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Case Report

Radial Nerve Palsy Secondary to Congenital Constriction Ring Syndrome in a Neonate: A Case Report and Literature Review

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ABSTRACT

Congenital constriction band syndrome (CBS) is a rare condition characterized by either partial or complete circular constrictions around limbs or digits. Possible consequences include acrosyndactyly of the digits, terminal amputations, and localized swelling with digital edema distal to the constrictions. Occasionally, a constriction band may lead to injury of a peripheral nerve. We report a case of CBS in a newborn presenting with radial nerve palsy caused by a constriction band at the level of the arm. Surgical release of the congenital constriction band was performed at 1 month of age, including neurolysis of the radial nerve. As a result, the nerve was decompressed, and hand function was restored. Given the rarity of radial nerve palsy as a complication of congenital constriction bands, this case enriches the limited literature by emphasizing the importance of early diagnosis and prompt surgical intervention to optimize functional outcomes. It also highlights the need for increased clinical vigilance and provides practical insights into managing isolated radial nerve palsies.

Key words: Congenital constriction band syndrome, constriction ring syndrome, amniotic band syndrome, radial nerve palsy, case report

INTRODUCTION

Congenital constriction band syndrome (CBS) is a rare condition characterized by a spectrum of congenital abnormalities, primarily affecting the limbs. It results from fibrous bands that partially or completely encircle fetal body parts, leading to constrictions that may cause acrosyndactyly, terminal amputations, or distal swelling with digital edema. [1]

This condition is a common cause of terminal congenital limb malformations, with a reported incidence of 1 in 15,000 live births. [2] It is also estimated to account for 178

cases per 10,000 miscarriages. It affects both sexes equally, and there is no evidence of hereditary transmission. [1, 2]

CASE REPORT

We report the case of a 1-day-old full-term newborn delivered spontaneously and vaginally, with a birth weight of 3.4 kg, who presented with CBS of the upper limb, causing radial nerve palsy. There was no maternal history of drug use or abuse during pregnancy.

Physical examination revealed a constriction band encircling the proximal third of the left upper arm, accompanied by radial nerve palsy, as evidenced by the absence of wrist and finger extension (**Figure 1**). Hand perfusion was intact, with both radial and ulnar pulses easily palpable. Left shoulder and elbow function were fully preserved. The child had no additional abnormalities or comorbidities. There was no family history of similar anomalies.

Electromyography and nerve conduction studies could not be completed as the patient was unable to tolerate the examination.

At the age of 1 month, the patient underwent surgical intervention. A complete circumferential excision was performed, removing the constriction band and underlying fibrous tissue within the subcutaneous layer and fascia, along with a 1 to 2 mm margin of surrounding healthy tissue. The wound was closed in a standard side-to-side fashion (**Figure 2**). In our case, early intervention with nerve decompression was successful, resulting in complete recovery of hand function without sequelae (**Figure 3**).

Figure 1: Constriction band located in the proximal third of the left upper arm, causing radial nerve palsy.

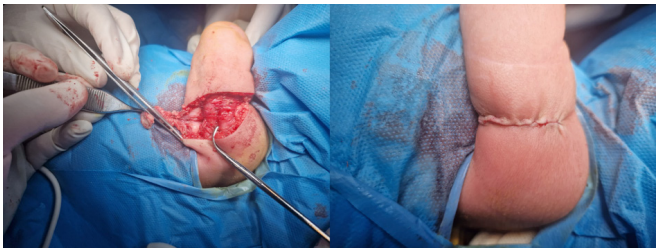

Figure 2: Intraoperative view illustrating the complete circumferential excision of the constriction band on the limb.

Figure 3: Long-term postoperative follow-up at 17 months reveals full functional restoration of the left upper extremity and complete resolution of radial nerve palsy.

DISCUSSION

Congenital CBS is known by various names, including amniotic band syndrome, congenital ring syndrome, constriction ring syndrome, amniotic band sequence, amniotic disruption sequence, Streeter's dysplasia, pseudoainhum, annular grooves or defects, ADAM complex (Amniotic Deformation, Adhesion, and Mutilation), and intrauterine amputation. [1–3]

Despite the existence of multiple theories, the etiopathogenesis of CBS remains unclear. There are two main and conflicting theories suggesting either intrinsic or extrinsic causes affecting the embryo or fetus.

The intrinsic theory, proposed by George Streeter, suggests that a defect in the subcutaneous germplasm leads to soft tissue necrosis followed by healing, resulting in the formation of constriction bands. According to this theory, a disruptive event occurs during blastogenesis, causing soft tissue to slough off without the formation of amniotic bands. Subsequent healing of the affected area leads to the development of constricting rings, which cause localized developmental defects.

The extrinsic theory, which is the most widely accepted, was proposed by Richard Torpin. He suggested that entanglement of the limbs in defects or free strands of amniotic fluid results in CBS, from which the other eponyms have originated. [2]

Clinical features

The prenatal history may include oligohydramnios, premature uterine contractions, and fluid leakage. However, the mother often reports an otherwise uncomplicated pregnancy. Although prenatal diagnosis by ultrasonography is occasionally possible, for most parents, the affected child comes as a surprise. Newer techniques, such as three-dimensional ultrasonography, may improve prenatal detection, and high-resolution ultrasound—along with advances in fetoscopic surgery—may eventually allow in utero treatment of amniotic bands. [3, 4]

The clinical presentation of CBS varies widely depending on the severity and depth of the constrictions. These may range from superficial, incomplete bands to deep, circumferential rings that can cause near-complete congenital amputations. Typically, the bands are located distally on the limbs; however,

multiple constrictions may be present within the same limb. Upper extremity involvement is more frequent than lower extremity involvement. The digits are the most commonly affected parts of the limbs, especially the three central digits (index, middle, and ring fingers). [1, 5]

Amputations may occur when constriction bands completely obstruct blood flow to a fetal limb, inhibiting its growth and causing tissue necrosis. Fenestrated syndactyly is the next most frequent anomaly, occurring in nearly half of all cases. The constrictions may cause digits to fuse with adjacent or nonadjacent digits, resulting in either complex syndactyly or acrosyndactyly (or both), with distal fusion and a proximal sinus or cleft between the digits. [1–3, 6, 7]

Syndactyly, hypoplasia, brachydactyly, symphalangism, symbrachydactyly, and camptodactyly have been reported in up to 80% of patients with congenital band syndrome. [7]

Significant neurovascular impairment may be present distal to the constriction band. Impaired venous and lymphatic drainage can cause swelling of the limb distal to the constriction. As the child grows, the band may worsen and become symptomatic. When constriction bands cross the body, they can result in a congenital fissure of the chest wall, a condition known as thoracoschisis. [2]

Around 7% of children with CBS present with craniofacial anomalies, including cleft lip and cleft palate. [1, 8] The prevalence of clubfoot ranges from 12% to 56%. These feet are often rigid and more difficult to treat. Most cases show peroneal muscle weakness (paralytic form) and are typically associated with ipsilateral constriction bands. The underlying cause is often deep bands that compress the peroneal nerve, leading to compression neuropathy, direct muscle injury, or even compartment syndrome. [1, 2, 9, 10]

Angular deformities, bone dysplasia, and pseudarthrosis may also occur beneath constriction bands in both upper and lower extremities. Additionally, leg-length discrepancy occurs in approximately 25% of patients.

The differential diagnosis includes Michelin tire baby syndrome, ainhum, hair thread tourniquet syndrome, transient neonatal radial nerve palsy (wrist drop), and brachial plexus injury. [2]

Peripheral nerve palsy

Peripheral nerve palsy Several rare case reports have described peripheral nerve palsies caused by CBS, emphasizing the critical importance of early diagnosis and intervention. Uchida and Sugioka identified three cases of nerve palsy among patients with constriction bands proximal to the wrist. [11] Weeks reported a neonate with total radial nerve palsy and dysfunction of the median and ulnar nerves due to a severe constriction band of the arm. [12] Weinzweig and Barr described successful early surgical release in a neonate with triple nerve involvement. [13] Mohan et al. presented a 1-week-old neonate with ulnar nerve motor dysfunction and triple nerve decompression, showing early recovery after surgery. [14] Jones et al. reported isolated ulnar nerve palsy with good long-term outcomes following early surgical release. [15] Richardson presented a case of delayed neurolysis in congenital radial nerve palsy, which was unsuccessful. [16]

Our case adds to this limited body of literature by illustrating that complete radial nerve palsy due to a congenital constriction band can still fully recover when managed with timely surgical intervention—even when not performed immediately after birth. Unlike some previously reported cases where urgent decompression was performed within days of life, our patient underwent successful decompression at 1 month of age, demonstrating that early intervention within the neonatal period remains effective for optimal functional recovery.

Treatment

There are three primary clinical scenarios in which congenital constriction bands require surgical intervention: acute release in neonates to salvage a compromised limb or digit; release of deep bands causing vascular and/or neurologic compromise; and cosmetic release of superficial bands. [1, 7]

Occasionally, neonates are born with severely compromised digits or limbs due to tight constriction bands. In such cases, urgent surgical release is necessary to restore perfusion and prevent permanent damage or loss of the limb or digit.

Deep bands that extend to or beyond the deep fascia may impair both vascular and neurologic function. Historically, a staged excision approach has been favored in such cases. In this method, only half of the band is excised initially to preserve skin viability, with the second half removed after a 6- to 12-week interval. This approach is believed to reduce the risk of necrosis due to impaired venous and lymphatic drainage. However, recent evidence suggests that single-stage circumferential release can often be performed safely, even for deep bands, without increased risk of complications. [1, 3, 17]

During surgical release, the constriction band and associated fibrous tissue are completely excised. This includes affected subcutaneous tissue, fascia, and occasionally muscle. A 1 to 2 mm margin of surrounding healthy tissue is also removed to ensure complete excision. [2, 18]

CONCLUSIONS

Isolated radial nerve palsy as a consequence of congenital CBS is a rare occurrence. Our reported case contributes to the limited literature by highlighting the effectiveness of early surgical decompression. Early circumferential excision of the constriction band and neurolysis of the radial nerve resulted in complete recovery of hand function without residual deficits.

PATIENT CONSENT

Written informed consent was obtained from the patient for the publication of this case report and all associated images.

AUTHORS' CONTRIBUTION

All authors have significantly contributed to the work, whether by following the case at the bedside, conducting literature searches, drafting, revising, or critically reviewing the article. They have given their final approval of the version to be published, have agreed with the journal to which the article has been submitted, and agree to be accountable for all aspects of the work.

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CONFLICT OF INTEREST

None.

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