

Congenital Gangrene of the Upper Limb in a Big Premature: A Case Report and Literature Review

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Abstract: Congenital gangrene limb is a rare condition. In the majority of cases, no cause is identified and its treatment is not codified. We report the case of 28 weeks of gestation premature male of a nondiabetic mother who presented at birth ischemic gangrene of the right upper limb with severe respiratory distress hospitalized in neonatal intensive care unit, undergoing trans-humeral amputation in front of the installation of a febrile syndrome. We discuss the different aetiopathogenic theories based on literature review.

Keywords: congenital gangrene; premature; ischemia; febrile syndrome; amputation.

INTRODUCTION

Congenital gangrene limb is rare [1,2] and very few cases have been reported in the literature since the first described by Martini in 1828 [1]. It is serious because that could jeopardize the vital and functional prognosis. In most cases, no cause is identified [1,3] and the treatment is not codified [4].

We report a case of congenital gangrene of the right upper limb in a premature baby of 28 weeks of gestation and discuss the diagnostic, etiological, therapeutic and evolutionary aspects by a review of the literature.

CASE REPORT

A male premature was born after 28 weeks of gestation, from a 32-year-old mother, third gesture and third par, not known diabetic or hypertensive. She benefited from four prenatal consultations and the serological test was negative. The birth was vaginal with premature rupture of membranes (PROM), in cephalic presentation, the APGAR score was 8/10/10 at the 1st, 5th and 10th minute, weight 1260 g. At birth, the gynecologist found an ischemic aspect of the forearm and right hand with areas of cutaneous necrosis, cyanosis extended to the lower third of the arm, absence of humeral, radial and cubital pulse. The patient also had a 3/10 respiratory distress according to the silverman score, for which he was transferred to a neonatal intensive care unit for the management of prematurity and respiratory distress. After admission to the neonatal intensive care unit, the patient was put in an incubator with monitoring and oxygen therapy.

The clinical examination found a cold pulseless limb, and absence of mobilization of the same limb, placards of cutaneous necrosis, fingers in claw,

heart rate at 164 beats / min, respiratory rate at 58 cycles / min. A blood assessment on admission revealed the presence of anemia with hemoglobin at 8 g / dl, serum sodium and serum potassium normal, normal crash test with normal fibrinogen and D-dimer levels.

A Doppler-echo of the upper limb realized objectified: arterial flow present at the right axillary artery and the brachial artery up to its 1/3 lower, we do not perceive any flow. With no flow at the radial and ulnar arteries. A Transthoracic cardiac ultrasound made back normal.

The treatment consisted of a continuous injection of unfractionated heparin curative dose with aPTT oversight and a triple antibiotic therapy based on ciprofloxacin, vancomycin and amikacin. The evolution was marked by appearance the day after his admission of a febrile illness with a temperature at 38.6 degree, an amputation of the right arm to the middle third was performed to avoid septic shock. The patient died on day 15 of his stay with nosocomial pneumonia.



Fig-1: image showing advanced ischemia of the upper limb a few hours after birth

DISCUSSION

Congenital gangrene limb is exceptional and raises the problem of its etiology, which remains unknown in the majority of cases [1, 3]. Many aetiopathogenic hypotheses are emitted including uterine abnormalities, abnormal presentations with externalization of the upper limb, shoulder dystocia, oligohydramnios, amniotic flanges, infections, cord circulars, umbilical catheterization, maternal diabetes, prematurity, dehydration, polycythemia, coagulopathies, transfusor-transfused syndrome, obstetric trauma [1,5,2,3,6,7]. For Tanvig *et al.* [6], Arshad *et al.* [8], the starting point for this gangrene is ischemia due to intrauterine compression or thromboembolism.

Of the 151 cases of acute limb ischemia reported by Kayssi *et al.* Over a 13-year period, only 14 cases (9%) were non-traumatic [9]. The literature of this condition is often confined to clinical cases [10]; reflecting its rarity. The positive diagnosis of acute ischemia is clinical. Imaging invalidates the diagnosis for a pre-therapeutic purpose. Doppler ultrasound is often sufficient, but in some cases, arteriography or CT angiography can be used [11].

In our case, the diagnosis was based on the clinic and the Doppler ultrasound confirmed the diagnosis. Acute ischemia in children is a polymorphic condition. The etiologies are multiple, well dominated by arterial catheterization in 91 to 92%. Non-traumatic etiologies vary according to age group [11]. Intrauterine ischemia occurs before birth, lesions are visible from birth and most often involve the upper limb. Only about twenty cases of intrauterine ischemia have been reported in the literature [12].

The case reported by Carr *et al.* [13] nevertheless approaches ours. Indeed, it was a premature birth, which featured ischemic gangrene of the hand and right forearm associated with infarction of the right cerebral hemisphere discovered at the age of seven months [12].

Favoring factors are numerous and are of malformative origin, infectious, obstetrical or metabolic. The maternal diabetes is most often incriminated. The different etiologies reported are represented in this table [12].

| | |
|--------------|---|
| malformative | Prematurity, congenital heart disease, amniotic flanks, congenital arterial abnormality, transfusor-transfusion syndrome in twins |
| infectious | Syphilis, rubella, chickenpox |
| metabolic | Maternal diabetes, thromboembolic disease, protein C deficiency, thrombocytopenia |
| obstetric | Pre-eclampsia, eclampsia, obstetric trauma, premature rupture of membranes, strangulation of the limb by the umbilical cord, oligohydramnios, presentation abnormality in utero, placental infarction |

The element common to all these etiological factors is an arterial insufficiency leading to ischemia of the limb [13]. This deficiency is the result of either intrauterine compression or arterial thrombosis [12].

According to this table, several etiologies can explain the ischemia of the limb in our case namely the great prematurity and premature rupture of membranes. And according to the results of Doppler thrombotic origin is also implicated, thrombosis of the brachial artery, the thrombus could come as in the case described by Carr *et al.* [13] from a placental infarction, released into the fetal circulation, migrated through the foramen ovale to the left ventricle and then ejected into the aorta. The brachiocephalic arterial trunk being the first collateral branch of the aortic arch, it is the first conduit borrowed by a thrombus from the heart [12]. Although the anatomopathological examination of the placenta was not performed in our case, it remains a hypothesis that can explain the thrombotic process in our case. The placental abnormality may be infarction or fetal thrombotic vasculopathy [14].

Acute ischemia of newborns related to heterozygosity of the MTHFR 677C-T gene mutation, and a mutation of the factor V Leiden gene, have been reported [11]. This polymorphism may be at the origin of a thermolabile protein with a reduced enzymatic activity that can alter the metabolism of folate and induce a moderate rise in the plasma level of homocysteine. This amino acid has a deleterious effect on the endothelial cell and promotes thrombotic complications [14].

Indeed, the homozygous state of the MTHFR 677C-T gene mutation considered being an arterial and venous thrombogenic factor, but the causal link remains controversial. For the heterozygous state, some authors mention it as a thrombogenic factor by the slight increase in homocysteine. However, recent studies have concluded that there is no clinical significance for this condition. In our case, the genetic anomaly could not be formally retained in the absence of a homocysteinemia dosage [14].

Concerning in utero thrombosis of limbs, only 4 cases having as risk factor a 677C> T polymorphism of MTHFR gene mutation have been published. Alioglu *et al* published the case of a newborn who presented at birth ischemia of the right lower limb. Thrombophilia workup revealed a heterozygous status of the MTHFR

677C-T gene mutation associated with mild hyperhomocysteinemia. Khriesat *et al* reported the case of a premature baby was born after 33 weeks of gestation who presented at birth arterial ischemia of his right upper limb. The etiological workup confirmed the presence of a double heterozygosity of the MTHFR 677C-T gene mutation and factor V of Leiden. Homocysteine was slightly elevated. More recently, McKasson and Golomb have reported the cases of two newborns who had upper limbs ischemia at birth. The genetic study showed a homozygous state for the MTHFR 677C-T gene mutation in both cases [14].

In the literature, there are other thrombophilic abnormalities, constitutional or acquired, diagnosed in newborns who presented at birth arterial limb ischemia: heterozygous protein C deficiency, factor V Leiden in the heterozygous state, low activity of plasminogen and presence of anti-phospholipid antibodies [14].

In addition, the administration of some drugs such as indomethacin during pregnancy, hypernatremic dehydration may be associated with some forms of acute ischemia of the newborn. [11]. Daskalaki *et al.* Reported acute ischemia in an infant due to infection with *Streptococcus beta-hemolytic group A* [10]. But literature reports cases of idiopathic acute ischemia [11].

There is no consensus on the management of acute ischemia in children. But the ability of this particular group of patients to rapidly develop arterial collateral, advocates more and more towards a non-interventional first-line attitude even in case of arterial trauma [11]. Medical treatment consists of unfractionated heparin or enoxaparin, tissue plasminogen activator, aspirin or warfarin depending on the context. In case of failure, other minimally invasive treatment such as sympathetic nerve blocks to lift arterial spasm. The surgery consists of an embolectomy or most often bypass surgery, but requires knowledge in microsurgery to ensure the quality of the gesture [11]. New drugs such as streptokinase and urokinase have also demonstrated their effectiveness. Indeed; Brevière GM *et al.* had reported a series of five newborns treated effectively by urokinase administered in infusion at a dose of 1000 to 4000 U / kg per hour. This antithrombotic treatment is indicated in reversible ischemia and cannot be used in the gangrene stage [12]. Aydin *et al* in Turkey have successfully treated a case of congenital gangrene of the upper limb through

negative pressure therapy or vacuum-assisted closure therapy [3].

Limb amputation is reserved for outdated forms when there is a furrow delimiting necrosis [12]. In our case, the patient did not spread to medical treatment and amputation was performed in front of the installation of febrile syndrome for avoid a state of septic shock.

CONCLUSION

Congenital gangrene limb is rare. His dramatic outcome is amputation. It is a tragedy for the functional future of the newborn, and a major psychological trauma to parents. It is a rare and complex phenomenon, whose etiology remains obscure. The diagnosis must be made at birth to allow adequate and multidisciplinary care. But, one must keep in mind that idiopathic acute ischemia exists.

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