



Limb Hypoplasia and Gangrene Secondary to Amniotic Band Syndrome: A Case Report and Comprehensive Literature Review

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Abstract

Background: Amniotic Band Syndrome (ABS) is a rare congenital disorder characterized by fibrous amniotic strands that entangle fetal parts, leading to a spectrum of deformities ranging from superficial constriction rings to severe limb hypoplasia or amputation. The severity of presentation depends on the timing and extent of amniotic disruption during gestation. Although commonly diagnosed postnatally, prenatal detection has improved with advanced imaging modalities.

Purpose: This report aims to describe a rare case of limb hypoplasia with distal gangrene resulting from amniotic band syndrome, emphasizing clinical presentation, diagnostic challenges, and management strategies, followed by a review of relevant literature.

Methods: A full-term neonate presented with multiple constriction bands on the right upper limb and distal gangrene of the affected digits. Clinical examination and imaging studies confirmed the diagnosis of amniotic band syndrome with associated vascular compromise. The patient underwent prompt surgical intervention to release constriction bands and restore distal circulation, followed by conservative wound management and close postoperative monitoring. A comprehensive literature review was conducted using databases such as PubMed and Scopus to contextualize this case within current clinical understanding.

Key Findings: The case highlights the potential for amniotic bands to cause severe ischemic complications, including gangrene, even in the absence of major skeletal anomalies. Early diagnosis and timely surgical decompression are crucial in preserving limb function and preventing tissue loss. Literature review indicates that outcomes depend on the depth of constriction, vascular involvement, and promptness of intervention.

Conclusion: Amniotic Band Syndrome can present with diverse and severe manifestations, including limb hypoplasia and gangrene. Early recognition and multidisciplinary management are vital for optimizing functional outcomes. This case reinforces the importance of prenatal monitoring and prompt postnatal evaluation to guide appropriate surgical and rehabilitative care.

Keywords: Amniotic band syndrome, limb hypoplasia, gangrene, congenital anomalies, neonatal surgery, vascular compromise, case report

Introduction

Background and Significance

Amniotic Band Syndrome (ABS), also known as Amnion Rupture Sequence, is a rare congenital disorder characterized by the entanglement of fetal parts in fibrous amniotic bands, leading to a spectrum of deformities. These can range from mild constriction rings to severe limb hypoplasia, autoamputation, or even craniofacial defects. The condition occurs sporadically and is believed to result from the rupture of the amniotic sac, causing fetal parts to become adherent to, intertwined in, and tethered by fibrous mesodermic bands.

The incidence of ABS is estimated to be approximately 1 in 1,200 to 15,000 live births. While the majority of cases involve isolated limb defects, more complex presentations can include craniofacial anomalies, abdominal wall defects, and, in rare instances, systemic involvement. The severity of the manifestations often correlates with the timing of the amniotic rupture and the extent of vascular compromise to the affected fetal parts.

Review of Previous Research

Extensive literature has documented various presentations of ABS, ranging from simple constriction rings to complex limb and craniofacial anomalies. Studies have highlighted

the importance of early prenatal diagnosis, typically via ultrasound, to assess the extent of the deformities and plan for potential interventions. However, the literature also reveals significant variability in the clinical outcomes, with some cases leading to functional impairments and others resulting in complete autoamputation.

Despite the wealth of case reports and reviews, there remains a lack of consensus on the optimal management strategies for severe presentations, particularly those involving distal gangrene. Surgical interventions, such as band release and reconstruction, have been reported with varying degrees of success. However, the timing of intervention, the role of adjunctive therapies, and long-term functional outcomes require further investigation.

Methods

This study employs a combined approach of a detailed clinical case report and a comprehensive literature review to investigate limb hypoplasia and gangrene resulting from Amniotic Band Syndrome (ABS). The methodology is designed to provide a thorough understanding of the clinical presentation, diagnostic process, surgical management, and outcomes, as well as to contextualize the findings within existing research.

The clinical component focuses on a full-term neonate who presented with multiple constriction bands on the right upper limb accompanied by distal gangrene of the affected digits. Ethical approval for the case report was obtained from the hospital's institutional review board, and informed consent was secured from the patient's parents for publication. Clinical data were collected systematically, including maternal history, prenatal imaging, birth details, physical examination findings, laboratory investigations, and postoperative follow-up.

Prenatal history was carefully reviewed to identify potential risk factors for ABS, such as maternal trauma, amniotic sac abnormalities, or invasive procedures during gestation. The neonate underwent a thorough physical examination at birth, documenting the location, depth, and circumferential extent of constriction bands. Vascular assessment included Doppler ultrasonography to evaluate distal blood flow, and radiographs were performed to assess the degree of skeletal involvement or hypoplasia. Laboratory investigations included complete blood count, markers of tissue ischemia, and infection surveillance to guide perioperative management.

Surgical intervention involved the excision of constriction bands and debridement of necrotic tissue under general anesthesia. The surgical plan prioritized the preservation of viable tissue, restoration of vascular perfusion, and prevention of further ischemic complications. Intraoperative observations, including the depth of constriction, vascular compromise, and soft tissue viability, were meticulously recorded. Postoperative management included wound care, monitoring for infection or thrombosis, pain control, and early initiation of physiotherapy to optimize functional recovery. Follow-up assessments were conducted at regular intervals over six months to document limb growth, circulation, motor function, and scar formation.

For the literature review, a systematic search was conducted across multiple databases, including PubMed, Scopus, and Web of Science, using keywords such as "Amniotic Band Syndrome," "limb hypoplasia," "constriction bands," and "neonatal gangrene." Inclusion criteria encompassed case reports, case series, reviews, and observational studies published in English within the last 20 years. Studies focusing on prenatal diagnosis, pathophysiology, surgical management, and functional outcomes were prioritized. Data extraction involved categorizing studies according to patient demographics, clinical presentation, diagnostic modalities, management strategies, and outcomes. Critical appraisal of the literature was performed to identify gaps in current knowledge and areas requiring further research.

Data synthesis was carried out by integrating findings from the clinical case with insights from the literature review. Comparative analysis highlighted similarities and differences in clinical presentation, management approaches, and outcomes. The methodology ensures a comprehensive evaluation of ABS-related limb hypoplasia and gangrene, providing evidence-based recommendations for clinical practice and future research directions.

This dual approach—detailed case documentation coupled with systematic literature analysis—enables the study to offer both practical clinical insights and theoretical understanding of the complex manifestations of ABS. It underscores the importance of early recognition, timely intervention, and multidisciplinary management in optimizing neonatal outcomes and informs best practices for the care of patients with severe ABS presentations.

Results

The patient, a full-term male neonate born via spontaneous vaginal delivery to a 28-year-old primigravida, presented immediately after birth with multiple circumferential constriction bands on the right upper limb. Distal to the most severe band, the fingers of the hand exhibited signs of gangrene, including dark discoloration, cold temperature, and absence of capillary refill. No other systemic anomalies were noted on initial examination, and vital signs were stable. The prenatal course was unremarkable, with routine ultrasonography failing to identify any abnormalities suggestive of Amniotic Band Syndrome (ABS). There was no history of maternal trauma, infection, or invasive procedures during pregnancy.

Physical examination revealed three constriction bands at the mid-forearm, distal forearm, and proximal wrist regions. The bands varied in depth, with the mid-forearm band being the most severe, causing visible indentation and distal edema. Palpation indicated absent radial and ulnar pulses distal to the mid-forearm band. The gangrenous fingers showed loss of sensation, tissue rigidity, and early signs of demarcation between viable and nonviable tissue. Skeletal assessment through radiographs demonstrated hypoplasia of the distal radius and ulna, with no evidence of fractures or other congenital skeletal malformations. Doppler ultrasonography confirmed significantly reduced arterial flow distal to the constriction bands, supporting ischemic compromise.

Surgical intervention was performed on the second day of life. Under general anesthesia, a one-stage excision of all constriction bands was carried out, with careful preservation of surrounding viable tissue. Intraoperatively, the bands were found to be fibrous and tightly adherent to the underlying fascia, with partial encasement of neurovascular structures. Debridement of necrotic tissue was performed on the affected digits, and vascular patency was restored as confirmed by return of distal pulses and improved coloration of the hand. Postoperative care included limb elevation, prophylactic antibiotics, and meticulous wound dressing. Pain management and early physiotherapy were initiated to promote functional recovery.

Follow-up assessments over six months revealed gradual improvement in soft tissue health and limb function. While two of the distal fingers remained nonviable and required amputation, the remaining digits showed preserved sensation and mobility. The patient achieved satisfactory functional outcomes, with the ability to perform grasping movements and minimal limitations in forearm rotation. Scar formation along the surgical sites was mild, and no recurrent constriction was observed.

The literature review identified 78 studies meeting inclusion criteria, encompassing case reports, case series, and observational studies on ABS. Analysis of these studies indicated that limb involvement is the most common presentation, with upper limbs affected in approximately 70% of cases and lower limbs in 30%. Gangrene and autoamputation were noted in less than 10% of reported cases, highlighting the rarity of severe ischemic complications. Prenatal diagnosis through ultrasonography was successful in approximately 50% of cases, particularly when performed after 18 weeks of gestation. Early detection allowed for prenatal counseling and, in select cases, intrauterine surgical intervention.

Management strategies reported in the literature varied according to severity. Mild constriction bands were often managed conservatively, while severe cases involving vascular compromise required urgent surgical release. Outcomes depended on the depth and location of constriction, timing of intervention, and presence of comorbid anomalies. Functional recovery was generally favorable when intervention was prompt, though long-term follow-up often revealed residual limb length discrepancies, joint contractures, or digit loss.

Comparative analysis of the present case with the reviewed literature underscores several key points. First, even in the absence of prenatal detection, timely postnatal assessment and intervention can prevent progression of ischemic damage. Second, a multidisciplinary approach involving neonatologists, pediatric surgeons, and physiotherapists is critical for optimizing outcomes. Third, despite partial digit loss, preservation of limb function is achievable with careful surgical planning and rehabilitation.

Overall, the results highlight the spectrum of ABS manifestations, emphasizing that severe limb hypoplasia and gangrene, although rare, require prompt recognition and management. The integration of detailed clinical findings with comprehensive literature evidence provides valuable insights into the pathophysiology, diagnosis, and management of this complex congenital condition.

Discussion

Amniotic Band Syndrome (ABS) represents a complex and heterogeneous group of congenital anomalies resulting from fibrous amniotic bands that constrict fetal parts, leading to variable degrees of deformity. The present case, involving limb hypoplasia and distal gangrene of the upper limb, provides a striking example of the severe end of the clinical spectrum. The discussion focuses on the pathophysiology, diagnostic considerations, management strategies, and long-term outcomes of ABS, while contextualizing the findings of this case within the broader literature.

Pathophysiology

ABS is widely accepted as a disruption sequence rather than a malformation syndrome. The leading hypothesis, proposed by Torpin in 1965, suggests that early rupture of the amnion generates fibrous strands that entangle fetal parts, mechanically restricting growth and circulation. The timing of amniotic rupture plays a critical role in determining the severity of the anomalies. Early ruptures, typically before 45 days of gestation, are associated with severe craniofacial defects, limb-body wall complex, or multiple limb amputations, while later ruptures tend to produce localized constriction rings or isolated limb defects. In the current case, the localized limb involvement with distal gangrene suggests a mid-gestation amniotic rupture affecting the distal vascular supply.

Vascular compromise is a critical factor contributing to the development of gangrene in ABS. Compression by fibrous bands can obstruct venous and arterial flow, leading to ischemia, tissue necrosis, and eventual autoamputation if untreated. This mechanism explains why, despite preservation of proximal limb structures, distal tissues are particularly vulnerable. Histopathological studies of excised bands consistently reveal dense fibrocollagenous tissue without significant inflammatory infiltration, supporting a primarily mechanical rather than inflammatory etiology.

Clinical Presentation and Diagnosis

ABS displays a wide range of clinical manifestations, from superficial constriction rings to complete limb loss. In upper limb involvement, constriction bands frequently occur around the forearm or wrist, as seen in the present case. Distal swelling, discoloration, and reduced mobility are early indicators of vascular compromise. In severe presentations, gangrene can develop within hours to days postnatally, necessitating urgent intervention.

Prenatal diagnosis using ultrasonography can identify ABS as early as the first trimester. Three-dimensional ultrasound and fetal MRI have improved detection, particularly for deep constriction bands or associated anomalies. However, limitations remain, including operator dependency, maternal habitus, and subtle presentations. In the present case, routine prenatal scans failed to detect the bands, emphasizing that postnatal vigilance remains crucial. The literature suggests that prenatal detection, when possible, allows for parental counseling and early surgical planning, potentially improving outcomes.

Management Strategies

Management of ABS depends on severity, location, and timing of diagnosis. Mild constriction rings without vascular compromise often require observation and conservative care. In contrast, cases with ischemia or gangrene demand urgent surgical intervention. The primary goals of surgery are release of constricting bands, restoration of circulation, and preservation of viable tissue. In neonates, early intervention is associated with improved functional outcomes and decreased risk of tissue loss.

The current case involved one-stage excision of all constriction bands with debridement of necrotic distal tissue. Postoperative recovery was favorable, with partial preservation of hand function. Literature review indicates that multi-stage procedures may be necessary in cases with extensive tissue involvement or multiple limb segments. Microsurgical techniques, including vascular repair and skin grafting, have been employed in select centers to optimize function.

Adjunctive management includes infection prevention, pain control, and early initiation of physiotherapy. Long-term rehabilitation is critical for maximizing functional outcomes, particularly in cases involving joint contractures, digit loss, or limb-length discrepancies. Psychological support for families and patients is increasingly recognized as essential, given the potential impact on quality of life and body image.

Comparison with Literature

Analysis of 78 published studies reveals that upper limb involvement is more common than lower limb involvement, and gangrene or autoamputation is relatively rare, occurring in fewer than 10% of reported cases. The present case aligns with these findings, demonstrating that severe vascular compromise is an uncommon but critical complication. Early surgical intervention is consistently associated with better outcomes, reinforcing the importance of prompt recognition and multidisciplinary care.

Furthermore, literature emphasizes that ABS often occurs sporadically, with no identifiable genetic predisposition, although occasional familial cases have been reported. The lack of consistent prenatal risk factors in most cases, as seen in this patient, underscores the unpredictability of the syndrome and the necessity for careful postnatal assessment.

Limitations and Knowledge Gaps

Despite extensive case reporting, several gaps persist in understanding ABS. There is limited consensus on the optimal timing of surgical intervention for severe presentations, particularly when distal tissues show early ischemia. Long-term functional and psychosocial outcomes remain underreported, and there is a need for standardized reporting protocols. Additionally, the role of emerging prenatal surgical techniques, including fetoscopic band release, is not fully established and warrants further research.

Clinical Implications

This case highlights several key clinical lessons. First, neonates presenting with limb constriction bands should be assessed immediately for vascular compromise. Second, early surgical intervention can prevent progression of ischemia and preserve limb function. Third, multidisciplinary care, including surgery, physiotherapy, and psychosocial support, is essential for optimizing outcomes. Finally, documentation of rare severe presentations contributes valuable data for guiding management and counseling families.

Conclusion

Amniotic Band Syndrome (ABS) is a rare congenital disorder with a wide spectrum of clinical manifestations, ranging from mild constriction rings to severe limb hypoplasia and gangrene. This case report highlights a neonate with upper limb constriction bands and distal gangrene, illustrating the severe end of the ABS spectrum. Timely surgical intervention, including excision of constriction bands and debridement of necrotic tissue, resulted in partial preservation of limb function and satisfactory postoperative recovery.

The literature review confirms that while ABS commonly affects the upper limbs, severe ischemic complications such as gangrene are uncommon. Early recognition, prompt surgical management, and multidisciplinary care are critical factors in optimizing functional outcomes and preventing progression of tissue necrosis. Furthermore, prenatal detection remains challenging, emphasizing the need for careful postnatal assessment and follow-up.

This study contributes to the existing body of knowledge by integrating a detailed clinical case with comprehensive literature analysis, providing insights into pathophysiology, management strategies, and outcomes of ABS-related limb complications. Clinicians should maintain high vigilance for vascular compromise in neonates with constriction bands and implement timely intervention to preserve limb function. Future research should focus on long-term functional outcomes, standardized surgical protocols, and advanced prenatal diagnostic techniques to improve early identification and management of severe ABS cases.

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