



Incontinence pigmenti (Bloch–Sulzberger syndrome) in the neonatal stage: A case report.

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Abstract

Introduction: Incontinence pigmenti (IP) is a rare neurocutaneous disorder caused by a hereditary alteration linked to the X chromosome. A clinical case is presented.

Case report: A newborn girl who was evaluated at 48 hours of life for skin lesions with erythematous vesicle-pustular blisters located on her left lower limb presented at birth. The lesions spread, increasing in number, to other regions and sustaining irritability.

Diagnostic workshop: A skin biopsy was performed, and incontinence pigmenti was diagnosed. A genetic study revealed a dominant genodermatosis linked to the X chromosome, associated with the IKBKG gene mutation. Hematological, chemical, and hormonal control studies were standard. There was no neurological or ophthalmic involvement.

Evolution: The patient with lesions in the vesicular phase received topical zinc oxide and calamine treatment. During disease progression, oxacillin was added to infected papule-type lesions. She was discharged in good condition and remained under observation.

Conclusions: In the present case, differential diagnoses such as bullous impetigo, bullous pemphigoid, neonatal herpes, cytomegalovirus, mastocytosis, hereditary epidermolysis bullosa, or toxic erythema were proposed.

Keywords:

Incontinence Pigmenti, Genodermatosis, Woman, Case report

Abbreviations

IP: incontinence pigment.

Supplementary information

No supplementary materials are declared.

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Author contributions

Santiago Chavesta Aray: Conceptualization, data curation, Fund acquisition, Project administration, Supervision.

Evelyn Ordóñez González: conceptualization, formal analysis, methodology.

Letty Muzzio Prott: Conceptualization, data curation, formal analysis, funding acquisition, research, writing - original draft.

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Availability of data and materials

The data sets used and analyzed during the present study are available from the corresponding author upon reasonable request.

Introduction

Incontinence pigmenti (IP) is a rare neurocutaneous disorder caused by a hereditary alteration linked to the X chromosome, with multisystem involvement characterized by initial vesicular-bullous lesions in bands. These linear or dotted lesions follow Blaschko's lines. The illness is usually present or appears shortly after birth. IP was first described by Garrod in 1906, but the first complete description was attributed to Bardach, Bloch, Siemens, and Sulzberger during the 1920s; based on its clinical and histopathological characteristics, it became known as Bloch–Sulzberger syndrome. It has a worldwide distribution, but its prevalence is still unknown. However, its incidence is 1 in every 40,000 or 50,000 live births [1]. It occurs almost exclusively in women with a 20:1 ratio since nearly all men die in the perinatal stage unless their karyotype is 47XXY or they present somatic mosaicism and hypomorphic mutation [2, 3]. This entity is caused by the deletion of exons 4-10 of the IKBKG gene, which is present in the Xq28 locus in 80 to 90% of cases. This gene encodes the NEMO subunit of the protein kinase complex, which is an essential inhibitor of antiapoptotic and proinflammatory cellular signaling mediated by NF-kappa B. Consequently, dysregulation of the production of cytokines, chemokines, and adhesion molecules leads to the predisposition of mutant cells to apoptosis. Other mutations have been associated with IP, such as point missense mutations in exons 2 to 10 [4].

Dermatological findings are the first to appear and are divided into four stages: vesicles, generally perinatal; warty lesions; hyperpigmentation, which follows a characteristic pattern; and atrophic or scarring hypochromic lesions. Skin alterations may be present at birth or appear in the first 15 days of life; over time, they fade, eventually disappearing during puberty and adulthood. The difficulty of diagnosis lies in the fact that the fourth stage of the disease is a late manifestation, probably underdiagnosed, which consists of hypopigmented atrophic alterations in the lower limbs. In some cases, residual neurological lesions remain, such as mental retardation or severe motor and ocular impairments [5]. Patients present a variable phenotype due to the inactivation of one of the X chromosomes (heterochromatinization) very early in embryogenesis, determining the variable effects on different organs. The involvement of the central nervous and ocular systems determines the prognosis. Its early detection is essential since there is no curative etiological treatment, and treatment with supportive measures for the different accompanying disorders

must be carried out [6]. We decided to present this case for these reasons and since it is a rare genodermatosis.

Clinical case

A female neonate born by vaginal delivery at a health center and assessed at 38 weeks of gestation by the Ballard scale, weighing 2950 grams, appropriate for gestational age, with an Apgar of 7/9 and a perinatal history of a 31-year-old mother with a controlled pregnancy and non-consanguineous parents. She was discharged after 48 hours of life. On the third day of life, she came to our institution due to skin lesions, such as erythematous vesicle-pustular blisters, which are located on her left lower limb and are present since birth, as the days spread, increasing in number to other regions, in addition to sustained irritability (Figure 1).

Figure 1. Lesions present in the patient.



The vesicles are erythematous with pustular blisters, following a linear path in the left lower limb.

Physical exam

During the physical examination upon admission, attention was drawn to a dermatosis spread to the anterior trunk, posterior trunk, upper extremities, lower extremities, genital area, and gluteal area. The lesions are characterized by erythematous papules, vesicle-type, pustular, and yellowish content. Some lesions adopted a linear pattern, and others were curvilinear. The lesions converged, forming erythematous plaques with fine peeling in some areas. The size of the lesions was variable. No involvement was found in the face, oral mucosa, or ocular area (Figure 2). The neurological examination was expected, and the rest of the physical examination showed no abnormalities.

Figure 2. Lesions present in the patient.

Disseminated dermatosis.

The initial diagnosis was sepsis associated with neonatal dermatosis. First-line antibiotic treatment based on ampicillin and gentamicin was initiated.

Given the persistence of disseminated dermal lesions, the dermatology service was consulted on the third day of hospitalization. According to the diagnostic criteria of Landy and Donnai, the patient was suspected of having incontinentia pigmenti in the vesicular phase. A histological study of the biopsy taken from one of the gallbladders of the left leg confirmed the diagnosis. Topical treatment with zinc oxide and calamine was initiated.

The genetics service evaluated the patient where the diagnosis of dominant genodermatosis linked to the X chromosome, associated with the *IKBKG* gene mutation, was made. This entity led to a screening of the ophthalmological and neurological associations, reporting it as usual.

During its evolution, over-infected papule-type lesions were added ([Figure 3](#)), which required evaluation by the Infectious Diseases service, where antimicrobial coverage with oxacillin was indicated, with new blood cultures reported without bacterial growth.

Figure 3. Evolution of the patient's injuries.

Injuries superinfected.

Evolution

The patient was discharged at 28 days of age with lesions still in stage 1 ([Figure 4](#)).

Figure 4. Evolution of the patient's injuries.

Injuries improved at the end of treatment.

Auxiliary diagnostic methods

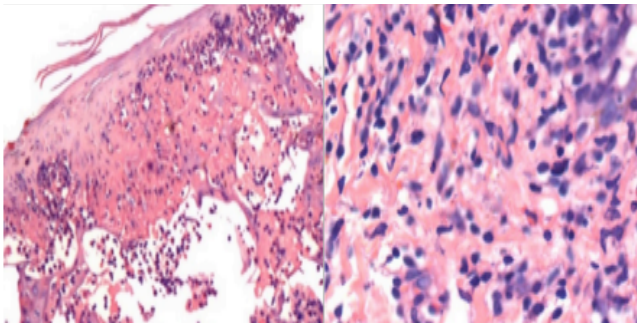
Paraclinical parameters included admission blood count, severe eosinophilia (greater than 3,000/ μ l), white blood cell count, neutrophil count, lymphocyte count, red blood cell count, and platelet count within normal ranges. A peripheral blood smear revealed 12,000 $\times$ 10⁹/L leukocytes, 41% segments, 11% lymphocytes, 48% eosinophils (4,290/ μ l absolute), and 192,000 $\times$ mm³ platelets, with no alterations in

the red series. Negative acute phase reactants. Hepatic, pancreatic, and renal functions were preserved. VDRL serology – HIV nonreactive, TORCH negative. Blood cultures on two occasions were negative—no growth. Simple cranial tomography revealed no alterations in the brain parenchyma and slight ventricular asymmetry with a predominance of the left hemisphere. The fundus was normal.

Skin biopsy: left leg vesicles

Direct examination and skin culture were negative. Pathological anatomy reports included epidermis with acanthosis, hyperkeratosis, abundant dyskeratocytes and exocytosis of eosinophils, eosinophilic spongiosis, intraepidermal and subcorneal vesicles with abundant eosinophils, and perivascular inflammatory infiltration of eosinophils in the papillary and reticular dermis (Figure 5).

Figure 5. Histopathological diagnosis.



Final pathological diagnosis: incontinentia pigmenti stage 1 (vesicle-erythematous-bullous).

Evolution

Currently, the patient is under the control and monitoring of a multidisciplinary team that includes pediatrics, dermatology, neurology, ophthalmology, genetics, and, in the future, dentistry.

Discussion

Incontinentia pigmenti is a genetic disease belonging to the group of genodermatoses. It is a rare multisystem disorder whose incidence is 1 in every 40,000 live births. However, it is probably more common because its presentation is variable and can quickly go unnoticed. The lesions are possibly confused with those of other entities, such as viral or bacterial infections and reactions such as toxic erythema in the neonatal period and the initial stages [1]. It is transmitted in a dominant form linked to the X chromosome, and only women suffer. The ratio of affected women/men is 20:1; however, in other studies, an incidence of up to 40:1 has been

reported since, in men, it is lethal due to the intrauterine complications that occur [2].

In 2000, Smahi and collaborators reported that more than 80% of patients had a deletion of exons 4-10 of the nuclear factor- κ B essential modulator (NEMO) gene, which encodes the homonymous protein that activates the NF- κ B transcription factor. NF- κ B is a homodimeric or heterodimeric protein formed by subunits belonging to the Rel protein family (such as p50, p52, cRel, RelA, and RelB). Under basal conditions, it is found in the cytoplasm bound to an inhibitor called I κ B (an inhibitor of κ B). The phosphorylation of NF- κ B is carried out by a protein complex called IKK (I κ B kinase), which is formed by three subunits, IKK1 (also called IKK- α), IKK2 (or IKK- β) and NEMO (or IKK- γ). In response to stimuli such as interleukin 1- β (IL-1- β), tumor necrosis factor α (TNF- α), and lipopolysaccharides (via Toll-like receptors or TLRs), I κ B is phosphorylated by IKK and then subjected to proteolysis. Cleavage of I κ B allows NF- κ B to migrate to the nucleus, where it binds to DNA and increases the transcription of genes that promote cell survival, including proinflammatory cytokines, such as TNF- α , IL-1, and IL-6; chemokines, such as MIP-1 α (macrophage inflammatory protein-1 α), RANTES (regulated upon activation, normal T-cell expressed and secreted); adhesion molecules, such as E-selectin and VCAM1 (vascular cell adhesion molecule 1); antiapoptotic proteins, such as c-IAP-1/2, AI, Bcl-2, and Bcl-XL; proliferation promoters (cyclin D1); microbicidal molecules (defensins); and generators of other intermediates (inducible nitric oxide synthase). NF- κ B is not activated in NEMO-deficient cells. Interestingly, NEMO has no catalytic function in the enzyme complex but only a structural and regulatory function [7].

Because these cytokines are produced in response to NF- κ B, they are considered to be secreted by NEMO (+) cells, perhaps in response to necrosis of adjacent NEMO (–) cells that release cellular debris and heat shock proteins. This generates a vicious cycle since TNF- α and IL-1 increase the death of adjacent NEMO (–) cells susceptible to apoptosis and activate NF- κ B in the same NEMO (+) cells, amplifying the apoptotic stimulus. However, another study revealed that NEMO+ cells are not essential for initiating an inflammatory response, so the presence of NEMO keratinocytes is sufficient. Eotaxin is also postulated to be important in producing PI skin lesions [8].

Eotaxin is a chemotactic cytokine specific for eosinophils. Immunohistochemical analysis of histological skin sections with vesicles and verrucous plaques revealed eotaxin expression in areas with eosinophil infiltration. Thus, it is proposed that eotaxin promotes the chemotaxis of eosinophils

toward the epidermis, which releases proteases that degrade tonofilaments and desmosomes, causing spongiosis and vesicle formation [9].

The diagnosis is made clinically based on a sequential history of skin lesions and their characteristics. Landy and Donnai (1993) recommended diagnostic criteria before discovering the causative gene (Table 1).

Table 1. Diagnostic criteria for pigmentary incontinence.

Patients without a family history of pigmentary incontinence	
Major criteria	
1. Typical neonatal rash: eosinophilia + vesicles and erythema. 2. Blaschko lines , mainly on the trunk, which disappear at puberty. 3. Alopecia and linear atrophy.	
Minor Criteria	
1. Tooth abnormalities 2. Nail abnormalities. 3. Alopecia. 4. Retinopathy.	
Patients with a family history of pigmentary incontinence.	
1. Typical neonatal rash: eosinophilia, typical hyperpigmentation, alopecia and linear atrophy. 2. Male pattern baldness. 3. Retinopathy. 4. Tooth abnormality 5. Recurrent abortions of male fetuses.	

At least one of the main criteria is necessary for a diagnosis. The primary criteria were typical neonatal skin rash and hyperpigmentation, linear or atrophic skin lesions, and affected first-degree relatives. The minor criteria are dental involvement, alteration of nerves, and retinal disease. The sensitivity and specificity of these criteria have yet to be determined, and their correlation with the most recent molecular findings has yet to be proven. However, they can still offer valuable guidance to clinicians. Wood's lamp assessment may be helpful in older children to make pigmentation abnormalities more evident. Other diagnostic characteristics include the presence of eosinophilia and the use of a histological skin biopsy that provides diagnostic confirmation [10-12].

IP is characterized by linear skin lesions (Blaschko lines) that represent the migration routes of embryonic cells that begin to be expressed at birth and spontaneously evolve in four stages due to their histopathological correlation [10] (Table 2).

Stage 1 is inflammatory vesicular-bullous with spongiosis, with bands of intraepidermal vesicles filled with eosinophils that are generally present at birth or in the first weeks of life, as evidenced in the patient and resolve at 4-6 months of age. In stage 2, warty, hyperkeratotic lesions and acanthosis are observed, as well as dyskeratotic or necrotic keratinocytes that can appear in areas other than stage 1; these lesions resolve in 6 months. Stage 3 consists of macules with linear hyperpigmentation that gradually increase following the Blaschko lines; epidermal thinning and vacuolization of the basal layer are also observed. Stage 4 is characterized by hypopigmentation and atrophy [11].

The major criteria describe the typical dermatological findings associated with the entity, which make the clinical diagnosis, together with the minor criteria associated with other anomalies that may be evident [13].

In addition, it is essential to differentially diagnose each stage from other entities and thus pay attention to the suspicion of incontinence pigmenti. The first stage can be confused with infections (bacterial and herpes simplex virus), toxic erythema, and epidermolysis bullosa. The second stage, with linear epidermal nevus and linear and swirling nevoid hypermelanosis, may be identical to the third stage [14].

Its early detection is, therefore, essential since there is currently no etiological treatment, and symptomatic treatment of the different alterations must be carried out. Neurological complications occur in 30-50% of patients and include mental retardation, seizures, spastic paralysis, microcephaly, brain malformations (cerebrovascular events), hyperactivity, and cerebellar ataxia. In general, these patients who present with repeated seizures present with acute encephalopathy in the neonatal period.

Among neurocutaneous disorders, IP is the third most common form of clinical presentation of epileptic seizures, and hypsarrhythmia is an electrical phenotype. The first corresponds to tuberous sclerosis (20%), and the second corresponds to neurofibromatosis type 1. Skin lesions of the scalp located mainly in the vertex in 38% of patients are frequently associated with underlying brain lesions. This characteristic constitutes an expression of genetic lyonization in these patients, the process by which some cells of exact embryological origin express the defect, and others do not.

The eyes can show the following alterations: microphthalmia, papillitis, retinopathy, and eyelid deformity. These alterations are frequently associated with CNS involvement and can appear in the first year of life, requiring early ophthalmological check-ups.

Table 2. Phases of pigmentary incontinence.

Phase	Description	Distribution	Histology
Vesicular: 80%. Two weeks to four months of age at .	Blascko 's lines.	Lower limbs, trunk, head, neck.	Intraepidermal vesicles , spongiosis , dyskeratosis and eosinophilic and neutrophilic inflammatory infiltration.
Warty: 70%. They disappear after six months or persist into adulthood.	Warty plaques on an erythematous base, with a linear arrangement, following Blascko 's lines.	Distal third of the lower limbs ^b .	Acanthosis, hyperkeratosis and papillomatosis.
Hyperpigmented: 90%. Progressive regression in adolescence.	Blaschko 's lines.	Lower limbs, trunk, armpits, inguinal region.	Extensive pigment incontinence.
Hypopigmented: 30-70%. Permanent. They appear in adolescence.	Alopecic and anhidrotic areas.	Back of the legs and arms.	Absence of pigment, as well as eccrine glands.

- a. The lesions they can appear years later with illnesses febrile.
- b. They may or may not correspond to sites of previous injury in the vesicular phase .

The dentition in these children frequently presents with hypodontia or anodontia (approximately 43%), delayed eruption, constitutional deformities (conical teeth), and enamel alterations, the most common extracutaneous involvement. Both temporary and permanent teeth are affected. At the bone level, the alterations are characterized by hemivertebrae, scoliosis, spina bifida, syndactyly, ear anomalies, extra ribs, and skull deformities. Another characteristic alteration of these patients is periungual keratotic tumors, located mainly in the toes between puberty and the 3rd decade of life. They can progress toward spontaneous regression or sometimes toward continuous growth with pain, nail dystrophy, and bone destruction of the distal phalanx (despite their benign histology, in these cases, they require removal). Congenital hypothyroidism, myasthenia gravis, and Wilms tumor have been described in some cases of IP. However, the link cannot be established; liver evaluation has recently been suggested, thus further expanding medical care for patients with this condition.

Treatment is decided based on extracutaneous anomalies since skin lesions are benign. Skin lesions are self-resolving, and their treatment can be symptomatic. The use of topical retinoids at 0.05% for treating verrucous lesions with good results has been described, as has the use of topical corticosteroids and emollients. The multidisciplinary team included several specialists, of whom the ophthalmologist stands out since close ophthalmological surveillance is required for the first years of life due to the possible complications mentioned. Experts suggest monthly control from birth to 4 months, quarterly control until one year of age, semiannual control until three years of age, and annual

control. Ophthalmic treatments may include photocoagulation for fibrovascular proliferation and vitreoretinal surgery for retinal detachments. Dental care will be carried out periodically to detect complications during oral check-ups. During the onset of neurological abnormalities, interconsultation will be made with him, among other specialties, to optimize behaviors and improve his quality of life.

On the other hand, the update in the diagnosis proposed by Rosser does not include skeletal alterations, including short stature, hemivertebra, kyphosis, scoliosis, supernumerary clavicle, hip dysplasia, hemiatrophy, equinovarus foot, and syndactyly in the toes. In addition, cardiopulmonary disorders, such as atrial septal defects, left ventricular endomyocardial fibrosis, tricuspid regurgitation, tetralogy of Fallot, and pulmonary hypertension, can occur even in the absence of cardiovascular alterations. Additionally, these criteria do not mention the risk of contracting recurrent infections. Future updates will likely be described to expand on the main clinical features that may be present. Genetic evaluation is crucial to facilitate understanding of the entity and allows it to be confirmed through molecular genetic study, which is practical in doubtful cases. The prognosis is generally good, but periodic evaluation by the interdisciplinary team must be carried out, as already mentioned. Family genetic counseling is equally important; in the presence of a dominant X-linked inheritance pattern, which is usually lethal in men, a female patient with the entity has a 50% risk of having daughters with IP. In the case of a male fetus, abortion can occur in 50%, and if an affected child appears, a cytogenetic study is indicated [15].

Finally, it is essential to highlight that IP is considered a multisystemic genodermatosis, which requires early multidisciplinary patient follow-up and genetic counseling for the family. Therefore, the early diagnosis of these patients is based on identifying the initial skin lesions. Thus, IP should be considered when diagnosing vesiculobullous erythema and neonatal pustulosis.

Conclusions

IP is a rare, potentially genetic severe disease whose most prominent clinical manifestations are found in the skin and are the basis for early diagnosis in the neonatal stage. Genetic assessment is recommended in all cases, and genetic counseling to parents about the risk of transmission to their future offspring is crucial. Neurological findings and ophthalmic alterations constitute the most severe disease and are the main prognostic factors. To date, there is no curative treatment; the main thing that can be provided is support measures and treatment of accompanying disorders. In the face of a multi-system disorder, medical follow-up must be carried out long-term and in an individualized and interdisciplinary manner, including evaluations by the Pediatrics, Dermatology, Neurology, Ophthalmology, Dentistry, and Genetics services.

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Statements

Ethics committee approval and consent to participate

Clinical cases are not needed.

Publication consent

The authors have permission to publish the images and photographs from the patient's parents.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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