Cutaneous Scarring and Hyperpigmentation at Birth: Congenital Herpes Simplex Infection Resembling Incontinentia Pigmenti

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INTRODUCTION

Congenital herpes simplex virus (HSV) infection is a rare infection associated with severe morbidity and mortality. It typically results from perinatal exposure to HSV from maternal genital lesions. Most newborns with neonatal HSV infections are asymptomatic at birth but become ill within a few days. On the other hand, in utero exposure to HSV can cause congenital infection and can present with lesions at birth. These lesions consist of skin scarring and hyperpigmentation which resemble other cutaneous conditions. Characteristic vesicles are commonly absent.

CASE REPORT

We report the case of a well appearing, afebrile male newborn who was born at GA 39w2d via NSVD to a 23-year-old G3P2 mother. Newborn examination on Day of Life 1 (DOL1) was remarkable for hyperpigmented patches with mild scaling and skin scarring noticed on the back and axilla. Mother's prenatal labs and imaging were normal and there was no known history of genital herpes. Due to the noticed skin findings, lab workup was done including blood count and liver enzymes which were normal. A presumptive diagnosis of Incontinentia Pigmenti was made and the patient was discharged home to follow up with dermatology. Because one vesicular lesion was also noted on his right hand, HSV polymerase chain reaction (PCR) swabs from the lesion and the mucous membranes, along with blood, were collected before discharge, per Pediatric Infectious Disease recommendations.

Subsequently, the newborn developed generalized vesicular rash while at home however remained clinically stable. On DOL4, the HSV PCR came back positive for HSV 2 from the lesions, mucous membranes and blood. Thus, the newborn was readmitted for further work up and intravenous acyclovir therapy. HSV PCR from the cerebrospinal fluid (CSF) was negative. Head ultrasound done on DOL7 revealed nonspecific hypoechoic lesions concerning for calcifications. The newborn was then treated with intravenous acyclovir for 21 days and the skin lesions improved. He was discharged home on DOL25 and was continued on suppressive therapy with valacyclovir for 6 months to prevent recurrence of vesicles.

CASE REPORT

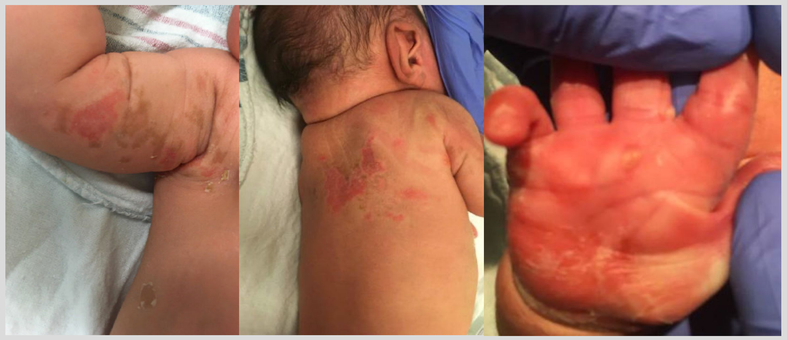


Figure 1. DAY OF LIFE 1 - Cutaneous scarring and hyperpigmentation at birth

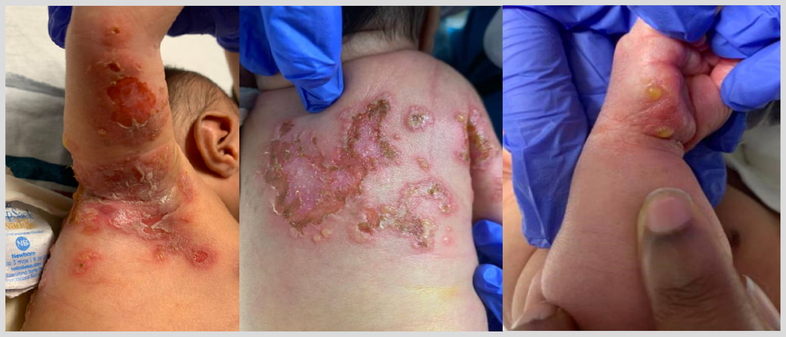


Figure 2. DAY OF LIFE 4 - Flare up of generalized vesicular rash with areas of peeling

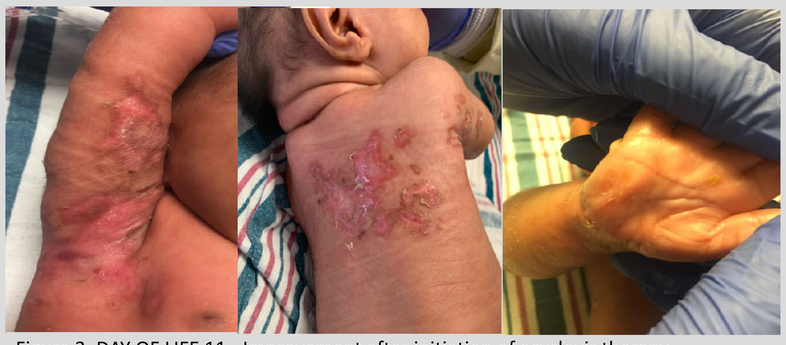
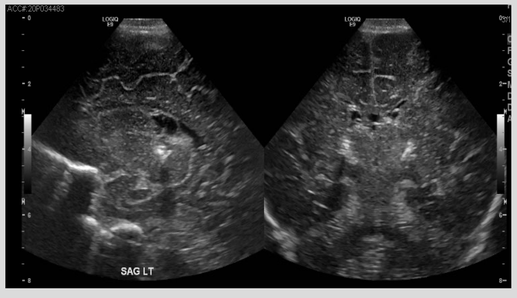


Figure 3. DAY OF LIFE 11 - Improvement after initiation of acyclovir therapy



Head Ultrasound

Figure 4. DAY OF LIFE 7 - Nonspecific hypoechoic lesions concerning for intracranial calcifications

DISCUSSION

Congenital herpes simplex infection is caused *in utero* HSV transmission from mother-to-child. It accounts for only 5% of neonatal HSV infections. HSV transmission can also occur during the peripartum period (85%) as a result of viral shedding from the genital tract around the time of delivery, or postnatally (10%) due to direct contact with HSV-infected persons.

Congenital HSV is characterized by the triad of cutaneous, eye and neurologic manifestations present at birth. The history, physical exam and investigation findings of this patient are consistent with a diagnosis of congenital HSV. The initial findings might be confused with the findings of Incontinentia pigmenti which occurs from birth to the first 2 weeks of life. Incontinentia pigmenti (IP) is a rare genetic disorder that affects the skin, eye, teeth, and central nervous system. The characteristic skin findings of IP are usually present at birth. Other blistering skin conditions to be considered include transient neonatal pustular melanosis and erythema toxicum neonatorum. These are usually benign, noninfectious and self limiting.

Using polymerase chain reaction, a diagnosis of congenital HSV can be established by obatining swabs from the nasal, oropharyngeal, umbilical, anal, and skin lesions. Testing the blood and the CSF is important to assess the severity of the disease. Treatment with intravenous acyclovir is indicated for flare up and active lesions. Typically, patients will be also placed on 6 months of suppressive therapy with oral acyclovir.

Our case highlights the importance of maintaining a high index of suspicion of any neonatal skin lesion despite absence of concerning maternal history or characteristic vesicular lesions. Early identification of congenital HSV and early initiation of acyclovir therapy is pivotal in mitigating long term sequelae.

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