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# Konjenital İktiyozisli Preterm İnfantta Ağır Seyreden Kandida Sepsisi

# Severe Candida Sepsis in a Preterm Infant with Congenital Ichthyosis

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## ÖZ

İktiyozlar klinik olarak heterojen olup, derinin kalıtsal keratinizasyon bozukluğudur. Klinik, yaygın cilt kuruluğundan hiperkeratoza kadar oldukça değişkendir. Dehidratasyon, sepsis, elektrolit dengesizliği gibi komplikasyonların önlenmesi sağ kalım açısından büyük önem taşırken, prematür yenidoğanlarda komplikasyonlar daha ağır seyredebilir. Bu çalışmada, konjenital iktiyozisli prematür bebekte gelişen dirençli candida sepsisini **sunduk.**

**Anahtar kelimeler:** preterm, konjenital, iktiyozis, kandida, sepsis

## ABSTRACT

Ichthyosis which is an inherited keratinisation disorder of the skin, is heterogeneous as clinically. This disease's clinic is considerably variable from widespread skin dryness to hyperkeratosis. While the prevention of complications such as dehydration, sepsis and electrolyte imbalance, is very important in respect to survival, in premature neonates, more severe complications may be seen. In this case, we reported that a premature infant who developed catastrophic candida sepsis with congenital ichthyosis.

**Keywords:** preterm, congenital, ichthyosis, candida, sepsis

## INTRODUCTION

Ichthyosis means "fish skin".Ichthyosis which is seen with dryness, flaking and peeling in the skin, is an inherited disorder (1, 2). Ichthyosis Vulgaris and X-linked ichthyosis are the most commonly seen types and congenital ichthyosis is seen less often. Infants were born with a clear membrane (collodion membrane) which is similar in structure to parchment paper covering the whole body of the infant (3). This membrane does not function like normal skin, hyperkeratosis is seen in the pathophysiology and it tears spontaneously within the first 1-2 weeks. Although ichthyosis itself is not life-threatening, the emergence of complications such as dehydration, infections and electrolyte imbalance cause severe morbidity and mortality. In preterm infants, the incidence of complication is increased and complications have a more fatal course (4, 5). In the Neonatal Intensive Care Unit (NICU), candida infections are the main reason for fungal sepsis and related to morbidity and mortality (6).

In this case, we reported catastrophic candida sepsis in a premature infant diagnosed with ichthyosis.

## CASE

A premature, male infant, delivered by the normal vaginal route at 28 weeks with 1500 gr birthweight was admitted to our institution because of respiratory distress syndrome and the diagnosis of ichthyosis from an external centre. The infant was the 4th pregnancy and 3rd live birth of a 22-year old healthy mother. There was no consanguinity between the parents, the first child was 3 years old and healthy, and the other child had been born prematurely and had died at 5 days postnatal. In the patient's first physical examination, there were oedema of the eyelids, evident bilateral ectropion and crusted, fish-scale type lesions over all the scalp. The skin which's appearance is hyperemic and shiny covered the whole body as a membrane, and there were ecchymotic lesions on both feet (Figure).

Appropriate fluid and ventilation support was started to the patient. The patient was consulted to the Dermatology Department and treatment was started that contains vaseline care for the trunk and castor oil, salicylic acid and olive oil applied to the hair twice a day (10-day treatment). At the same time, umbilical catheter was placed and ampicillin was started with gentamicin for empirical treatment. On the

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8th day of follow-up, sepsis was clinically determined with WBC increased from 7.67 109/L to 12.02 109/L, thrombocytes decreased from 120 109/L to 39 109/L and negative CRP increased to 34 mg/L and procalcitonin was positive at 2.8 ng/L. Blood culture was taken. Treatment was continued with vancomycin and meropenem. At the same time, nystatin was started as prophylactic because of extended-spectrum antibiotic treatment. The acute phases did not recover and the patient had a fever, amphotericin B was started for the fungal infection on the 10th day of follow up. The patient continued to deteriorate clinically. In the blood culture, candida albicans was determined on the 12th day of life. This agent was sensitive to caspofungin. caspofungin was added to the treatment but despite all the supportive treatment, the patient died on 14th day of life.

## DISCUSSION

For the diagnosis of ichthyosis, also known as collodion infant, physical examination and skin findings are sufficient. The cheeks have a red, tight and shiny appearance, and palms of the hands and soles of the feet are usually involved. In the diagnosis of the disease, the presence of ectropion is an important finding also eclabion, hypohidrosis and alopecia are accompanying other findings. Unlike congenital ichthyosiform erythroderma, in this disease, erythroderma is either not seen at all or is seen very mild. The autosomal recessive inheritance pattern suggesting a sibling history and parental consanguinity is also important in diagnosis (3, 7). In the current case, there was no positive sibling history and no parental consanguinity. As the infant was premature, the subcutaneous fat tissue was less than normal and the skin was clear, therefore the patient had an erythrodermic appearance.

In ichthyosis patients, heat and fluid loss develops from the skin because of the integrity of the epidermis is impaired. Consequently, hypernatremic dehydration often occurs. The risk of mortality is increased associated with pyoderma and sepsis (5). The prevention of occurence of complications such as dehydration, sepsis, electrolyte imbalance and pneumonia is very important in respect to survival.

Topical agents are often used in treatment. In some studies at the experimental stage it has also been shown that systemic drugs could be used (5, 8).

In premature infants with very low birthweight, such as the current patient, complications such as these are expected to be observed at a more severe degree than in full-term infants with ichthyosis. In the current patient, hypernatremic dehydration and electrolyte imbalance were observed, then candida sepsis developed, the patient did not respond to treatment and was lost. In NICUs, the occurrence of invasive candida infection is one of the most significant reason for morbidity and mortality (6). Candida albicans fungemia can be treated with amphotericin B and its liposomal forms without 5-Flucitosin or fluconazole (9). Despite the initiation of amphotericin B treatment to the current patient, there was no clinical recovery.

Caspofungin belongs to the new antifungal agent group of echinocandins and has fungicidal activity against candida strains. There are few current examples of the use of caspofungin in neonates. In one previous study, the combination of amphotericin B and caspofungin was shown to be beneficial in a premature infant with ichthyosis (10). However, there has been no study yet that has tested combined treatment in comparison to monotherapy in preterm infants with high risk faktors such as ichthyosis.

Based on current knowledge, although antifungal monotherapy, especially amphotericin B, is the first choice in neonates with invasive candida, as combined therapy with caspofungin it could also be an option for patients with invasive candidiasis that does not respond to antifungal monotherapy (10). In the case reported here, treatment was started with amphotericin B as monotherapy, but the patient did not respond to treatment and it was exitus.

In the follow-up of such cases, it must be taken into consideration that catastrophic invasive fungal infection could develop. In premature infants with additional risk factors thought to have invasive fungal infection, combined therapy with caspofungin should be considered at the forefront of treatment. There is a need for further studies to determine the optimal antifungal therapy for neonates with systemic candidiasis that does not respond to antifungal monotherapy.

## REFERENCES

1. Oji V, Tadini G, Akiyama M, et al. Revised nomenclature and classification of inherited ichthyoses: results of the First Ichthyosis Consensus Conference in Sorèze 2009. J Am Acad Dermatol 2010; 63(4): 607-41.

2. Schmuth M, Gruber R, Elias PM, et al. Ichthyosis update: towards a function-driven model of pathogenesis of the disorders of cornification and the role of corneocyte proteins in these disorders. Adv Dermatol 2007; 23: 231-56.

3.Fischer J: Autosomal recessive congenital ichtiyosis. J Invest Dermatol 2009; 129 (6): 1319-21.

4 . DiGiovanna JJ, Robinson-Bostom L. Ichthyosis: etiology, diagnosis, and management. Am J Clin Dermatol 2003;4 (2): 81-95.

5. Dyer JA, Spraker M, Williams M. Care of the newborn with ichthyosis. Dermatol Ther 2013; 26 (1):1-15.

6. Leibovitz E. Neonatal candidosis:clinical Picture, management controversies and consensus, and new therapeutic options. J Antimicrob Chemother 2002; 49: 69-73.

7. Shwayder T, Ott F. All about ichthyosis. Pediatr Clin N Am 1991;38 (4) :835-57.

8. Van Gysel D, Lijnen RL, Moekti SS, et al. Collodion baby: a follow-up study of 17 cases. J Eur Acad Dermatol Venereol 2002;16 (5): 472–5.

9.Odio CM, Araya R, Pinto LE et al. Caspofungin therapy of neonates with invasive candidiasis.Pediatr İnfect Dis J 2004; 23 (12):1093-7.

10. Haase R, Kreft B, Foell J, Kekulé AS, Merkel N. Successful treatment of Candida albicans septicemia in a preterm infant with severe congenital ichthyosis (Harlequin baby). Pediatr Dermatol. 2009; 26 (5): 575-8.