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Sindrome delle briglie amniotiche: caso clinico con interessamento dei quattro arti

Amniotic Band Syndrome: case report with four limbs involvement

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Riassunto

Introduzione. La sindrome delle briglie amniotiche è una malattia congenita rara caratterizzata dalla presenza di solchi o anelli di costrizione che causano deformità negli arti dei neonati. Presentiamo un caso con grave coinvolgimento di tutti gli arti.

Materiali e metodi. Dopo un primo intervento eseguito 24 ore dopo la nascita per risolvere il grave linfedema del piede destro, il paziente è stato sottoposto nell'arco dei 2 anni successivi a diversi interventi per acrosindattilia delle mani.

Risultati. Il paziente ha sviluppato un'ottima capacità di manipolazione degli oggetti ed, eccetto una lieve ipometria destra, ha avuto un normale sviluppo degli arti inferiori e dello schema motorio del passo.

Conclusioni. Le manifestazioni della sindrome delle briglie amniotiche sono molto variabili e lo stesso paziente può presentare deformità di diverso tipo. Richiede pertanto un trattamento personalizzato, spesso a più step chirurgici e volto in prima istanza al ripristino della funzionalità.

Parole chiave: briglie amniotiche, solchi amniotici, malformazione congenita, acrosindattilia

Summary

Introduction. *Amniotic band syndrome is a rare congenital disorder characterized by constriction rings that cause deformities in the limbs of newborns. We present a case with severe involvement of all limbs.*

Materials and methods. *After a first surgery performed 24 hours after birth to resolve the severe lymphedema of the right foot, the patient underwent several surgical procedures over the next 2 years for acrosyndactyly of the hands.*

Results. *The patient developed an excellent ability to manipulate objects and, except for a slight right hypometry, he had a normal development of the lower limbs and of gait pattern.*

Conclusions. *The manifestations of amniotic band syndrome are enormously variable and the same patient can present different types of deformities. It therefore requires a personalized treatment, often involving multiple surgical steps and aimed primarily at functional recover.*

Key words: *amniotic bands, amniotic rings, congenital deformity, acrosyndactyly*

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Conflitto di interessi
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Introduction

Amniotic band syndrome is a congenital disorder characterized by constriction rings that cause deformities or amputations in newborns. The main cause of such deformities is the early rupture of the amniotic sac during the early stages of pregnancy, leading to the formation of bands that trap the fetus. The severity and clinical variability of the syndrome can affect any part of the body, but the extremities are particularly at risk. The most common deformities in the upper limbs include distal constriction rings, lymphedema, intrauterine amputations, and acrosyndactyly. These deformities can lead to significant disabilities in the patient, requiring personalized treatment that often occurs in several stages during childhood. In this article, we present a severe case of amniotic band syndrome with involvement of four limbs[1].

Case report

A male infant of African American origins was born at a gestational age of 34 weeks. Immediately after birth a severe form of amniotic band syndrome was diagnosed with involvement of all four limbs as follows:

- Acrosyndactyly of the five rays of the right upper limb; all fingers except the fourth presented only the proximal phalanx (Fig. 1)

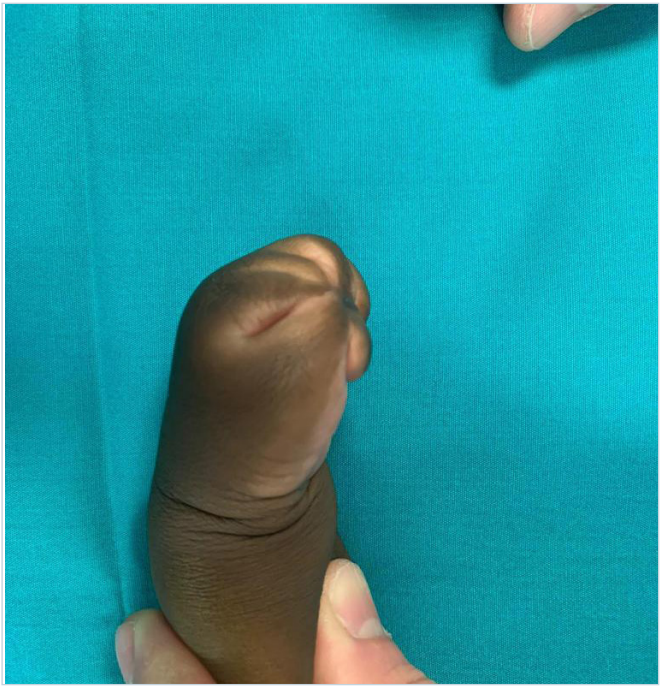

Figure 1. Acrosyndactyly of the five rays of the right upper limb at birth.

Figure 2. A deep circumferential amniotic band at the distal third of the leg associated with severe lymphedema and equinovarus foot of the right lower limb; Syndactyly and amputation of all toes of the left foot.

- Acrosyndactyly of the first four rays of the left upper limb; all fingers except the fifth presented only the proximal phalanx
- A deep circumferential amniotic band at the distal third of the leg associated with severe lymphedema and equinovarus foot of the right lower limb; (Fig. 2)
- Syndactyly and amputation of all toes of the left foot. (Fig. 2)

At birth, the most concerning aspect was the constriction ring to the right leg associated with a severe lymphedema, type II according to Patterson classification. The same foot presented also a clubfoot deformity. The state of soft tissue was so troublesome that on the first day of life, it was decided to proceed with the removal of the amniotic band at the level of the right leg using the Upton technique in one stage[2]. In the following months, the lymphedema gradually reduced, but the excess of skin and soft tissue prevented the application of cast according to Ponseti method. At 10 months, debulking of the right foot was performed using a multiple Z-plasty technique. At 11 months, after the removal of the sutures from the right leg, correction of clubfoot deformity began using the Ponseti technique. As for the acrosyndactyly of the hands, at 7 months, the opening of the first web space of the left hand was performed using the Jumping Man flap, along with the opening of the first, third

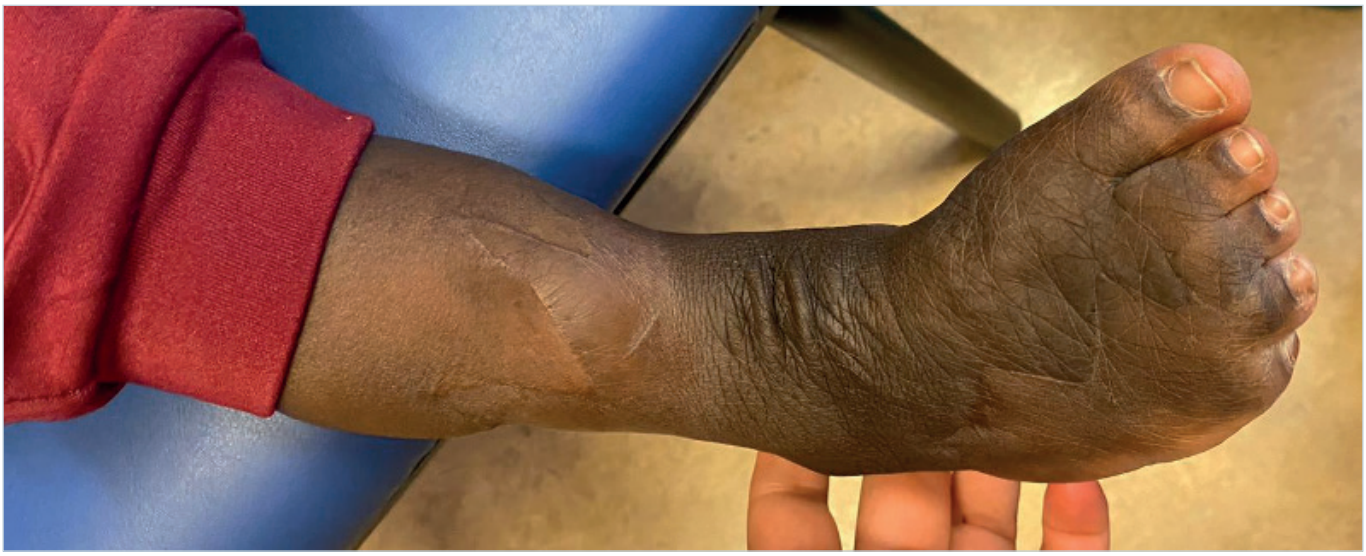

Figure 3. The right lower limb after treatment.

and fourth web spaces and the separation of the apical bone bridge between the second and the third fingers of the right hand. At 12 months, Achilles tenotomy was performed on the right side, and simultaneously, the II web space of the left hand was opened using a dorsal and volar triangular flap and a full thickness graft from the volar region of the wrist. At 15 months, the initiation of walking and good gripping and mobilization of objects were achieved (Fig. 3)

Discussion

Amniotic Band Syndrome (ABS) is a rare and still poorly understood congenital pathology. Many different etiological theories have been proposed over time, and the most famous are Streeter's intrinsic theory and Torpin's extrinsic theories[3]. The first one suggested that constriction rings were due to defects in germ plasma, while the second hypothesized that the cause was external compression. Anyway, many authors have extensively studied the clinical manifestation of this syndrome, which can enormously vary from patient to patient, and even in the same patient we can observe different types of deformities. ABS is characterized by the presence of fibrous bands which may entangle fetus limbs in utero causing different types of deformities[4]. The constriction rings generally involve more segments, and they can be of different deepness. Sometimes, they are very superficial resulting in a little dimple with any functional problem. Instead, deeper bands can lead to amputation, vascular compromise and severe lymphedema, nerve palsies and secondary deformities such as acrosyndactyly. In this article, we have reported a severe case of ABS with involvement of all limbs. We found many points of reflection in this case. First, regarding the type of surgery for deep constriction ring, it is debated if the best procedure is a single-stage or two stage technique. Two stage surgery was initially recommended for treatment of circumferential constriction bands whereby the first half of the band is excised, followed by the other half of the band later. This is done with the aim to prevent disruption of limb circulation distal to the band[5]. However, since the circulation of the limb distal to the constriction band is supplied by the myocutaneous arteries and endosteal blood supply, single-stage surgery is possible, as demonstrated by many Authors[6-7]. We opted for a single stage procedure with multiple z-plasties, with a satisfactory outcome in terms of wound healing, scar quality, and limb function. Another important aspect was the coexistence of lymphedema and clubfoot. Contrary to other cases reported in literature, in our case, the excess of skin and soft tissue after ring correction and edema reduction prevented the early application of corrective casts according to Ponseti method[8]. Casting was possible only after a second surgery of debulking of the soft tissue of the foot especially on the dorsal aspect. Despite the delay of the treatment, we were able to obtain a satisfactory foot correction and at present the child has experienced no relapse. When ABS affected hands, the result is often acrosyndactyly which is a constriction ring around adjacent digits, with distal soft-tissue webbing and epithelial sinus tracts at the base of the proximal phalanx. This is a very challenging deformity for the hand surgeon. According to Walsh, we can distinguish three types of acrosyndactyly: mild acrosyndacyly with three phalanges and two interphalangeal joints in the affected dig-

its, typically with long clefts between the fingers; moderate acrosyndactyly with two phalanges and one interphalangeal joint; severe acrosyndactyly presents only stubby fingers, with one phalanx and no interphalangeal joints[9]. Our case is an example of severe acrosyndactyly, as all hand fingers, except two, presented only one phalanx. The surgical goal in these cases is early separation combined with commissural deepening. Multiple surgical steps are often necessary to obtain a functional hand, because fingers could be fused and distorted and sometimes It may be difficult to determine which fingertip goes to which finger. We can conclude that treatment of ABS must be often individualized, the intervention should start as early as possible in the first months of life especially in case of severe deformity of the hands or severe lymphedema. Parents should be informed about the need for multiple corrective surgeries and that limb function takes precedence over aesthetic outcome.

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