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Upper Limb Ischemia at Birth: A Case Report from Saudi Arabia



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Abstract



Keywords

amniotic band syndrome; gestational diabetes; intrauterine limb ischemia; maternal diabetes; neonatal limb ischemia; The aim of this case report: is to present and discuss the clinical features, diagnostic challenges, management strategies, and outcomes of two cases involving upper limb ischemia in neonates. By examining these cases, we aim to contribute to the understanding of the underlying causes, diagnostic pathways, and potential interventions for neonates presenting with upper limb ischemia, emphasizing the importance of early diagnosis and appropriate management to minimize complications and enhance patient outcomes. Discussion Summary: ABS presents a range of fetal abnormalities caused by fibrous bands in the womb, leading to deformities and developmental interruptions. Limb constriction rings are common, potentially resulting in limb loss or digit deformities. Neonatal limb ischemia, particularly acute limb ischemia, is rare but associated with catheterization. In utero compression ischemia, often affecting the brachial artery, can lead to limb ischemia. Risk factors include maternal conditions like diabetes, dehydration, and more. Proper delivery planning is crucial for macrosomic infants. Diagnosis and management of neonatal limb ischemia are challenging. Doppler ultrasound is the primary diagnostic tool, with fetal CT enhancing prenatal assessment. Thrombolytic therapy, including alteplase and heparin, is a common treatment. Conclusion: This case study underscores the importance of timely intervention, thorough prenatal care, and multidisciplinary collaboration in obstetric and neonatal care. It also highlights the need for ongoing research to enhance our understanding of these conditions and improve patient care.

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1 Introduction

Amniotic Band Syndrome comprises a spectrum of fetal abnormalities linked to the presence of fibrous bands that seem to ensnare or encircle different fetal structures within the womb, ultimately causing deformities, irregularities, or interruptions in development (Seeds et al., 1982). Constriction ring anomalies in the limbs are commonly observed. These typically affect fingers or toes, sometimes accompanied by swelling in the distal part of the limb. These constrictions can result in the loss of limbs or digits, and on ultrasound scans, they may give the appearance of syndactyly (pseudosyndactyly). The outlook for individuals affected by these bands typically varies from case to case, contingent upon the severity of the condition and the timing of medical intervention (Society for Maternal-Fetal Medicine; Gandhi et al., 2019). The most common manifestations associated with amniotic band syndrome are constriction ring defects affecting the extremities, and these defects are typically present in almost all clear instances of the syndrome (Lamrissi et al., 2022). Clubfoot, although not a constriction-related defect, is found in approximately one-third of cases (Seeds et al., 1982; Baker & Rudolph, 1971). It's considered a rare diagnosis as a 5-year study in Australia identified 25 cases of amniotic band syndrome, and an additional 15 new cases were recognized through the study's selection process. This results in an annual prevalence of 2.03 cases per 10,000 births over the study duration (Bower et al., 1993; Romero-Valdovinos et al., 2014).

Acute limb ischemia is defined as a sudden decrease in limb perfusion that threatens the viability of the limb (Norgren et al., 2007). According to a study by Schmidt and Andrew, the incidence of ALI in the neonatal population is low, at 2.4 per 1,000 newborns admitted to intensive care units. Of these events, 89% are associated with catheterization (Schmidt & Andrew, 1995). In utero compression ischemia pertains to the constriction of the primary artery that supplies a limb due to an external factor during the fetal stage. This compression leads to ischemia and, if prolonged, can result in the necrosis of the limb. This phenomenon is specifically observed in the brachial artery and the upper limb, denoted as in utero Volkmann's ischemia. Risk factors for this condition encompass oligohydramnios and amniotic bands, which exert extrinsic pressure on the limb. While this occurrence remains relatively uncommon, it has gained substantial recognition, with numerous documented cases in the medical literature (Arshad & McCarthy, 2009; Ragland III et al., 2005; Carr et al., 1996).

2 Case Presentation

Case Report 1

A woman presented to the ER on May 29, 2023, at 12:10, she is a 30-year-old Saudi female, G2P1+0 (with one previous cesarean section), complaining of post-term pregnancy. Her Last Menstrual Period (L.M.P) was on the 20th of August 2022, and her Estimated Due Date (E.D.D) was expected on the 27th of May 2023, indicating a gestational age (G.A) of 40 weeks and 2 days. An early ultrasound on November 29th, 2022, marked her gestational age at 13 weeks and 3 days, confirming her current gestational age as 39 weeks and 2 days. She had a history of managing type 2 diabetes with metformin (Glucophage) and dietary adjustments. A previous

cesarean section had been performed due to concerns of fetal macrosomia. The patient was examined, showing a stable condition, consistent fundal height (38 weeks), cephalic fetal presentation, and pathological cardiotocography (CTG). An emergency cesarean section was conducted, resulting in the birth of a 4 kg baby girl who was intubated and diagnosed with right-sided upper limb ischemia, along with Erb's palsy.

The surgical intervention involved a full-term baby girl with right upper limb ischemia, evidenced by the absence of Doppler WAVE signals in the brachial, radial, and ulnar arteries. Vascular surgery was performed, including an intraoperative brachial artery procedure below the elbow to address spasms. A small arteriotomy incision was made, and thrombolytic therapy with the injection of 5cc of Actilyse was administered proximally and distally, resulting in improved arterial flow. The patient was kept on a heparin infusion and was eventually discharged home from the NICU in good general condition with fully restored limb function.

(Parents refused to include pictures of the baby)

Case Report 2

A woman was admitted on June 26, 2023, at 21:00, from the ER to the labor ward, she was a 35-year-old Saudi female, G7P5+1 (with all previous normal spontaneous vaginal deliveries), with her last delivery occurring 1.5 years ago. Her Last Menstrual Period (L.M.P) was on September 23rd, 2022 and her Estimated Due Date (E.D.D) was expected ON June 30th, 2023, putting her at precisely 39 weeks and 3 days of gestational age (G.A). She had complained of labor pain and decreased fetal movement over the past three days. Her past medical history included gestational diabetes mellitus managed through dietary measures (Suryasa et al., 2021). The patient had not received consistent antenatal care during this pregnancy. Upon examination, the patient appeared well, with normal vital signs. Her physical assessment showed a gestational age of 40 weeks, left occiput transverse cephalic presentation, big size baby, and a thick cervix with a 2 cm os. Cardiotocography (CTG) results were unreactive in the ER, leading to admission to the labor ward as nonreassuring CTG. The patient was subsequently taken for a cesarean section. The outcome was a 5.8 kg baby boy who was intubated and diagnosed with upper limb ischemia.

The surgical intervention involved a full-term baby boy with a macrocosmic birth weight. He presented with a left forearm lesion and multiple levels of amniotic bands at the elbow, mid-forearm, and wrist. The bands were incomplete at the elbow and mid-forearm but complete at the wrist. Doppler ultrasound showed good signals over the left axillary artery and brachial artery but none over the brachial artery at the elbow level, radial and ulnar arteries, and palmar arches. Urgent surgical intervention was necessary for the release of constricting amniotic bands.

As of August 18, 2023, the baby was breathing room air with no congenital anomalies. While the left upper limb was not completely healed, it was under the follow-up care of the plastic surgery team. The baby exhibited a good range of movement and was undergoing physiotherapy. He remained under observation at the NICU.

The two cases initially encountered challenges in reaching a final diagnosis, with considerations including amniotic band compression, venous obstruction, compartment syndrome due to swelling (Dandurand et al., 2009), and upper limb avulsion. However, a definitive diagnosis was ultimately established, leading to the subsequent surgical intervention.



Figure 1. "Case 2" appearance of the hand on birth showing the left upper limb, featuring edematous skin, black discolouration of fingers, and macerated skin on the arm and forearm

3 Results and Discussions

The aetiology of prenatal ischemia can be classified into different categories, including intrauterine limb ischemia stemming from prenatal thrombosis, embolism, and in-utero compression (Sentilhes et al., 2003). This condition is relatively uncommon, with a global incidence ranging between 1 in 1,200 to 1 in 15,000 births (Andrew et al., 2001).

Risk factors associated with prenatal ischemia include neonatal hypofibrinolysis, underdeveloped anticoagulation response, anticoagulant factor deficiencies, and maternal conditions such as diabetes, dehydration, polycythemia, sepsis, congenital heart disease, and anticoagulant system anomalies (Nowak-Göttl et al., 2001; Andrew et al., 2001; Carr et al., 1996). Emboli can arise from both maternal and fetal origins, with placental emboli traversing the foramen ovale, often causing arterial events, including lodging in the brachial artery leading to upper limb necrosis (Chandrasekaran, 2021).

In our cases, both mothers had diabetes mellitus (DM), one with type 2 DM was on Glucophage and diet, and the second was diagnosed during pregnancy and managed with diet alone (Ma et al., 2012; Arshad & McCarthy, 2009). Fetal emboli, commonly from ductus arteriosus or cardiac defects, can traverse the venous-arterial pathway (Wang et al., 2018). Neonatal ischemia in twins with twin-twin transfusion syndrome is related to the recipient twin's polycythemia (Winer et al., 2008).

On the other hand, in utero compression ischemia, typically affecting upper limbs, emerges from external arterial pressure, inducing ischemia and potential necrosis. Oligohydramnios and amniotic bands adding external pressure pose risks (Sentilhes et al., 2003). Like the cases presented, a history of gestational diabetes mellitus (GDM) might have contributed to the pathology of infantile ischemia (Ma et al., 2012). In the second case, the delivery was intricate due to the presence of a macrosomic infant, and the mother had previously undergone a cesarean section. The newborn's weight exceeded the 90th percentile, making scheduling a cesarean section before labor a prudent choice to mitigate potential complications (Chandrasekaran, 2021).

In a case report detailing six instances of intrauterine vascular insufficiency, it was observed that three of the expectant mothers had a diagnosis of Diabetes Mellitus (Arshad & McCarthy, 2009). Furthermore, the

study highlighted associations between intrauterine thrombosis and various factors, including: Emboli originating from the closure of the ductus arteriosus, Placental-related thrombosis, Thrombotic events in the umbilical arteries, Cases of prematurity, Instances involving maternal diabetes, which were associated with venous thrombosis, Dehydration-related cases, Occurrences linked to polycythemia, Cases involving deficiencies in Protein C, Protein S, and antithrombin III, Thrombotic events related to sepsis, Cases associated with congenital heart disease, Events linked to the presence of musculoelastic ridges within blood vessels (Turnpenny et al., 1992; Wang et al., 2018).

In a comprehensive analysis of 47 cases documented since 1941, the causative factors behind this condition were distinctly discernible in only six instances, with four cases directly attributed to compression resulting from the encircling umbilical cord (Torpin, 1965). Additionally, pregnancy-related hypertension emerged as a concurrent factor in seven cases, while oligohydramnios was noted in six instances. These findings underscore the complex and multifactorial nature of this condition, with potential contributions from various maternal and fetal factors (Sentilhes et al., 2003).

To delineate the types and levels of constriction, Type I, or mild constriction, is characterized by a superficial indentation in the skin and soft tissue, and it does not involve lymphedema. Type II, or moderate constriction, is associated with distal deformities, acrosyndactyly, or the presence of weakened or even discontinuous neurovascular or musculotendinous structures. Vascular function is often not compromised in Type II constrictions, and the presence of lymphedema may vary. Type III constrictions are characterized by severe, ring-like enclosures that progressively obstruct lymphatic, venous, or arterial circulation, potentially resulting in soft tissue loss. These Type III constrictions may or may not be accompanied by soft tissue damage (Weinzweig, 1995).

The diagnosis and management of neonatal limb ischemia remain challenging. In both cases discussed, medical practitioners encountered multiple potential diagnoses before arriving at the final diagnosis of upper limb ischemia. Initially, both cases faced challenges in reaching a definitive diagnosis, with considerations such as amniotic band compression, venous obstruction, compartment syndrome due to swelling, and upper limb avulsion (Arshad & McCarthy, 2009; Roy et al., 2002).

The primary imaging modality employed in neonatal arterial or venous thrombosis diagnosis is Doppler ultrasound. While contrast angiography is the established diagnostic standard for thrombosis in adults, its application in neonates is constrained by its invasive nature. Ultrasound offers distinct advantages, notably its noninvasive character and widespread accessibility. Nonetheless, diagnosing amniotic band syndrome (ABS) in utero presents considerable challenges. Fetal CT is employed as a secondary diagnostic tool to enhance prenatal assessment, enabling a detailed evaluation of the fetal skeletal structure (Haase & Merkel, 2008; Çöloğlu et al., 2014; Arshad & McCarthy, 2009).

An expert panel on the management of arterial thromboembolic events in neonates recommended that therapy should be individualized based on the extent of thrombosis and the urgency of the clinical situation. In the two cases presented, thrombolytic therapy, including alteplase and heparin, was administered post-surgery, which is a common treatment approach (Manco-Johnson et al., 2002; Arshad & McCarthy, 2009).

To achieve a favorable postoperative outcome, a well-organized newborn intensive care unit, along with surgical proficiency and access to microsurgical instruments, are essential factors (Özgenel et al., 2000). It's important to note that each case has a unique prognosis, which depends on various factors. In the two cases, one infant was discharged home in a satisfactory condition, while the other remained under NICU observation. This highlights the challenges in predicting prognosis, underscoring the need for further research to gain a comprehensive understanding. Notably, studies revealed that complete resolution of the clot within the first three months of age was observed in 68% (17/25) of the treated neonates and 86% (6/7) of the untreated neonates. An ischemic limb caused by a vascular spasm may exhibit an inherent tendency to spontaneously restore normal circulation, but long-term complications were identified, including chronic wounds affecting two patients in the affected limb and one patient presenting with limb length discrepancy (Wang et al., 2018; Arshad & McCarthy, 2009).

The importance of these two cases lies in their portrayal of issues that require immediate attention and intervention. The first case highlights the need for monitoring during pregnancy when the mother has type 2 diabetes and has previously had a cesarean section. The occurrence of limb ischemia and Erb's palsy in the newborn emphasizes the importance of assessing and managing birth-related injuries diligently. In these cases, the presence of diabetes mellitus and an otherwise uncomplicated obstetric history emphasizes the

significance of regular prenatal care and fetal monitoring (Van Elteren et al., 2011). The fact that the newborn experienced artery thrombosis and upper limb ischemia underscores the need for assessments during childbirth to promptly address any potential complications. Together, these cases emphasize the importance of intervention, collaboration among healthcare professionals, and thorough prenatal and perinatal care to ensure optimal outcomes for both mothers and babies facing diverse obstetric challenges (Sentilhes et al., 2003; Chandrasekaran, 2021).

4 Conclusion

These cases underscore the critical issues we face in obstetrics and neonatal care. The first case highlights just how important it is to closely monitor pregnancies involving mothers with type 2 diabetes, especially those who've had previous cesarean sections. The fact that the newborn in this case experienced both limb ischemia and Erb's palsy drives home the need for thorough assessments and careful management of any birth-related injuries.

These cases also emphasize the vital role of regular prenatal care and fetal monitoring, as well as the necessity for healthcare professionals to work together effectively. The presence of diabetes in one of the cases, alongside an otherwise uncomplicated obstetric history, really underscores the importance of consistent prenatal care to swiftly deal with any potential complications.

Moreover, the second case, where arterial thrombosis and upper limb ischemia occurred, highlights the need for very meticulous assessments during childbirth to address potential issues promptly. Together, these cases emphasize the crucial need for intervention, collaboration among healthcare providers from different disciplines, and thorough prenatal and perinatal care to ensure the best possible outcomes for both mothers and infants facing various obstetric challenges. They also underscore the importance of ongoing research to enhance our understanding of these complex medical conditions and to continually improve patient care in these situations.

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