CASE REPORT

Congenital gangrene of the extremities in a newborn

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Abstract

Gangrene of the extremities in the newborn is extremely rare at birth. Less than 100 cases have been reported worldwide. Its etiology is obscure in many cases; however, some factors have been associated with it in the newborn, which include vascular injury and embolism. We report a case of a baby with congenital bilateral lower limb gangrene caused by thromboembolic phenomenon from retroplacental hematoma following abruptio placentae and highlight the challenges of managing such condition in resource-poor setting.

Key words: Extremities, gangrene, newborn

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Introduction

Intrauterine vascular insufficiency with bilateral lower limb gangrene at birth is a rare but well recognized phenomenon. Since the first case described by Martini in 1828, according to Gross,[1] less than 100 cases[2] have been reported so far. The etiology is obscure in most cases. This is being reported due to the rarity of the condition and difficulties of managing it in resource-poor setting.

Case Report

A 66-hour-old baby girl was referred to us on account of blueness and inability to move both the lower limbs since birth. The baby was a product of 36 weeks gestation. Pregnancy was supervised in a private hospital and was uneventful until the 36th weeks of gestation, when mother developed spontaneous vaginal bleeding which lasted for 38 hours before delivery. There was no history of hypertension, diabetes mellitus, renal diseases, thrombotic or hemorrhagic tendency in the mother, siblings, or other family members. Drug use during pregnancy was limited to routine antenatal drugs. Antepartum ultrasonography revealed abruptio placentae with a normal live fetus. Baby was delivered in the hospital, where pregnancy was supervised, by emergency caesarian section and had Apgar scores of 3 and 5 at 1 and 5 minutes, respectively. Prompt airway management viz-à-viz clearing of the airway and positive airway pressure ventilation was instituted. No history of umbilical catheterization or administration of parenteral drugs. She responded to the resuscitation but her lower limbs remained limp. At the age of 12 hours, the left lower limb became bluish up to the groin and swollen while the right leg had blackish areas on the last three toes and back of the leg up to the mid-thigh. This prompted the private hospital to refer the patient to the University Hospital.

At presentation, the baby was moderately dehydrated, jaundiced, and pyrexic. The left lower limb was edematous, bluish, and cold up to the hip joint, with blisters on the dorsal aspect of the foot and lateral part of the leg, the largest blister measuring 3 × 4 cm. There was a line about 2 cm below the inguinal ligament demarcating the healthy proximal portion from the dead distal part [Figure 1]. The left dorsalis pedis, popliteal, and femoral arterial pulses were absent. The right lower limb was hyperaemic with bluish areas on the posterior

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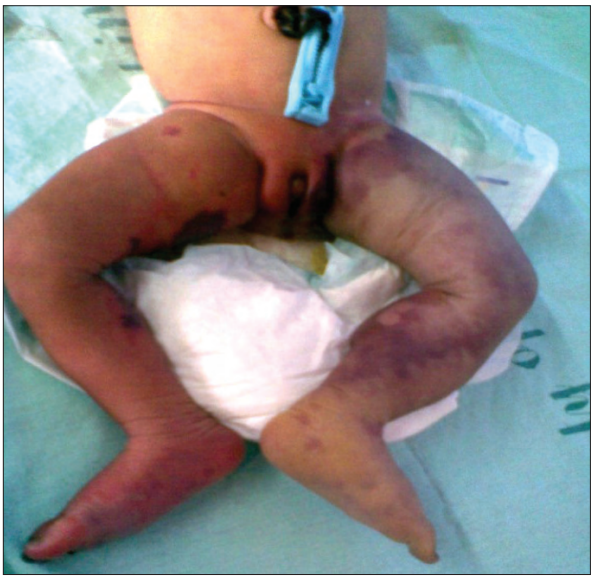

Figure 1: Bilateral lower limb gangrene in a newborn

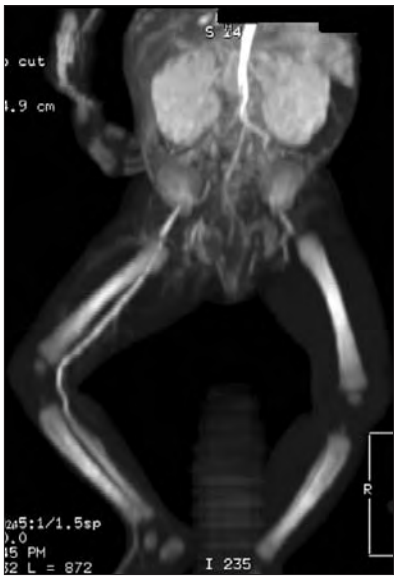

Figure 2: Contrast angiography showing right femoral artery stenosis and absent left femoral and popliteal vascular markings

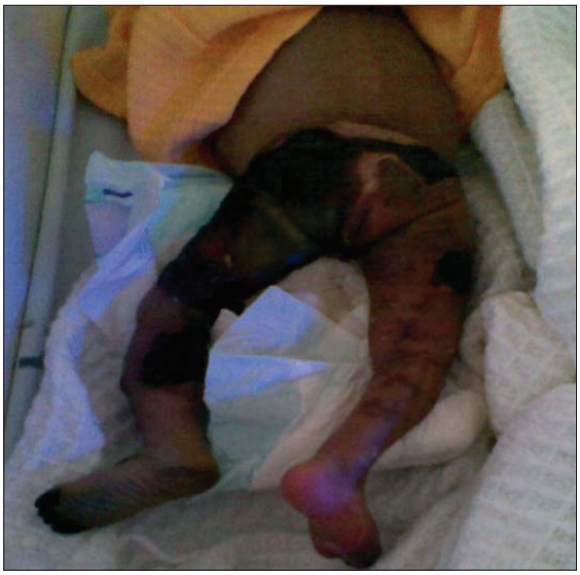

Figure 3: Progression in the limb gangrene

aspect, up to the gluteal area. The first, second, and third lateral toes also showed dark bluish discoloration.

The arterial pulsations in the right lower limb were barely palpable. The leg was tender with no spontaneous movement. Examination of the systems was normal except for hepatomegaly of 4 cm and vulval edema. Her blood pressure was 60/40 mmHg.

Laboratory investigations revealed hemtocrit of 41%, total leucocyte count of 9.0 × 10^9/l with 60% neutrophils, 24% lymphocytes, 1% myelocytes, and 5% stab forms. The platelet count was 326 × 10^9/l. Total serum bilirubin was 326 μmol/l and conjugated fraction was 8 μmol/l. The prothrombin and activated partial thromboplastin times were similar to the control, while the serum biochemistry was within normal range. Doppler studies were not possible but contrast computed tomography angiography performed revealed stenotic right femoral artery. In addition, the arteries of the left lower limbs were not visualized up to the level of the division of the common iliac artery [Figure 2].

An impression of bilateral lower limb gangrene was made. The limbs were elevated and warm compress applied. Intravenous tissue plasminogen activator was prescribed at 0.1 mg/kg/h but it was unavailable. Subcutaneous enoxaparin 1.6 mg/kg 12 hourly, intravenous antibiotics and oral ibuprofen was commenced as well as intensive phototherapy, fluid, and caloric supply.

The right femoral and popliteal pulsations became stronger by the third day of commencing enoxaparin but the left lower limb gangrene progressed [Figure 3].

Amputation of the gangrenous parts could not be done due to its extensive nature. The baby developed features of multiple organ failure and died on the 13th day of admission. The parent declined offer for post mortem autopsy.

Discussion

Gangrene of the lower limbs is rare in newborns.[3] This is the first case seen in our center in over 35 years of its existence. Although the cause of gangrene in newborn is either obscure or unknown in many cases, careful analysis of the sequence of events surrounding the insult may offer a clue in determining the likely etiology.

Ischemic changes present at birth suggests intrauterine etiopathogenesis.[4-6] Since our patient's limbs showed ischemic changes at birth, some peripartum events must have been responsible. Acute limb ischemia is most commonly caused by thromboembolic phenomenom.[7] The possible cause in the present patient is an embolus dislodged from the retroplacental haematoma that resulted from abruptio placentae. Microemboli on the maternal side of the placenta could be driven by the high-pressure (70 mmHg) in the spiral arteries into the low-pressure (10 mmHg) intervillous space.[8] The continuing influx of blood from spiral arteries exerts pressure on the content of the intervillous space.[8] With separation of the placenta, the basal membrane of syncytiotrophoblast and fetal capillaries at the exchange zones[8] could be disrupted thus weakening the placental barrier and allowing the emboli easy access to the fetal capillaries and hence the umbilical vein and fetal circulation. The emboli could have gained access to the fetal aorta and got trapped in the femoral arteries. A strong association between placental thrombosis and fetal somatic thrombi has been described by Kraus and Acheen.[6] In that study, autopsy findings demonstrated that 37.5% of fetuses with significant placental thrombi had associated somatic thrombi.

The angiographic finding of right femoral artery stenosis in our patient suggests the presence of underlying congenital vascular abnormalities. Gangrene of lower limb secondary to developmental abnormality of femoropopliteal artery was reported by Hefelfinger *et al*.[9] in 1971. Therefore, detailed radiological evaluation of the vascular tree of newborns with gangrene is mandatory. The presence of congenital vascular malformations in the milieu of a potentially thrombogenic circulatory changes at birth[10] and the hypofibrinolytic neonatal state (characterized by dysfunctional and decreased concentration of plasma plasminogen and tissue plasminogen activators as well as severe transient deficiency of antithrombin III, protein C, and protein S)[10] places the newborn at risk of thromboembolism.

Babies developing peripheral gangrene often have a history of abnormal delivery and perinatal asphyxia,[4] as in the case of our patient. The role of asphyxia in the pathogenesis of gangrene in this patient was however uncertain. Although some authors[2-5,10-12] have cited other factors like polycythaemia, hypernatraemia, systemic infection, hypothermia, umbilical catheterization, hyperglycemia, maternal diabetes mellitus, congenital heart diseases, amniotic constriction bands, and constriction by umbilical cord in the pathogenesis of gangrene in the newborn, none of these was present in our patient. In addition, congenital thrombophilia, though a rare condition, is a remote possibility. Assaying for plasminogen, anti-thrombin III, protein C, and protein S activities would have been a worthwhile venture, but ours is a community that lack the wherewithal for such esoteric investigations.

Thrombolysis could have been achieved with recombinant tissue plasminogen activators but these were not available, hence an anticoagulant, enoxaparin, was relied upon to prevent further thrombotic episodes. Combination of a thrombolytic agent with an anticoagulant was recommended by Arshad and McCarthy[13] in neonates with limb thrombosis. Complete clot lysis is usual in 75% of cases.[13] The poor response in our case could be due to lack of plasminogen activator in our treatment modality.

Early surgical intervention is usually not advised, because often the gangrene is yet evolving and not properly demarcated. In our case, early surgical intervention was not undertaken with the above in mind, and later, the extensive nature of the gangrene and its subsequent evolution to involve the posterior aspect of the trunk up to the level of the third lumbar vertebrae precluded surgery. Surgical intervention for such extensive gangrene is a task fraught with extremely high risk of intra and postoperative morbidity.[14] Salvation of the patient and the gangrenous limbs may be possible in technologically advanced communities where facilities for intensive interventions abound. Highly intensive treatment modalities required for newborns with extensive gangrene of this nature are often not available in resource poor communities, thus posing a great challenge in the management. Supplies of cost effective and technologically appropriate facilities for intensive care are therefore of urgent need in developing countries.

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