Management Of Congenital Volkmann Syndrome: A Case Report

A Kane, C Diouf, Y Sghair, N Mohamed, E Adda, H Tandia

Citation

A Kane, C Diouf, Y Sghair, N Mohamed, E Adda, H Tandia. *Management Of Congenital Volkmann Syndrome: A Case Report*. The Internet Journal of Pediatrics and Neonatology. 2019 Volume 21 Number 1.

DOI: 10.5580/IJPN.54406

Abstract

Congenital Volkmann syndrome is an exceptional condition with an etiology and a management that are controversial. We are reporting the case of a newborn girl from a twin pregnancy that was seen for edema of the right hand and forearm with cyanosis and ulcerations in patches and paralysis of the wrist and bent fingers. The other twin was a macerated stillborn. The limb echodoppler, x-ray, and blood tests were normal. Fasciotomy was performed on the second day of life with immediate favorable result. At 18 months, the child had delayed psychomotor development, slight dorsiflexion of the hand and thumb adduction while the other fingers are normal.

The diagnosis and the faciotomy must be done the earliest for a convenient evolution of the congenital Volkmann syndrome.

INTRODUCTION

Congenital Volkmann syndrome is an exceptional condition characterized by the observation at birth of cutaneous lesions of the upper limb associated with a paralyzed wrist and fingers. Its etiologies and its management are controversial [1]. We had reported a case supported in our service and done a review of the literature.

CASE REPORT

We report about a newborn female who was sent to us twelve hours after her birth for cyanosis of the right hand. She was born from a twin pregnancy with the other twin a macerated stillborn. Caesarean delivery was at 37 weeks due to metrorrhagia caused by the fall of the mother.

Physical examination, showed a temperature at 37 ° C and a weight of 2900g. There was edema of the right upper extremity taking the hand and the forearm with cyanosis and ulcerations in plates as well as paralysis of the wrist and fingers flexing (Fig 1). The rest of the exam was normal. Echodoppler of the right upper limb showed a permeability of the vascular axes. Biological assessment and X-ray of the limb were normal. After several opinions, the diagnosis of congenital Volkmann syndrome was retained and aponevrotomy performed on the second day of life. Immediate operative follow-up was favorable with hand and finger resurfacing and scarring of the wound after 3 weeks (Fig 2). 18 months after surgery, the child had a slight dorsiflexion of the hand and adduction of the thumb while the other fingers had a normal mobility (Fig 3). There was also a delay in standing and walking.

Figure 1

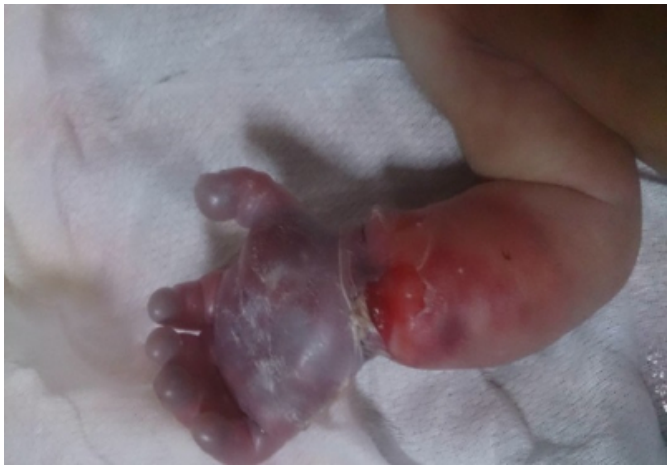


Figure 2

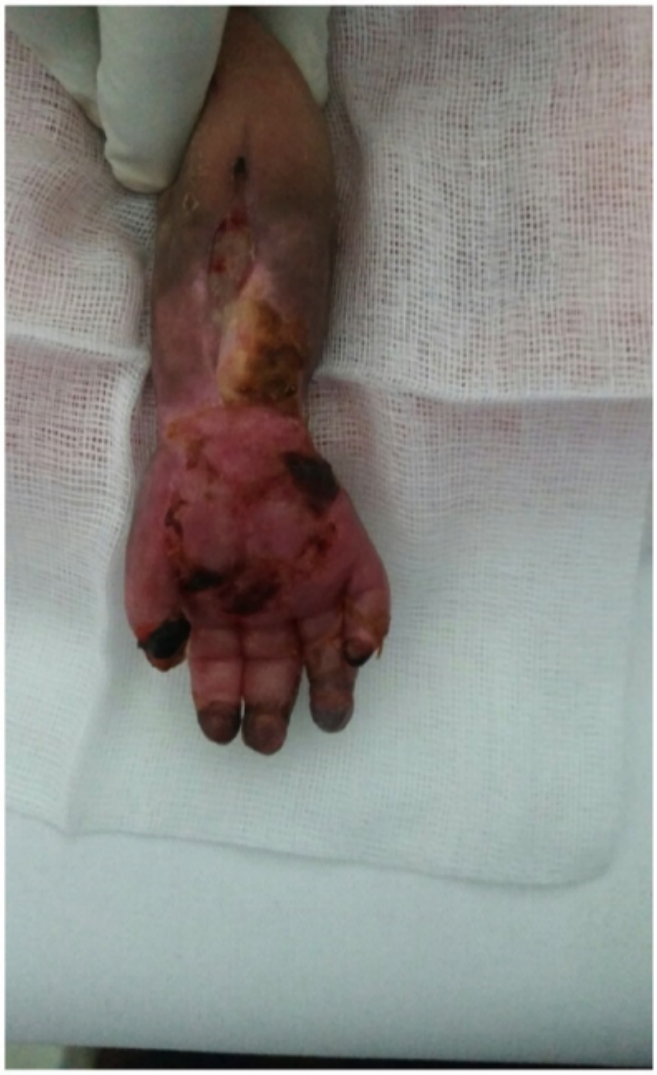
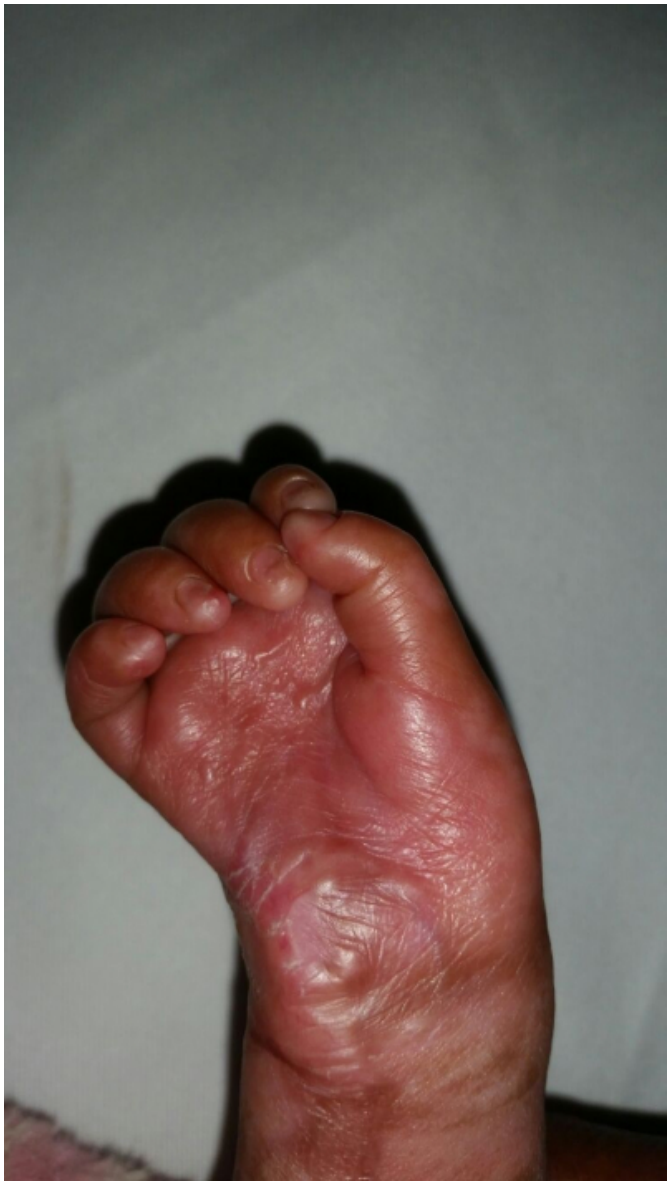


Figure 3



DISCUSSION

Volkmann's syndrome, in its acquired form, is a well-described pathology, unlike the congenital form, which is exceptional with an average case reported each year [2]. The etiology is still poorly known but mechanical factors would be incriminated including the compression of the limb by a deceased twin (case of our patient?), amniotic band, circular cord, and perinatal ischemia would be a factor favoring [3]. The diagnosis is clinical and is based on the observation at birth of an edema of the forearm and hand associated with cutaneous ulcers and motor deficit of the wrist and fingers [4]. However, certain dermatological conditions, including congenital cutaneous aplasia, congenital chickenpox, necrotizing fasciitis, epidermolysis bullosa... can look alike pathology [5]. In addition, a careful neurological

examination should be performed to detect signs of perinatal ischemia [6].

Congenital Volkmann syndrome is a surgical emergency consisting of a wide fasciotomy of the muscular compartments of the forearm [7]. In cases where the fasciotomy has not been performed, some authors recommend local care to heal the lesions followed by free transfer of a musculocutaneous flap from a re-innervated latissimus dorsi to one of the motor branches. median nerve around the age of 7 months [8] or correction osteotomies from the age of 18 months [9]. The evolution of this pathology is considered disappointing (member with low functionality) by almost all the authors even if the treatment was early [10].

CONCLUSION

Although rare, the Congenital Volkmann syndrome is to be considered in front of any newborn with cutaneous lesions of the upper limb associated with an impairment of the wrist and fingers. Discharge aponevrotomies must be performed urgently to hope for a satisfactory function of the limb.

References

1- Martin B, Treharne L. Neonatal compartment syndrome. Ann R Coll Surg Engl. 2016 ;98(7) : 111-3
2- Ragland R, Moukoko D, Ezaki M, Carter PR, Mills J. Forearm compartment syndrome in the newborn: report of 24 cases. J Hand Surg Am. 2005;30(5):997-1003
3- Dandurand M, Michel B, Fabre C, Stoebner P, Meunier L. Neonatal Volkmann's syndrome. Ann Dermatol Venerol. 2009;136(11):785-9
4- Raimer L, McCarthy RA, Raimer D, Colome-Grimmer M. Congenital Volkmann ischemic contracture: a case report. Pediatr Dermatol. 2008;25(3):352-4
5- Cham PM, Drolet BA, Segura AD, Esterly NB. Congenital Volkmann ischaemic contracture: a case report and review. Br J Dermatol. 2004;150(2):357-63
6- Pavlidis E, Spagnoli C, Duca M, Pisani F. Association between neonatal Volkmann's syndrome and perinatal ischemic stroke: review of the literature. Acta Biomed 2015;86(3):213-9
7- Plancq MC, Buisson P, Deroussen F, Krim G, Collet LM, Gouron R. Successful Early Surgical Treatment in Neonatal Compartment Syndrome: Case Report. J. Hand Surg. 2013;38(6): 1185-8
8- Trimaille A, Kerfant N, Le Rouzic-Dartoy C, Henry A, Hu W. Free re-innervated Latissimus Dorsi musculocutaneous flap for treat congenital Volkmann ischemic Contracture: A case report. Ann Chir Plast Esthet. 2014;59(3):200-3
9- Goubier JN, Romaña C, Molina V. Neonatal Volkmann’s compartment syndrome. A report of two cases. Chir Main. 2005;24(1):45-7
10- Tetreault AK, Axibal DP, Scott FA.]. Neonatal Compartment Syndrome Treated Within the First 24 Hours of Life. Orthopedics. 2018;41(5):731-3

Author Information

Ahmed Kane
Centre Hospitalier National de Nouakchott

Cheikh Diouf
Centre Hospitalier de Ziguinchor

Yacoub Mohamed Sghair
Centre Hospitalier Mère Enfant de Nouakchott

Nagi Sidi Mohamed
Centre Hospitalier National de Nouakchott

Elhadj Adda
Centre Hospitalier National de Nouakchott

Hadya Tandia
Centre Hospitalier Mère Enfant de Nouakchott