

Neonatal Volkmann syndrome: About a case

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Abstract

Neonatal Volkmann syndrome is rare. It must be evoked when skin lesions associated with wrist and finger paralysis are present. We present, with a long delay, a patient who presented a Volkmann syndrome. No etiology was found, as well as no associated brain lesion. Several surgical interventions were necessary to improve the functional prognosis. Volkmann's syndrome must be recognized in order to search for associated lesions and to propose an emergency aponeurotomy. Regular follow-up should be performed to avoid bone deformities and retractions.

Keywords: Volkmann; Necrosis; In Newborn; Ischemic; Aponeurotomy

1. Introduction

Neonatal Volkmann syndrome is rare. It is a neuromuscular ischemia secondary to an increase in interstitial pressure in a closed anatomical space (osteoaponeurotic compartment). Its occurrence in the neonatal period is exceptional and its etiology is not well defined. However, it must be diagnosed early in order to allow optimal treatment. The exact causes of neonatal Volkmann syndrome are not known. Mechanical compression factors are mainly considered: constrictive amniotic band, compression by a deceased twin, malposition of the arm, coiled umbilical cord, local or general factors that contribute to a difficult extraction [1-2]. Only about ten cases have been described in the literature. We present one patient with neonatal Volkmann's syndrome with a long delay.

2. Observation

Male newborn from a monitored pregnancy conducted at 37 SA; delivery by high route for suspected acute fetal distress on RCF with oligohydramnios; Apgar was 10/10; birth weight 2550 g; 34-year-old mother G5P3 with notion of 1 early abortion and 1 death per partum in the context of perinatal asphyxia; having as antecedent a gestational diabetes under insulin and dysthyroidism discovered at delivery. The infectious anamnesis was negative with no medication or toxicity taken during the pregnancy and no similar case in the family. The newborn was admitted at 1 day of age for skin necrosis of the right forearm with respiratory distress rated at 1/10 according to the Silverman score.

On clinical examination, the newborn presented phlyctenes and edema on the anterior and lateral aspect of the forearm and right hand associated with cyanosis.

The thromboembolic workup, the echodoppler of the right upper limb and the echocardiogram were normal.

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Figure 1 Phlyctenes on the anterolateral aspect of the forearm at birth suggesting neonatal Volkmann syndrome



Figure 2 Necrosis with fibrinous ulceration

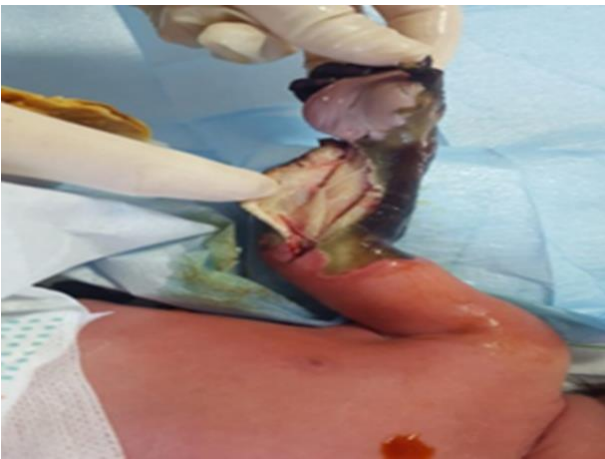


Figure 3 Aponeurotomy



Figure 4 Necrosis despite aponeurotomy

A bullous detachment with necrosis and deep fibrinous ulceration of the medial and anterior high forearm and elbow crease, moderate digital flexion (Fig. 2). Distal pulses were difficult to perceive because of the edema. The evolution was marked by the abolition of the radial and ulnar pulses; there was no biological inflammatory syndrome, no hyperleukocytosis, no sign of disseminated intravascular coagulation (DIC), and the bacteriological skin samples were negative. No skin biopsy was performed. A probabilistic antibiotic therapy was started. A transfontanellar ultrasound was normal.

An aponeurotomy of the anterior and posterior lodge of the right forearm was proposed with passive mobilization started in parallel.

The evolution was marked by extensive necrosis of the hand and forearm with cardboard skin leading to a trans-radio-ulnar amputation with radial and ulnar osteotomy at the level of the proximal middle 1/2 of the forearm.

Subsequently, necrosis spread to the arm as well, resulting in a second complete amputation of the entire limb.



Figure 5 Extensive arm necrosis



Figure 6 Amputation of the necrotic limb

3. Discussion

Neonatal Volkmann's syndrome is rare and associates skin lesions such as phlyctenes or erosions, a compartment syndrome and paralysis of the forearm muscles. It should not be confused with aplasia cutis congenital (extensive skin lesions, especially on the skull) or gangrene of the extremities (complete necrosis of the extremities) [1-2].

Currently, the pathophysiology of Volkmann syndrome remains unknown. However, some authors have proposed several theories, sometimes associated, including: traumatic (compression against the sacrum in the context of oligohydramnios; stricture of the upper limb by the umbilical cord; occipito-transverse presentation; iatrogenic by arterial catheterizations), thrombotic or septic [1-4]. Neurological manifestations are variable: hypo-mobility, flaccid paralysis, contractures.

The diagnosis is probably underestimated in the initial period, as evidenced by the retrospective series of Ragland et al [9] of 24 cases over a period of 20 years, despite the presence in all cases of cutaneous signs: only one case with surgical consultation and intervention within the first 24 hours, five cases seen within the first month and five cases within the first three months. The other cases were mostly seen after one year of age (mean age 6.5 years). Only one patient in this series, operated early, had a favorable evolution. All the other patients had severe complications: tissue loss with amputations of the fingers (three cases) or of the hand (one case), persistent compressive neuropathy (11 cases), muscle loss (ten cases), extensive bone damage (18 cases) with possible impact on distal growth (ten cases) [9]

A general clinical and biological examination should be performed to confirm the diagnosis and to look for predisposing factors such as dehydration, infection, maternal diabetes and amniotic bridges syndrome.

Armstrong and Page reported six cases of neonatal forearm necrosis with clinical and evolutionary characteristics of neonatal Volkmann syndrome [10]. They proposed for these observations the denomination of "intrauterine vascular insufficiency of the upper limb", in order not to ignore a possible embolic or thrombotic etiology

Moreover, for all the authors, an MRI is currently indispensable in order to search for associated cerebral vascular lesions [1-2].

A most of the time, the clinical examination is sufficient to determine the indication for an emergency surgical aponeurotomy. Indeed, even if it has been clinically proven that emergency decompression of the forearm improves muscle and nerve recovery [1-5], a secondary excision surgery is usually requested when the necrotic areas are delimited. In all cases, it must be associated with intensive rehabilitation using resting or dynamic splints in order to reduce the risk of sequelae [6]. Long follow-up is rarely reported in the literature. However, all the articles show that in the long term the functional results are not excellent [3-7-8].

Muscular lesions lead to a decrease in grip strength and mobility of the fingers. Ischemic lesions can cause retraction of the forearm muscles but also of the first commissure, placing the thumb in the palm. In addition, there are radiological lesions with shortening and widening of the epiphysis of the radius and ulna [2]. These bone lesions associated with muscle lesions lead to significant deformities resulting in a functionally poor hand or at a amputation. Therefore, the

follow-up of children with Volkmann syndrome must be prolonged in order to prevent complications by proposing surgery adapted to the deformities (first row resection, temporary fixation of the wrist, opening of the first commissure, digital arthrolysis...). However, the results are modest and the best treatment is preventive by informing the maternity hospitals so that an emergency aponeurotomy can be proposed to avoid amputation.

4. Conclusion

Congenital limb ischemia is favored by malformative, infectious, obstetric and metabolic factors. Maternal diabetes is most often incriminated (probably in our case). The diagnosis is clinical and can be confirmed by echodoppler and arteriography. Treatment is as conservative as possible based on heparin therapy and anti-thrombotic drugs (early stages) and cannot be useful at the gangrene stage. Amputation is reserved for outdated forms. Hence the interest of an early and multidisciplinary management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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