present, especially on the forearm, should alert the physician to the possibility of a neonatal compartmental syndrome.
- ▶ Prompt suspicion of this rare occurrence should lead to an urgent referral to a surgeon who can perform decompressive fasciotomy, the best way to decrease the extent of tissue loss and degree of functional impairment.
- ▶ At a later stage, other surgical treatments and intensive programme of physiotherapy allow some functional and aesthetical improvement of the involved limb.

Competing interests None.

Patient consent Obtained.

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