Combined Intrauterine Vascular Insufficiency and Brachial Plexus Palsy: A Case Report

Catherine Sinclair · Peter M. Murray ·
Sarvam P. Terkonda

Received: 4 June 2007 / Accepted: 17 August 2007 / Published online: 21 September 2007
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Abstract A unique case of combined intrauterine vascular insufficiency and complete brachial plexus palsy is described in a newborn delivered by cesarean section. Intrauterine vascular insufficiency resulted in a right below elbow amputation at 3 weeks of age. Amputation length was preserved after a pedicled thoracoabdominal flap. Function of the C5, C6, and C7 nerve roots returned to normal by 3 months of age.

Keywords Intrauterine vascular insufficiency · Ischemia · Obstetric brachial plexus palsy · Oligohydramnios

Introduction

Intrauterine vascular insufficiency is a rare clinical problem with a heterogeneous etiology. Less than 100 cases have been recorded [32] and only one-third have occurred in utero [5]. Irrespective of the cause, the end result of intrauterine vascular insufficiency is decreased perfusion and local ischemia, which can lead to tissue necrosis.

Another rare birth phenomenon is brachial plexus injury after cesarean delivery. The purpose of this case report is to present a child delivered by cesarean section with intrauterine vascular insufficiency and combined C5–T1 brachial plexus palsy.

Case Report

A 1.58-kg female was born to a healthy 27-year-old G1, P0 female after a 32-week gestation. The pregnancy was uncomplicated until 1 week before delivery when the mother was noted to have hypertension. At that time, no fetal movements were found on ultrasound and an emergent Cesarean section was performed. The infant was delivered without complication and the activity, pallor, grimace, appearance, respiration (APGAR) scores were 5/9 at 1 and 5 min.

At birth, the right arm was noted to be flaccid with hand swelling and blue discoloration to the midhumeral level. The brachial artery pulse was appreciated with Doppler to the level of the antecubital fossa. There was a weak radial pulse by Doppler, but no ulnar pulse. By day 7, the right upper extremity had demarcated at the level of the proximal forearm with swelling and mottling (Fig. 1). The digits were black indicating full thickness skin necrosis (Fig. 2). Initial treatment consisted of elevation and Silvadene dressing changes to the right antecubital fossa.

Protein C and protein S lab studies were normal. An initial thrombocytopenia of 47,000 platelets normalized by day 10 of life. The infant did not require platelet transfusion. Birth hematocrit was 57%. The infant had a high nucleated red blood cell count of 719 with a corrected white blood cell count of 11,000. Head CT was unremarkable. A positive Moro's reflex was noted on the

C. Sinclair
Ohio State University,
Columbus, OH, USA

P. M. Murray (✉)
Department of Orthopedic Surgery,
Mayo Clinic College of Medicine,
4500 San Pablo Road,
Jacksonville, FL 32224, USA
e-mail: murray.peter@mayo.edu

S. P. Terkonda
Mayo Clinic College of Medicine,
Jacksonville, FL, USA

Figure 1 Appearance of the right forearm at 1 week of age showing full thickness skin defect with obvious necrosis at the below elbow level.

left. No spontaneous shoulder or elbow motor activity was noted in the right upper extremity. A diagnosis of C5–T1 complete brachial plexus palsy was made. Upon initial evaluation, it was evident that the vascular insult had already occurred. The forearm was not tense and there were no physical findings indicating an acute compartment syndrome necessitating fasciotomy.

At 3 weeks of age, debridement of the forearm and disarticulation of the wrist were performed. Local wound care resulted in adequate granulation tissue formation. At 9 weeks of age, a 7×3.5 cm thoracoabdominal fasciocutaneous pedicle flap was used for coverage to preserve forearm length (Fig. 3). Postoperatively, a methicillin-resistant *Staphylococcus aureus* (MRSA) infection developed and was successfully treated with IV antibiotics. Partial recovery of the C5, C6, and C7 muscular function was noted on physical examination as demonstrated by antigravity shoulder elevation to 90°, antigravity biceps flexion to 90°, and elbow extension.

At 12 weeks, the pedicle flap was divided and all wounds were closed. Neurologic examination revealed

Figure 2 Palmar view of the hand showing full thickness skin necrosis.

Figure 3 Preoperative design of the thoracoabdominal fasciocutaneous pedicle flap used for coverage of the amputation stump.

complete recovery of C5, C6, and C7 innervated muscle function. Physical therapy was initiated to maintain shoulder forward elevation and external rotation.

Between 6 and 12 months of age, numerous attempts at prosthetic fitting were unsuccessful because of patient intolerance. At 4 years follow-up, the patient has normal shoulder and elbow function and is otherwise healthy.

Discussion

Intrauterine vascular insufficiency presents with vascular compromise leading to tissue necrosis in the extremity. The diagnosis given to a neonate with a necrotic or ischemic limb is intrauterine vascular insufficiency, intrauterine gangrene, or neonatal Volkmann's ischemia [3]. Causes or potential causes of gangrenous limbs of the newborn are coagulopathy, thromboembolism, rubella, varicella, congenital heart disease, polycythemia, eclampsia, fetal sepsis, venipuncture, umbilical artery catheterization, birth trauma, protein C, S, and antithrombin III deficiencies, constriction band syndrome, umbilical artery catheterization, maternal diabetes mellitus, prolonged rupture of membranes [13], oligohydramnios, mechanical compression, administration of sodium bicarbonate, compartment syndrome, and dehydration [3, 5, 11, 13, 14, 18, 21, 22, 25, 27–30, 32, 33]. Intrauterine vascular insufficiency has been attributed to a lack of methylenetetrahydrofolate reductase (MTHFR), oligohydramnios, or abnormal anatomy of the uterus [3, 19]. Other factors predisposing to vascular compromise in the newborn are prematurity and intensive treatment for other life-threatening illnesses [21]. Gross concluded that the most common cause was emboli from thrombosis during closure of the ductus arteriosus and umbilical arteries [10]. Today, emboli are most commonly felt to arise after cannulation of the umbilical vessels [4, 5].

Armstrong and Page reported six cases of intrauterine vascular insufficiency with established necrotic tissue at birth resulting from either thromboembolism or compression of the limb during intrauterine life [3]. Johnson et al. reported a case of bilateral upper extremity ischemia, oligohydramnios, and compression of the fetus resulting in amputation [18]. Carr et al. presented a case of neonatal extremity gangrene resulting from intrauterine embolization of infarcted placental substances [5]. Tebes et al. described the case of an infant born in compound presentation (hand behind the head) with ischemic forearm necrosis ultimately requiring amputation [30]. Hsi et al. reported a case of right upper extremity gangrene in a newborn of a diabetic mother [14]. Turnpenny et al. provided a case of congenital left forearm gangrene requiring surgical debridement and skin grafting. Thromboembolism to the brachial artery before delivery was felt to be the cause of the gangrene [32]. Nazer et al. have reported on brachial artery thrombosis as a result of paradoxical embolism from the renal vein passing through a patent foramen ovale [23].

It is important to differentiate between neonatal gangrene and necrotizing fasciitis in the newborn as the latter requires a much more aggressive treatment to manage potential sepsis and tissue destruction [13]. Necrotizing fasciitis is a rapidly progressive group-A, β -hemolytic *Streptococcus* infection affecting the subcutaneous tissues. Advancing cellulitis of the limb and subcutaneous crepitus are typically noted on physical examination. The limb in necrotizing fasciitis, although swollen, will generally appear well-perfused and pulses are identifiable. In contrast, patients with compartment syndrome of the forearm or hand will have a tensely swollen extremity and poor digital perfusion. Cellulitis is not present unless a secondary infection has occurred [31].

Infants with necrotic limbs, as in our case, should be treated with dressing changes, hydration, nutrition, antibiotics, and heparinization, allowing the eschar to separate spontaneously [3, 10, 12, 13, 27]. Skin viability may be improved with the use of topical 2% nitroglycerin. Ischemic limbs should be evaluated by arteriography as indicated [6, 27, 29, 33]. However, arteriography examination in the neonate, particularly in premature neonates, is challenging. We have found physical examination, including capillary refill assessment and palpation or Doppler assessment of pulses, to be reliable in assessing vascular integrity in these patients. In our patient, the level of vascular compromise was easily assessed on physical examination. In cases of tissue necrosis, amputation should be delayed as neonatal tissues have the ability to establish collateral circulation and survive [3, 5, 21, 29]. In cases of established gangrene, amputation is necessary [22].

Along with intrauterine vascular insufficiency, this patient had a complete brachial plexus palsy. The most

common type of obstetrical brachial plexus palsy involves the C5, C6, and C7 nerve roots [26]. Less commonly, involvement of the C5–T1 nerve roots is encountered in 9–26% of all obstetric brachial plexus palsies [24]. Patients with an upper lesion of the brachial plexus have the best prognosis with recovery in 79–100% of patients [16, 20]. Those patients with a C5–T1 plexus palsy see demonstrable recovery in only 50% [16, 20].

It has been thought that obstetrical brachial plexus palsy was caused by shoulder dystocia and increased traction during vaginal delivery. More recently, obstetrical brachial plexus palsy after cesarean section has been described but is considered rare. al-Quattan et al. found only 1% of 1,561 newborns with obstetric brachial plexus palsy were delivered by cesarean section [1]. Gherman et al. identified 17 cases of obstetric brachial plexus palsy after delivery by cesarean section [7]. Nine cases were excluded because of breech presentation and two were excluded because of difficulty with the cesarean delivery. The six remaining infants presented with profound C5, C6, and C7 involvement persisting beyond 12 months of age. This situation demonstrated a different mechanism of brachial plexus palsy as traction at delivery was eliminated as a cause in these patients. Long-standing in utero stretching of the brachial plexus could lead to brachial plexus palsy. Alfonso et al. presented a case of a newborn delivered by uncomplicated cesarean section with a history of decreased fetal movement of the right upper extremity noted at 18 weeks gestation [2]. They highlighted that brachial plexus injury can occur subsequent to intrauterine forces, not only from traction occurring during delivery.

Because of the rarity of obstetric brachial plexus palsy in babies delivered by cesarean section, al-Quattan et al. recommended examining for other potential causes such as constriction bands, uterine constraints, intrauterine maladaptation palsy, congenital aplasia of the roots of the brachial plexus, congenital varicella of the upper limb, or anoxia [1]. Septic arthritis, spinal cord injury, ischemia, or compression have been cited as other causes of obstetric brachial plexus palsy occurring in utero [20, 24].

Gilbert et al. found a 0.03% incidence of brachial plexus palsy among infants delivered by cesarean [8, 9]. Obstetric brachial plexus palsy is usually associated with overweight newborns (>4,000 g) in cephalic presentations or low birth weight infants with breech presentation [8]. It is postulated that brachial plexus lesions can derive from spontaneous forces in utero, independent of force applied to the head at the time of delivery [15, 17]. Jennet et al. reported brachial plexus palsy in infants delivered by cesarean after transverse lie presentation [16]. They speculated that obstetrical brachial plexus palsy developed because of prolonged malposition in utero. Oligohydramnios (diminished amniotic fluids) could also predispose the fetus to shoulder compres-

sion from malpositioning [15], explaining not only the obstetrical brachial plexus palsy in our patient but also the apparent intrauterine upper extremity vascular event. It is also possible that this child's upper limb palsy was potentiated by upper limb ischemia, although cases of ischemic polyneuropathy in the newborn have never been reported.

We present a unique case of a neonate suffering intrauterine vascular insufficiency of the upper limb with concomitant C5–T1 obstetrical brachial plexus palsy. The only indication of fetal distress in our patient was maternal hypertension late in pregnancy and fetal hypokinesia on prenatal ultrasound. It is important to recognize that fetal upper limb ischemia and brachial plexus palsy can occur subsequent to such factors as fetal malpositioning or oligohydramnios necessitating emergent cesarean delivery. We recommend prompt evaluation for intrauterine vascular insufficiency with every attempt to salvage the limb. Also, a careful brachial plexus examination should be performed in these infants. If possible, amputations should be delayed until clear demarcation of tissue necrosis is discerned. Preservation of limb length by local tissue coverage techniques may be important for later prosthetic use.

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