Amniotic Band Syndrome with the Involvement of Trunk: A Case Report

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Abstract

Amniotic Band Syndrome is a sporadic congenital disorder that may result in constriction bands, amputation and multiple craniofacial, visceral and body wall defects. Its incidence is estimated at approximately 1:5000 to 1:10,000 pregnancies. Band formation most frequently affects the distal segments, including the hand. We report a case of constriction amniotic bands involving the trunk. *(Iran J Dermatol 2010;13: 103-104)*

Key words: amniotic band syndrome, trunk , congenital, scar

Case Report

A 20-year-old man was visited at our clinic with congenital scar tissues on the trunk without the involvement of other organs or elsewhere. He was the result of a mature and normal delivery. There were no other problems in his past history. Mental and motor developments were normal. In examination, there were multiple spiral strands of atrophic scar tissues on the trunk. Limbs and head and neck were normal (Figure1). Family history for the same disorder was negative. Hair, nail and mucous membranes were normal. All these data was compatible with the diagnosis of amniotic band syndrome.

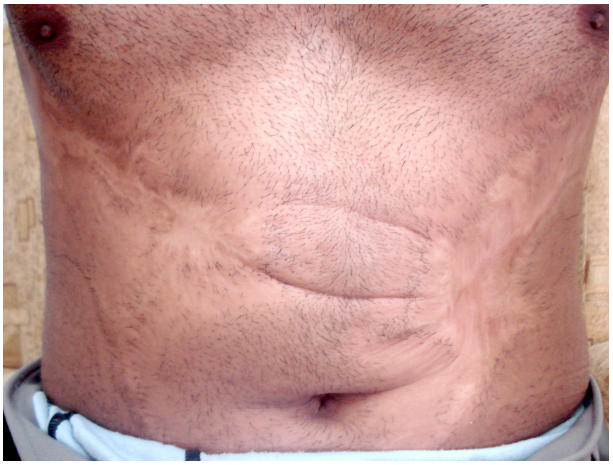

Figure1. *Multiple spiral strands of atrophic scar tissues on the trunk since birth*

Discussion

Amniotic fibrous bands may attach to various parts of the fetus or newborn and cause a variety of congenital malformations. The amniotic (constriction) band syndrome is characterized by distal ring constrictions, intrauterine amputations, and acrosyndactyly. Anecdotal cases involving central nervous system abnormalities (acrania, anencephaly, polymicrogyria, congenital bilateral perisylvian syndrome, neuronal heterotopia, septo-optic dysplasia, and spinal cord tethering) have been reported [1].

Two main theories have been proposed regarding the pathogenesis of amniotic bands: 1) the amniotic bands are the result of developmental defects occurring at the time of the formation of the germ disk and the amniotic cavity (endogenic origin), and 2) these bands generate from early rupture of the amnion (exogenic origin). Careful examination of the placenta is essential for a precise definition of the pathogenesis [2].

The variability in defect and severity has been attributed to differences in timing of the amniotic rupture [2]. Early rupture, within the first 45 days of gestation, leads to the most severe defects, including central nervous system and skull defects, facial clefts, cleft lip and palate, limb anomalies,

and major visceral defects. Early compression may also result in incomplete separation of the digits (syndactyly) or extra finger rays (polydactyly). When an amniotic rupture occurs after 12 weeks of gestation, constriction of isolated limb parts may occur more frequently than central nervous anomalies or clefts [3].

The exact cause of the constriction band in our patient was unknown. The concept of the amniotic band disruption complex suggests that amniotic rupture may have occurred late in the gestational stage.

Differential diagnoses include 1) Michelin tyre baby syndrome in which there is abnormal limb enlargement with folds rather than constriction 2) fetal varicella syndrome which is the result of maternal infection between 13 and 20 weeks of gestation. Fetal involvement with varicella (the risk is approximately 2%) usually presents with localized absence of skin, usually on limbs, due to intrauterine ulceration, dermatomal scar, papular lesions resembling connective tissue nevus, limb or digital hypoplasia associated with ocular and CNS abnormalities [4].

Because of the heterogeneous presentations of this disease, treatment is individualized. Timing of repair and surgical planning are important in improving its functional outcome. The most accepted method of treatment for constriction bands is excision of the ring and staged Z-plasty with fairly large flaps [5-8]. In addition, excision of the ring and a single-stage multiple Z-plasty repair has been described [7,9]. These techniques have been described for constriction band defects in the more common locations of the upper or lower limbs, trunks, or digits. Because of the unusual location of the constriction band defect, a simple excision of the band with multiple Z-plasties could not completely correct the deformity in our patient.

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