Reporte de Caso

Red blood cell exchange with peripheral venous access in a patient with sickle cell anemia and stroke

Recambio hemático con acceso venoso periférico en un paciente con drepanocitosis y accidente cerebrovascular isquémico

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Resumen

La drepanocitosis es la enfermedad huérfana más frecuente en el Valle del Cauca, se estima que es la principal causa de muerte por enfermedades huérfanas en Colombia en población que se autodenomina como afrocolombiana y mulata. El accidente cerebro vascular isquémico es una de las complicaciones más devastadoras de la enfermedad, su tratamiento requiere el empleo de recambio hemático automatizado el cual no se ha reportado hasta el momento en Colombia. El acceso venoso en los procedimientos de aféresis puede constituir una barrera de acceso y de oportunidad a la terapia. Se describe el caso de un paciente de 15 años con diagnóstico de drepanocitosis (HbSS) quien consultó a urgencias por presentar hemiparesia izquierda (fuerza 3/5), crisis dolorosa (escala visual análoga 10/10), con una tomografía axial computarizada que evidenciaba un accidente cerebrovascular isquémico. La última electroforesis de hemoglobina mostró un porcentaje de hemoglobina S del 95% y una hemoglobina de 10,5 g/dl. El servicio de hematología pediátrica consideró este escenario una indicación de recambio hemático, el cual se realizó con acceso venoso periférico requiriendo 12 unidades de glóbulos rojos leucorreducidos, fenotipados para el Rh y kell y compatibles y se utilizó la máquina Optia Spectra®. A las 48 horas el paciente presenta mejoría del dolor (escala visual análoga 0/10) y mejoría de la hemiparesia (fuerza 4/5), la electroforesis de hemoglobina S mostró un porcentaje del 8%. En Colombia es el primer caso descrito en la literatura de recambio hemático en un paciente con drepanocitosis con acceso venoso periférico y resultados exitosos.

Palabras clave: Drepanocitosis, Transfusión, Enfermedades cerebrovasculares, Hemoglobinopatía.

Abstract

Sickle cell disease is the most frequent orphan disease in Valle del Cauca, it is estimated that it is the main cause of death from orphan diseases in Colombia in a population that calls itself Afro-Colombian and mulatto. Ischemic stroke is one of the most devastating complications of the disease, its treatment requires the use of Red blood cell exchange (RBCX) which has not been reported in Colombia. Venous access in apheresis procedures can constitute a barrier to access and opportunity to this therapy. We describe the case of a 15-year-old patient with diagnosis of sickle cell

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disease (HbSS) who consulted the emergency room for presenting left hemiparesis (strength 3/5), painful crisis (visual analogue scale 10/10), with a computerized axial tomography that showed an ischemic stroke. The last hemoglobin electrophoresis showed a hemoglobin S percentage of 95% and a hemoglobin of 10.5 g / dl. The pediatric hematology service considered this scenario an indication for Red blood cell exchange (RBCX), which was performed with peripheral venous access requiring 12 units of leukoreduced packed red blood cells, phenotyped for Rh and kell and cross-matched, and the Optia Spectra® machine was used. At 48 hours, the patient presented pain relief (0/10 visual analog scale) and hemiparesis improvement (4/5 strength), the hemoglobin electrophoresis showed a hemoglobin S percentage of 8% afther the procedure. In Colombia, this is the first case described in the literature of Red blood cell exchange (RBCX) in a patient with sickle cell disease with peripheral venous access and successful results.

Keywords: Anemia, Sickle Cell, Erythrocyte, Transfusion, Stroke, Hemoglobinopathy.

Introduction

Sickle cell disease is a public health problem in Colombia, it is estimated that in Valle del Cauca approximately 4.5% of newborns are at least heterozygous for the hemoglobin S gene[1]. Therefore, the Colombian regulations declared it an orphan disease and began its surveillance through the National Institute of Health[2], unfortunately it is the cause of 82.7% of deaths from orphan diseases in the population that is recognized as mulatto, Afro-Colombian or Afro-descendant[3]. Sickle cell disease like other hemoglobinopathies is the result of human interaction with the parasite of the genus Plasmodium that cause of malaria, since this mutation confers resistance to infection and has occurred at least 5 times in the history of humanity in independent populations and without genetic flow between them[4].

The complications of the disease are systemic, they include an increased risk of suffering from pneumonia, asthma attacks, kidney failure, invasive infections by encapsulated microorganisms, meningitis and cerebrovascular accidents, among others[5]. The most feared complications and of recent attention are silent cerebrovascular infarcts that can occur in up to 35% of patients, cognitive impairment and non-silent cerebral infarcts which are associated with a significant deterioration of functional capacity and quality of life[6]. Therapeutic strategies consist of chronic red blood cell transfusions and Red blood cell exchange in order to reduce the percentage of hemoglobin S to a value less than 30%[7]. One of the main technical difficulties of the Red blood cell exchange lies in the need for central venous access to connect automated blood exchange devices to patients[8,9]. However, advances in technology are capable to perform apheresis procedures with peripheral venous access, thus reducing administrative barriers, costs, improving patient safety and optimizing the cost-utility and cost-effectiveness of care[10]. This article describes the treatment of a patient with a diagnosis of sickle cell disease and cerebral infarction who was successfully treated with blood exchange in the first 12 hours of care with peripheral venous access in the upper limbs.

Case Description

A 15-year-old male patient, originally from and from the municipality of Florida, Valle del Cauca, diagnosed with sickle cell disease (HbSS) on September 17, 2017, with a hemoglobin S percentage of 95%, with poor adherence to the treatment and no medical control. In July 2019 consulted at the local emergency room in his municipality for presenting 6 days of left hemiparesis associated with intense musculoskeletal pain, with an intensity on the visual-analog scale of 10/10, upon physical examination the patient was admitted with blood pressure 100/60, heart rate 110 beats per minute, respiratory rate 24 beats per minute and

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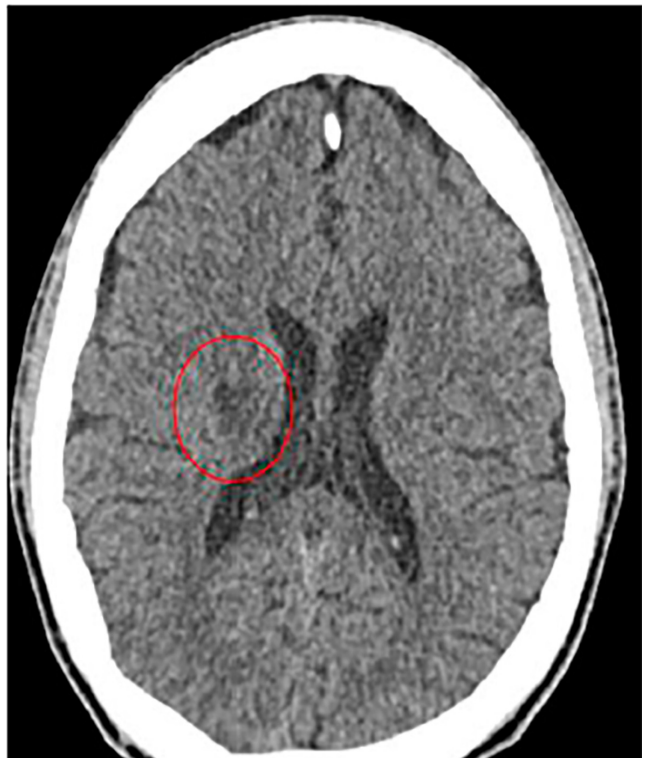
temperature 37.5° C. The thorax presented bilateral normoexpansion, no dullness to percussion, vesicular murmurs were heard in all lung fields, no egophony or whispered pectoriloquism was evidenced. A point of maximum intensity was palpated in the left midclavicular line, no thrill or turbulence was palpated, the heart sounds were rhythmic and regular without murmurs, the first and second sounds were reinforced, S3 was not auscultated, no murmurs were evidenced in the aortic mitral focus, pulmonary or tricuspid. The abdomen presented preserved air-fluid sounds, no signs of peritoneal irritation such as Blombeg, Rovsign, McBurney, rebound, psoas sign, thallus percussion were evident. The Murphy was negative. Costovertebral percussion was negative, as was renal percussion. The ascites wave was negative. Hepatomegaly and splenomegaly were negative. Neurological examination revealed left hemiparesis with a strength of 3/5 without cranial nerve involvement and left patellar and achillian hyporeflexia, without signs of meningeal irritation such as nuchal rigidity, Kernig or Brudzinski.

He is immediately referred to the Hospital Universitario del Valle, in the emergency service of this institution he is evaluated by pediatric neurology and hematology who find a patient with the described neurological deficit. Supportive medical management with intravenous fluids, morphine, high-flow oxygen was provided and requested a simple cerebral computed tomography scan and blood tests. The hemogram showed 15,130 leukocytes / µL, hematocrit 34.4%, hemoglobin 10.7 g / dl, platelets 583,000 per microliter, in the peripheral blood smear sickle cells were found. Clotting times were normal. Bilirubins were high, lactate dehydrogenase in normal ranges, as were electrolytes and creatinine. The computerized axial tomography revealed a focal hypodense area that compromised the body of the caudate nucleus and the anterior arm and part of the posterior arm of the internal capsule on the right side, in relation to the acute ischemic infarct (Figure 1). He was reassessed by the pediatric hematology service who considered performing automated blood exchange. The patient was transferred from the emergency room to the hematology hospitalization service where the vasculature of the upper limbs was evaluated and the nurse obtained intravenous access through cannulation of the antecubital veins. The patient was connected to the Optia Spectra(R) machine. Based in the previous hemoglobin S percentage, height and weigth, an erythorcyte volume of 3200 ml was calculated for a final hematocrit of 30%. The duration of the procedure was 3 hours, the flow was 45ml/minute and 12 units of packed red cell blood cells, leukoreduced, isophenotype for Rh and kell of were required. There were no adverse reactions to the transfusion, the patient tolerated the procedure satisfactorily.

Figure 1. Simple computerized axial tomography of the brain



Source: Radiology Service of the Hospital Universitario del Valle.

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Simple brain tomography showing a focal hypodense area that involves the body of the caudate nucleus and the anterior arm and part of the posterior arm of the internal capsule on the right side, in relation to an acute ischemic focus (red ellipse).

48 hours after the procedure, a new hemoglobin electrophoresis was performed, which reported a hemoglobin S percentage of 8% with absolute pain relief (0/10 visual analogue scale) without the need for opioid analgesics. On physical examination he persisted with left hemiparesis, but with improvement (strength in left hemibody 4/5). The hemogram reported hemoglobin of 10.5 g / dl and hematocrit of 30%. It is evaluated by physical medicine and rehabilitation who considered that the patient achieved progress and physical and occupational therapy was ordered in order to avoid loss of functional capacity and independence in activities of daily living.

Discussion

Red blood cell exchange (RBCX) in the treatment of sickle cell disease is a widespread practice in high-income countries, in the United Kingdom it is estimated that the use of existing machines will save that health system 13 million pounds[9,10]

. In Colombia, this is the first reported case of the use of blood exchange and its implementation with peripheral venous access in a patient with ischemic stroke, which, since it can be performed with peripheral catheters (Introcan Safety® 3 18G), reduces access barriers. In the present case, the use with the greatest evidence of automated blood exchange is shown, since its superiority over simple transfusion has been demonstrated in terms of mortality, technical efficiency (it allows achieving a hemoglobin S goal of less than 30%) and ease of operation in sickle cell patients with ischemic strokes[11]. Red blood cell exchange, poses challenges, firstly including it among specialists as a therapeutic tool. Secondly adapting the installed capacity of health services from the south west of Colombia. Thirdly, the demand and effort on the part of civil society to satisfy the demand for blood required by the procedure. Lastly, the standardization of a clinical practice guide that includes and disseminates it [10]. Despite the fact that the present case represents an advance in treatment, sickle cell disease persists as a public health problem[11]. The case described shows how the social determinants of health impact on access to treatment of the disease, which includes rapid dispensing of drugs (which in Colombia is precarious), access and opportunity to medical consultation with specialists who are experts in the disease, since the patient lost contact with the health system. This highlights the importance of a primary health care approach that includes orphan diseases within the collective imagination of the population, which allows decent and quality care for patients. In this sense, Colombia lags behind other countries where strategies and programs have been established that range from neonatal screening to work and community integration of patients[12,13].

Conclusions

Automated blood exchange with peripheral venous access as a therapeutic tool allowed the treatment of one of the most devastating complications of sickle cell disease in a young patient who presented with an ischemic stroke.

Patient perspective

The patient refers that the blood exchange improved the pain and he sees the importance of carrying out the medical controls “if it took away the pain I must return to medical control”, however, he also refers to fear of discrimination in the school environment “I want to go back to school I’m in the seventh year of high school and I haven’t been able

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to continue because the pain didn't leave me, but I don't want them to make fun of me ".

Conflict of interest

The authors declare no conflict of interest.

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