CASE REPORT

Neonatal Acute Compartment Syndrome as First Manifestation of Hematologic Disease: Case Report

Maria Luís Sacras[1], Catarina Ladeira[1], Sara Carmo[1], Paula Kjöllerström[2]

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Abstract

Hemophilia A, characterized by deficiency of functional plasma clotting factor VIII, is an X-linked disorder. Signs and symptoms depend on factor VIII levels. Severe hemophilia A (factor levels less than 1%) is usually diagnosed in the first two years of life. Pediatric compartment syndrome is most associated with trauma, but infectious and vascular causes must also be considered. We report a case of a newborn who presented with a severe hand hematoma after venous puncture, complicated by compartment syndrome. An isolated prolonged partial thromboplastin time was found, and hemophilia was suspected. Factor VIII levels less than 1% were consistent with severe hemophilia A. The patient received recombinant factor VIII peri-operatively. Fasciotomy was performed and the patient was kept on antibiotics until closure.

Keywords: Acute Disease; Compartment Syndromes/diagnosis; Compartment Syndromes/etiology; Compartment Syndromes/therapy; Fasciotomy; Hemophilia A/complications; Hemophilia A/diagnosis; Infant, Newborn

Introduction

Hemophilia A is an X-linked inherited hematological disease caused by factor VIII deficiency. Severe hemophilia can present early in life with easy bruising, bleeding and hemarthrosis. Intracranial hemorrhage is the most life-threatening complication in the neonate.[1] Acute compartment syndrome is an uncommon but serious complication in hemophilic patients. Uncontrolled bleeding, leading to an increase in interstitial pressure in a closed compartment, results in a decreased perfusion gradient which may lead to ischemia of the affected tissue, most commonly a limb.[2] A multidisciplinary approach facilitates an early diagnosis. Factor replacement and timely fasciotomy are crucial.

Case Report

A 1-day old male neonate born to healthy Nepalese non consanguineous parents was transferred to our center due to a right-hand hematoma after a venous puncture for blood sampling due to septic risk (mother with fever during labor).

He was delivered by C-section at 40 weeks of gestation, with an Apgar score of 9 and 10 at 1 and 5 minutes, respectively. The birth weight was 3154 g.

On admission he presented a large hematoma on the dorsal aspect of the right hand, extending from the proximal interphalangeal joints of the five fingers to the wrist proximally, with tension on palpation and increased capillary refill time (Fig. 1). Laboratory values showed a hemoglobin value of 13.3 mg/dL, a platelet count of 216 000 cells/L, elevated activated partial thromboplastin time of 1128.5 seconds, factor VIII was unmeasurable (factor IX and von Willebrand factor were within normal ranges).

A diagnosis of hemophilia A was assumed, and after intravenous administration of recombinant factor VIII (rFVIII) a fasciotomy was performed with immediate tension relieve and improved distal perfusion (Fig. 2).

On the post-operative period the right hand was kept elevated, with continuous monitoring of oximetry. Intravenous rFVIII therapy (five exposure days) and antibiotics were maintained during admission. Fasciotomy was stage-closed on days five and eight post-operative (Fig. 3).

The patient fully recovered and was discharged on the ninth post-operative day. He was revaluated two weeks after in the pediatric plastic surgery clinic, showing a favorable evolution. On follow-up hematology appointment, the diagnosis of hemophilia A was confirmed by molecular

1. Pediatric Surgery Department, Hospital de Dona Estefânia, Centro Hospitalar Lisboa Central, Lisboa, Portugal
2. Hematology Unit, Hospital de Dona Estefânia, Centro Hospitalar Lisboa Central, Lisboa, Portugal
Corresponding Author
Maria Luís Sacras
https://orcid.org/0000-0002-9935-0283
marialuissacras@gmail.com
Serviço de Cirurgia Pediátrica, Hospital de Dona Estefânia, Rua Jacinta Marto, 1169-045 Lisboa, Portugal
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studies. On follow-up consultations, the mother was confirmed to be a carrier with normal factor VIII levels, and after questioning, recalled her only brother having a bleeding disorder since infancy, which had never been investigated or treated. We assume it is highly likely severe hemophilia A.

The infant was kept on on-demand treatment during the first year of live and started prophylaxis at 13 months. So far, at 2 years old, evolution has been uneventful, with negative inhibitors.

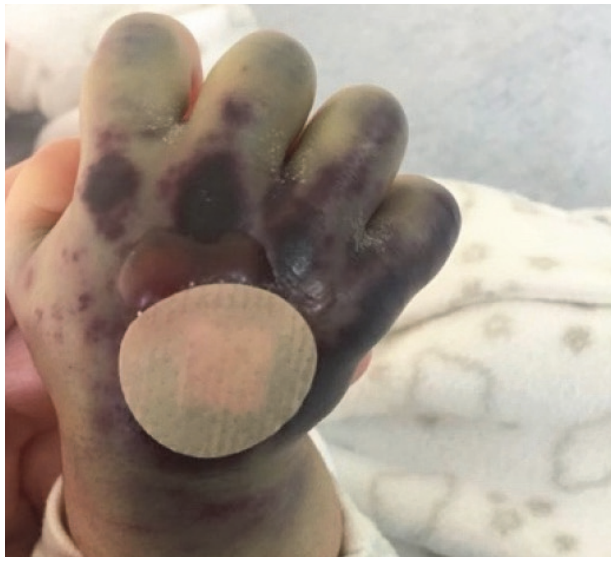

Figure 1. Hand hematoma complicated with compartment syndrome after venous puncture.

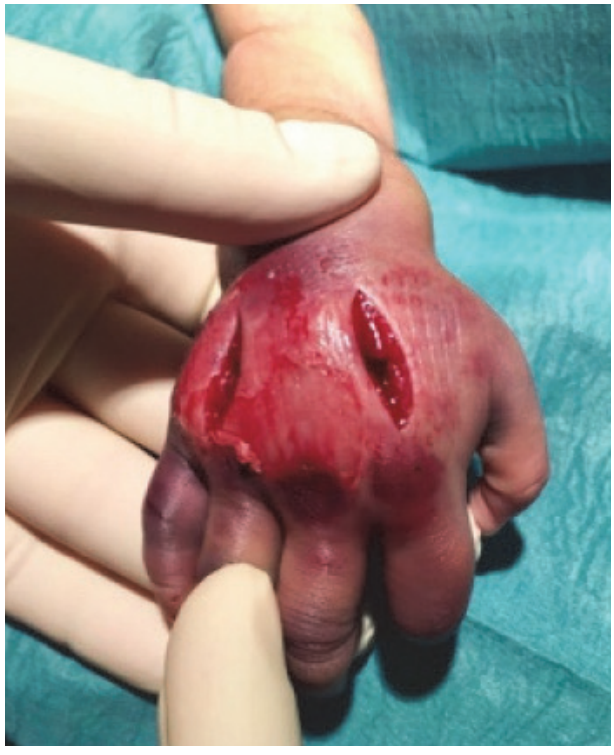

Figure 2. Hand hematoma immediately after fasciotomy.

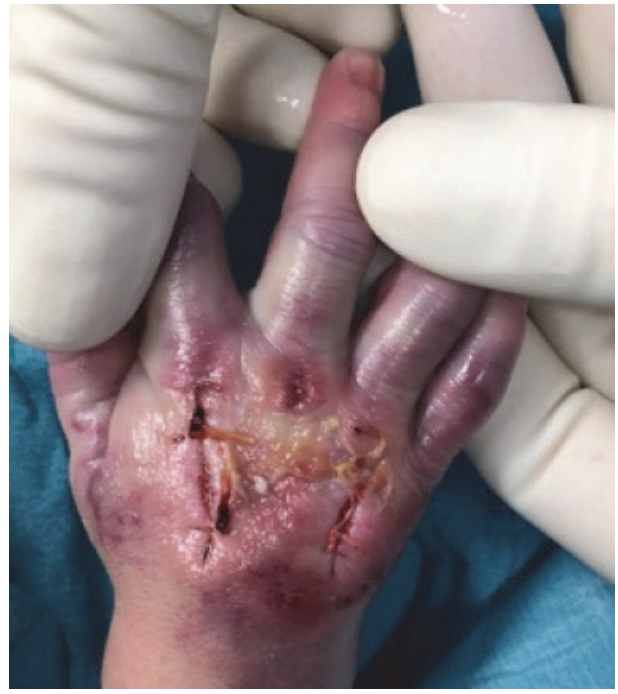

Figure 3. The affected hand at sixth post-operative day.

Discussion

Hemophilia A is an X-linked inherited disease which can have different manifestations depending on factor VIII levels. Severe deficiency (factor level below 1%) can present as spontaneous bleeding. While in older children muscle and joint bleeds predominate, in neonates the diagnosis is usually suspected after excessive bleeding or hematoma formation following venipuncture or intramuscular vitamin K administration.[3]

Compartment syndrome is a well-recognized, albeit rare manifestation of hemophilia A. In older children it is usually related to trauma, but in infants vascular and infectious etiologies are more common.[4] Hepatic and renal failure, leukemia, hemophilia, and snake bite must also be considered. In athletes the condition might be exercise related.[4,5] Neonatal compartment syndrome is a rare entity described on the upper extremities in relation to birth trauma.[4]

The clinical picture, classically described as the five P – pallor, pain, pulse weak / absent, paralysis and paresthesia – is difficult to recognize in children, making the diagnosis challenging. Instead, a three A system has been proposed: analgesic requirement increasing, anxiety and agitation.[2] Measurement of compartment pressure can be necessary to establish the diagnosis.

Once the diagnosis has been established, prompt treatment is required to prevent prolonged ischemia, which can compromise limb function. In children presenting with prolonged bleeding, a complete

blood count, prothrombin time and activated partial thromboplastin time evaluation are mandatory.[1] In the neonate, testing for factor levels should be considered even in the absence of a positive family history, since as much as 30% are *de novo* mutations.[3] Recombinant factor VIII is the mainstay of treatment and must be administered pre-operatively. In emergent cases fresh frozen plasma is also an option.[3] Removal of external sources of compression, limb elevation and emergent fasciotomy should be promptly performed.

Fasciotomy in children usually has good outcomes, with a recent systematic review reporting full recovery in 85% of the patients.[6] Although some reports suggest increased potential for full recovery in children,[4] a shorter time interval between injury and surgery avoids the risk of potential catastrophic consequences, such as amputation and loss of function.[7] Fasciotomy closure in pediatric patients requires an average of three surgeries.[8] Skin grafting and vacuum-assisted closure might be necessary.[2]

In this clinical report, compartment syndrome was the first manifestation of a congenital coagulopathy after a simple procedure (venous puncture). Prompt recognition and referral as well as a multidisciplinary approach were key for the effective treatment and favorable clinical evolution.

WHAT THIS CASE REPORT ADDS

- Compartment syndrome is rare in pediatric patients.
- In the absence of fracture, hematologic disorders should be sought, even without family history.
- The diagnosis can be challenging but an increasing need for analgesics is suspicious.
- Fasciotomy is the mainstay of surgical treatment and when timely performed leads to favorable outcomes.
- A multidisciplinary approach is needed to establish a timely diagnosis and provide the best possible treatment.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Consent for publication was obtained.

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Síndrome Compartimental Aguda Neonatal como Primeira Manifestação de Doença Hematológica: Caso Clínico

Resumo

A hemofilia A, caracterizada por deficiência do fator VIII de coagulação plasmática funcional, é uma doença ligada ao cromossoma X. Os sinais e sintomas dependem dos níveis de fator VIII. A hemofilia A grave (níveis do fator inferiores a 1%) geralmente é diagnosticada nos primeiros dois anos de vida. Em pediatria, a síndrome compartimental está mais associada a trauma, mas as causas infeciosas e vasculares também devem ser consideradas. Relatamos o caso de um recém-nascido que apresentou um hematoma grave nas mãos após punção venosa, complicado por síndrome compartimental. Foi determinado um tempo de tromboplastina parcial prolongado isolado e levantada a suspeita de hemofilia. Os níveis de fator VIII inferiores a 1% eram consistentes com hemofilia A grave. O doente recebeu fator VIII recombinante no perioperatório. Foi realizada uma fasciotomia e instituído tratamento antibiótico até o encerramento estar completo.

Palavras-Chave: Doença Aguda; Fasciotomia; Hemofilia A/complicações; Hemofilia A/diagnóstico; Recém-Nascido; Síndromes Compartimentais/diagnóstico; Síndromes Compartimentais/etiologia; Síndromes Compartimentais/tratamento