

Emergency approach in a rare congenital coexistence-ichthyosis and amniotic band syndrome

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ABSTRACT

Ichthyosis is caused by Mendelian cornification disorders. Hereditary ichthyoses are divided into non-syndromic and syndromic ichthyoses. Amniotic band syndrome involves congenital anomalies that most frequently cause hand and leg rings. The bands can wrap around the developing body parts. In this study, it is aimed to present an emergency approach to amniotic band syndrome accompanying a case of congenital ichthyosis. We were asked by the neonatal intensive care unit to consult on the case of a 1-day-old baby boy. On physical examination, congenital bands were found to be present on both hands, the toes were rudimentary, skin scaling was present on the entire body, and the consistency of the skin was stiff. The right testicle was not in the scrotum. Other system examinations were normal. However, the blood circulation in the fingers in the distal of the band had become critical. With the help of sedation, the bands on the fingers were excised, and after the procedure, it was observed that the circulation in the fingers was more relaxed than it had been before the procedure. Coexistence of congenital ichthyosis and amniotic band is very rare. Emergency approach to these patients is very important in terms of saving the limb and preventing growth retardation in the limb. As further developments take place in terms of prenatal diagnoses, these cases will be able to be prevented through the early diagnosis and treatment.

Keywords: Amniotic; band; ichthyosis.

INTRODUCTION

Ichthyosis is caused by Mendelian cornification disorders. Hereditary ichthyoses are divided into non-syndromic ichthyoses and syndromic ichthyoses. Non-syndromic ichthyoses usually include only skin lesions. However, syndromic ichthyoses have phenotypes that vary, depending on the genetic defects, and may not include only skin involvement but that of other organs as well. Syndrome forms such as Netherton, Sjögren Larsson, Refsum, Multiple Sulfatase Deficiency, Keratitis-Ichthyosis-Deafness, Congenital Hemidysplasia, Conradi-Hunermann-Happle, Dorfman-Chanarin Syndrome, and Hystrix-Like Ichthyosis with Deafness have been described in

the literature. However, none of these findings are associated with amniotic band syndrome.^[1]

Amniotic band syndrome involves congenital anomalies that most frequently cause hand and leg rings. The bands can wrap around the developing body parts partially or completely. This can result in atrophy in soft tissues and rudimentation in distal tissues. Its incidence is approximately 1/15,000. Gender and race differences are not observed, mostly seen sporadically, rarely familial cases have been reported.^[2,3]

In this study, it is aimed to present an emergency approach to amniotic band syndrome accompanying a case of congenital ichthyosis.

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CASE REPORT

We were asked by the neonatal intensive care unit to consult on the case of a 1-day-old baby boy who was born by spontaneous vaginal delivery. His mother was 21 years of age, and this was her fifth pregnancy. On physical examination, congenital bands were found to be present on both hands, the toes were rudimentary, skin scaling was present on the entire body, and the consistency of the skin was stiff. The right testicle was not in the scrotum. Other system examinations were normal. However, the blood circulation in the fingers in the distal of the band had become critical (Fig. 1-3).

There was kinship between the parents, but no disease in the mother, and she had no history of drug use during pregnancy. With the help of sedation, the bands on the fingers were excised, and after the procedure, it was observed that the circulation in the fingers was more relaxed than it had been before the procedure (Fig. 4 and 5).



Figure 1. Views of the amniotic bands in the right hand



Figure 2. Views of the amniotic bands in the left hand



Figure 3. Views of the rudimentary toes

We used vaseline and lanolin together in the treatment of our patient. We did not detect any problems in follow-up.

DISCUSSION

The pathogenesis of amniotic band syndrome is still unclear. The widely accepted theory is that amniogenic fibrous bands are the cause of the malformations that occur. The malformations are thought to be due to the intervention of the bands during embryonal development. If the amniotic bands occur before the 45th day of pregnancy, this may result in various cranial defects, facial clefts, and large organ defects; if the bands occur after the 45th day, limb constrictions, amputations, and pseudo-syndactyls may be seen.^[4] Our patient had only limb anomalies. In this patient, prenatal follow-up was not regular. We think that the amniotic bands in our patient developed in the last period of the pregnancy because they did not cause serious rudimentation in the extremities.

The term ichthyosis is used to describe a group of hereditary keratinization disorders of the skin, and it is primarily a single



Figure 4. View of the right hand after release



Figure 5. View of the left hand after release

gene disease. Prenatal diagnosis is made by fetoscopy, fetal skin biopsy, and microscopic examination of the amniotic fluid.^[5] Since prenatal follow-up was not regular in this patient, the diagnosis of ichthyosis was made clinically. Congenital ichthyosis includes various clinical phenotypes such as classical harlequin ichthyosis, non-bullous congenital ichthyosiform erythroderma, and classical lamellar ichthyosis. In some cases of ichthyosis, there may be extra-cutaneous involvements such as the central nervous system, immune system, and skeletal system, and undescended testicle may be detected.^[6] In our patient, the right testicle was not palpable.

The main purpose of the treatment was to prevent dehydration, moisturize the skin, and produce the softness of the stratum corneum. Although there is controversy in the literature, with some studies indicating that topical moisturizers may pose a risk for infection, the use of soft moisturizers such as petroleum jelly while paying attention to hygiene practices is currently the accepted treatment approach. Topical moisturizers should be applied as soon as the baby's movements are blocked by the surrounding membrane and are causing difficulties in breathing and with movements. Early intervention is very important in this rare disease, and proper care of the wound is vital.^[7]

Conclusion

Coexistence of congenital ichthyosis and amniotic band is very rare. Emergency approach to these patients is very important in terms of saving the limb and preventing growth retardation in the limb. As further developments take place in terms of prenatal diagnoses, we believe that these cases will be able to be prevented through the early diagnosis and treatment. In addition, multidisciplinary approaches play an

important role in the treatment and post-treatment care of these patients.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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OLGU SUNUMU - ÖZ

Nadir bir konjenital birlikteliğe acil yaklaşım – iktiyozis ve amniyotik bant sendromu

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İktiyozis, Mendel kornifikasyon bozukluklarından kaynaklanır. Kalıtsal iktiyozlar, sendromik olmayan ve sendromik iktiyozlar olarak ikiye ayrılır. Amniyotik bant sendromu, en sık el ve bacak halkalarına neden olan doğumsal anomalileri içerir. Bantlar gelişmekte olan vücut kısımlarını sarabilir. Bu çalışmada, konjenital iktiyozis olgusuna eşlik eden amniyotik bant sendromuna acil yaklaşımın sunulması amaçlandı. Yenidoğan yoğun bakım ünitesi tarafından bir günlük erkek bebek vakası için konsültasyon istendi. Fizik muayenede her iki elde doğuştan bantlar mevcuttu, ayak parmakları rudimenterdi, tüm vücutta ciltte pullanma ve cilt sertliği mevcuttu. Sağ testis skrotumda değildi. Diğer sistem muayeneleri normaldi. Ancak bantın distalindeki parmaklardaki kan dolaşımı kritik hale gelmişti. Sedasyon yardımı ile parmaklardaki bantlar eksizye edildi ve işlem sonrasında parmaklardaki dolaşımın işlem öncesine göre daha rahat olduğu gözlemlendi. Konjenital iktiyozis ve amniyotik bant birlikteliği oldukça nadirdir. Bu hastalara acil yaklaşım, ekstremitenin kurtarılması ve ekstremitede büyüme geriliğinin önlenmesi açısından çok önemlidir. Prenatal tanılar açısından gelişmeler yaşandıkça erken tanı ve tedavi ile bu vakaların önüne geçilebilecektir.

Anahtar sözcükler: Amniyotik; bant; iktiyozis.

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