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CASE REPORT

Vibrio Cholerae Septicemia and Harlequin Ichthyosis: Outstanding Association

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Abstract:

Background:

Harlequin ichthyosis is a rare and fatal fetal disorder, most often complicated by infection. Sepsis due to *Vibrio cholerae* is rare, especially in neonatology. The association between ichthyosis and *Vibrio cholerae* infection has never been described in medical literature.

Introduction:

We report the case of a newborn admitted to the neonatal intensive care unit, with a typical characteristics of Harlequin ichthyosis and a sepsis due to *Vibrio cholerae*, diagnosed with the blood culture. The outcome was fatal; characterized by death six hours after his hospitalization.

Conclusion:

Cases of Harlequin ichthyosis complicated with septicemia with *Pseudomonas Aeruginosa* have been described. *Vibrio cholerae* infections are usually confined to the gastrointestinal tract but some species, especially non-01 strains, can cause extra-intestinal infections including septicemia and meningitis.

Keywords: Harlequin ichthyosis, Infection, Newborn, Septicemia, *Vibrio cholerae*, *Pseudomonas aeruginosa*.

1. INTRODUCTION

The term ichthyosis derives from the Greek word “ichthus” which means “fish”. It includes a heterogeneous set of congenital or acquired pathologies characterized by xerosis cutis and the presence of excessive desquamation, reminiscent of fish scales. These pathologies are distinguished by their mode of transmission, age of onset, clinical presentation and the causal genetic anomaly [1]. Harlequin ichthyosis is a rare and fatal fetal disorder: the most serious autosomal recessive congenital ichthyosis and is most often complicated by infection.

Sepsis due to *Vibrio cholerae* is rare, particularly in neonatology, although sporadic cases have been reported in older patients [2]. The association between ichthyosis and *Vibrio cholerae* infection has never been described in literature.

We report an exceptional case, never before reported in medical literature, of *Vibrio cholerae* sepsis in a neonate associated with Harlequin ichthyosis, admitted to the neonatal intensive care unit.

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2. CASE REPORT

A newborn, female, from a cousin marriage, admitted on her second day of life for prematurity with neonatal dermatosis. The parturient aged 17, she was primiparous. The pregnancy was closely monitored and estimated at 31.9 weeks of gestation. There was no peculiar medical history, especially, there is not a diarrhea or a recent digestive disorders. The anamnesis was suggestive of infection, the delivery was vaginal, the Apgar was unprecise and the cry was immediate.

Upon admission the newborn was conscious, hypotrophic (-10P) with hypothermia, bradycardia, capillary recoloration time was prolonged. The skin was very hypertonic with large hyperkeratotic plaques separated by deep cracks, the hyperkeratosis of the ears, the ectropion and eclabium, the purulent fetid lesions, with a facial dysmorphism and an arthrogyposis (Fig. 1). The primitive reflexes were difficult to appreciate and the sucking reflex was absent.



Fig. (1). General appearance of congenital ichthyosis.

After initial measures, the newborn was treated by triple antibiotic therapy (in the first the antibiotics was probabilistic for all neonatal infection in our unit; we used 3rd generation Cephalosporin and Aminoglycoside (gentamicin); in addition, in this case Metronidazole because the newborn had a fetid lesions) and topical Vaseline for local application.

The Blood chemistry revealed a renal insufficiency at 0.74g/l, the lab tests for infections revealed neutropenia at 370/mm³, a C-reactive protein was 122mg/l, the blood culture recovered the following day had isolated *Vibrio cholerae*; this bacteria was sensible to this antibiotics: Cephalosporin, Fluoroquinolones, Sulfamides, Colistin Imipenem, Aminoglycosides, Norfloxacin and resistant to Amoxicillin. the rest of the biological test was normal.

The outcome was fatal six hours after hospitalization due to septic shock. We didn't realize a genetic study for newborn because of his early death and later, genetic counseling was programmed for the parents.

3. DISCUSSION

Ichthyoses are hereditary skin disorders associated with abnormalities of keratinization, forming a heterogeneous group of pathologies characterized by the accumulation of epidermal scales. Different criteria are applied for their classification and in particular the time of appearance of the clinical signs; distinguishing between congenital ichthyosis (from birth) and vulgar ichthyosis (first years of life) [3].

There are limited studies on the benefits of antibiotic or fungal prophylaxis in patients with Harlequin ichthyosis. Cases of Harlequin ichthyosis with patients dying from *Pseudomonas aeruginosa* sepsis despite broad-spectrum antibiotics have been described [4].

Vibrio cholerae sepsis is rare, particularly in neonatology, although sporadic cases have been reported in older patients [2].

Vibrio cholerae is a Gram-negative bacillus with a polar flagellum. Of the over 200 serotypes identified, only the enterotoxin producers of serotype 01 and 0139 can cause epidemics by infections especially of the gastrointestinal tract.

Non-toxigenic O1 strains and non-O1/O139 strains of *Vibrio cholerae* can cause diarrhea and sepsis, especially, in immunocompromised patients. In this case the newborn is considered an immunocompromised patient; in his history there isn't any gastrointestinal infection. However, the genotype, the virulence phenotype and the factors that allow strains to invade the bloodstream are not well understood [5].

First line antimicrobial therapy to eradicate *Vibrio cholera* is Tetracycline, Ciprofloxacin, Erythromycin, or Azithromycin, depending on resistance [6]. Acitretin, a systemic retinoid, is the drug of choice for ichthyosis because of its short plasma half-life, which reduces side effects. Before its introduction, mortality was about 70% in the first weeks of life. However, it is difficult to perform clinical trials evaluating efficacy because of the very low prevalence of Harlequin ichthyosis [7].

CONCLUSION

Although *Vibrio cholerae* septicemia is not common, and less so in neonates, it should be considered even in the absence of risk factors, since new germs are re-emerging.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

HUMAN AND ANIMAL RIGHTS

No animals/humans were used for studies that are the basis of this review.

CONSENT FOR PUBLICATION

A written informed consent was obtained from all patients when they were enrolled.

CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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Declared none.

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