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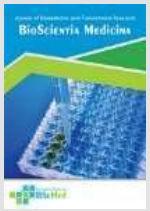

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Early-Onset Urticaria Pigmentosa in a 7-Month-Old Infant: A Case Report

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A B S T R A C T

Background: Urticaria pigmentosa (UP) is the most common form of cutaneous mastocytosis in children, accounting for 70-90% of cases. UP typically presents in infancy or early childhood with characteristic skin lesions that vary in appearance. The hallmark of UP is the presence of reddish-brown macules or papules that urticate or blister upon rubbing, a phenomenon known as Darier's sign. Histopathological examination of a skin biopsy is confirmatory, demonstrating an increased number of mast cells in the dermis. **Case presentation:** We report a case of early-onset UP in a 7-month-old infant boy who presented with multiple hyperpigmented macules all over his body. The lesions were pruritic and had progressively increased in size and number over the past two weeks. Dermatological examination revealed multiple erythematous macules on the face, hyperpigmented macules on the chest, abdomen, and extremities, hyperpigmented plaques on the legs, and a nodule on the back. The lesions exhibited a positive Darier's sign, characterized by swelling and redness upon rubbing. Systemic examination was unremarkable, with no evidence of hepatomegaly, splenomegaly, or lymphadenopathy. Dermoscopy of the lesions revealed a central pigment network and a light brownish peripheral structure, suggestive of UP. Histopathological examination of a skin biopsy confirmed the diagnosis of UP, demonstrating an increased number of mast cells in the dermis. The patient was treated with oral antihistamines (cetirizine) and topical corticosteroids (hydrocortisone) for four weeks, resulting in significant regression of the lesions. **Conclusion:** This case highlights the importance of recognizing the clinical features of UP in infants. Early diagnosis and appropriate management can help alleviate symptoms and improve the quality of life for affected children.

1. Introduction

Mastocytosis, a rare disorder characterized by the abnormal proliferation and accumulation of mast cells in various tissues, primarily the skin, presents a unique challenge in the realm of dermatological and immunological diseases. These cells, integral to the immune system's response to allergens and other triggers, release histamine and other inflammatory mediators. In mastocytosis, their excessive accumulation can lead to a range of symptoms, including skin lesions, itching, flushing, and gastrointestinal disturbances. Urticaria pigmentosa (UP), the most common form of cutaneous mastocytosis in children, accounts for 70-90% of cases, typically presenting in infancy or early childhood with characteristic skin lesions that vary in appearance. The hallmark of UP is the presence of reddish-brown macules or papules that urticate or blister upon rubbing, a phenomenon known as Darier's sign. These lesions are often pruritic and may be accompanied by flushing, dermatographism, or blistering. The diagnosis of UP is primarily clinical, based on the characteristic appearance of the skin lesions and the presence of Darier's sign. Dermoscopy, a non-invasive technique that allows visualization of skin structures at higher magnification, can aid in the diagnosis by revealing specific patterns associated with UP. Histopathological examination of a skin

biopsy is confirmatory, demonstrating an increased number of mast cells in the dermis.[1-4]

The prevalence of mastocytosis is estimated to be between 1:10,000 and 1:30,000, affecting both children and adults. However, the true prevalence may be higher due to underdiagnosis, as mild cases often go unrecognized. Cutaneous mastocytosis is the most common form of the disease, particularly in children. UP typically presents in infancy or early childhood with characteristic skin lesions that vary in appearance. The hallmark of UP is the presence of reddish-brown macules or papules that urticate or blister upon rubbing, a phenomenon known as Darier's sign. These lesions are often pruritic and may be accompanied by flushing, dermatographism, or blistering.[5-7]

The diagnosis of UP is primarily clinical, based on the characteristic appearance of the skin lesions and the presence of Darier's sign. Dermoscopy, a non-invasive technique that allows visualization of skin structures at higher magnification, can aid in the diagnosis by revealing specific patterns associated with UP. Histopathological examination of a skin biopsy is confirmatory, demonstrating an increased number of mast cells in the dermis. The management of UP primarily focuses on symptomatic relief and avoidance of triggers that can exacerbate symptoms. Antihistamines are commonly used to control itching and flushing, while topical corticosteroids may be prescribed for localized lesions. In most cases, UP in children resolves spontaneously by adolescence, although some patients may experience persistent or recurrent symptoms.[8-10] In this case report, we present a case of early-onset UP in a 7-month-old infant boy.

2. Case Presentation

This report describes a 7-month-old male infant presenting with a constellation of cutaneous manifestations suggestive of urticaria pigmentosa (UP). The patient's mother, the primary informant, detailed a history of progressively worsening skin lesions that began shortly after birth. The infant's mother reported the initial appearance of red bumps on the child's back two days postpartum. These lesions gradually spread to the chest and were treated with a topical ointment, after which they transformed into brownish patches. Subsequently, new reddish bumps emerged on the face, arms, and legs, prompting treatment with both topical ointment and oral syrup. Further lesions appeared on the neck and followed a similar pattern of evolution into brownish patches. The mother noted that new reddish bumps occasionally arose after breastfeeding or scratching. In addition to the cutaneous symptoms, the infant had recently experienced fever and a cold cough, for which he received treatment from a general practitioner. Notably, there was no history of facial angioedema, flushing, dyspnea, diarrhea, or abdominal distension. The family history was unremarkable for similar skin conditions. The infant was alert and cooperative during the examination. His vital signs were within the normal range for his age: pulse rate 100 beats per minute, respiratory rate 28 breaths per minute, and temperature 36.5°C. His height and weight were also appropriate, with a body mass index Z-score falling within the normal range. Systemic examination revealed no significant abnormalities. The infant's head, eyes, ears, nose, and throat were all within normal limits. There was no evidence of lymphadenopathy in the cervical, axillary, or inguinal regions. Examination of the chest and lungs revealed symmetrical chest expansion, clear breath sounds bilaterally, and no signs of respiratory distress. Cardiac examination revealed a palpable apical impulse, regular heart rhythm, and no murmurs or gallops. Abdominal examination was unremarkable, with no hepatosplenomegaly or palpable masses. Examination of the extremities showed no edema. Cutaneous examination revealed a diverse array of lesions distributed across the infant's body. Multiple erythematous macules were observed on the face, while hyperpigmented macules were present on the chest, abdomen, and right upper limb. Hyperpigmented plaques were noted on the legs, and a solitary nodule was identified on the back.

Importantly, all lesions exhibited a positive Darier's sign, characterized by localized swelling and erythema upon rubbing. Complete blood count revealed a hemoglobin level of 11.4 g/dL (normal range: 9.6-15.6 g/dL), a leukocyte count of 19.03 x 10^9/L (normal range: 4.0-12.0 x 10^9/L), and a platelet count of 508 x 10^9/L (normal range: 150-450 x 10^9/L). Notably, the leukocyte count was elevated with a predominance of eosinophils, suggesting an underlying inflammatory process. Coagulation studies, including prothrombin time (PT) and activated partial thromboplastin time (APTT), were within normal limits. Liver function tests, including serum glutamic-oxaloacetic transaminase (SGOT) and serum glutamic-pyruvic transaminase (SGPT), were also within normal limits. Renal function tests, including blood urea nitrogen and creatinine, were unremarkable. Blood glucose levels were within the normal range. Dermoscopic examination of the skin lesions revealed characteristic features suggestive of UP. Specifically, a central pigment network and a light brownish peripheral structure were observed. These findings, in conjunction with the clinical presentation and positive Darier's sign, further supported the diagnosis of UP. A skin biopsy was obtained from a representative lesion and submitted for histopathological evaluation. Microscopic examination revealed an increased number of mast cells in the dermis, confirming the diagnosis of UP. Based on the collective findings from the anamnesis, physical examination, dermatological examination, laboratory investigations, dermoscopy, and histopathological examination, a definitive diagnosis of urticaria pigmentosa (cutaneous mastocytosis) was established. This case underscores the importance of a comprehensive and multidisciplinary approach to the diagnosis of UP in infants. The early onset of symptoms, the characteristic morphology of the skin lesions, the positive Darier's sign, and the histopathological confirmation of mast cell infiltration collectively contributed to the accurate diagnosis in this case (Table 1).

The management of this infant with urticaria pigmentosa (UP) involved a dual approach: comprehensive parental education and targeted symptomatic treatment. A detailed discussion was conducted with the infant's parents to provide a thorough understanding of UP. This included explaining the etiology of the disease, emphasizing its generally benign nature and the typically favorable prognosis, particularly the high likelihood of spontaneous resolution before puberty. The discussion also encompassed potential complications and the importance of recognizing and managing them. A key component of the educational intervention was to empower the parents to identify and avoid potential trigger factors that could exacerbate the infant's symptoms. These triggers included certain medications (such as non-steroidal anti-inflammatory drugs), specific foods, local or systemic anesthetics, excessive heat, and friction. The parents were also informed about the possibility, albeit rare, of systemic involvement, which could manifest as whole-body flushing, shortness of breath, and diarrhea. Finally, the importance of regular follow-up examinations was emphasized to monitor the progression of the disease and to promptly detect any signs of systemic involvement. The specific treatment regimen consisted of both systemic and topical therapies to alleviate the infant's symptoms and improve his quality of life. Cetirizine, a second-generation antihistamine, was prescribed at a dosage of 2 mg once daily. Cetirizine is known for its efficacy in reducing pruritus and urticaria, which are common symptoms associated with UP. The oral solution formulation was chosen for ease of administration in this young infant. Hydrocortisone 2.5% cream, a mild topical corticosteroid, was prescribed for application twice daily to the affected skin areas. Topical corticosteroids are effective in reducing inflammation and relieving pruritus associated with UP lesions. The low potency of hydrocortisone cream was deemed appropriate for use in this infant, minimizing the risk of adverse effects associated with more potent corticosteroids. The infant was scheduled for a follow-up examination one month after the initial consultation. At this follow-up visit, the parents

reported significant improvement in the infant's condition. The parents reported that no new reddish patches or bumps had appeared, and the existing lesions had reduced in size and intensity of color, becoming more faded. The infant had not experienced any fever, diarrhea, nausea, or vomiting, indicating the absence of systemic involvement. The infant's vital signs remained within the normal range for his age. Dermatological examination revealed a marked improvement in the skin lesions. The erythematous macules had subsided, and the hyperpigmented macules and plaques had faded significantly. Based on the positive response to treatment, the initial treatment regimen was continued. The infant was to continue receiving cetirizine 2 mg once daily and hydrocortisone 2.5% cream twice daily to the affected skin areas. This case demonstrates the effectiveness of a combined approach to managing UP in infants. By providing comprehensive education to the parents and implementing targeted symptomatic treatment, the infant's symptoms were effectively controlled, and his quality of life was improved. The regular follow-up ensured ongoing monitoring of the disease progression and allowed for adjustments to the treatment plan as needed. The favorable outcome in this case highlights the importance of early diagnosis, appropriate management, and close follow-up in the care of infants with UP (Table 2).

Table 1. Anamnesis, physical examination, dermatological examination, laboratory, dermoscopy, histopathological and diagnosis.

Anamnesis	7-month-old boy with brown patches and bumps that felt itchy on face, chest, abdomen, back, neck, both arms and legs. Lesions increased in size and number since 2 weeks ago. Red bumps appeared on the back after birth, spread to the chest, treated with ointment, turned into brownish patches. New reddish bumps appeared on face, arms, and legs, treated with ointment and syrup. New reddish bumps appeared on neck, turned into brownish patches. Sometimes new reddish bumps appear after breastfeeding or scratching. Fever and cold cough, treated by a general practitioner. No history of facial angioedema, flushing, dyspnea, diarrhea, or swollen stomach. No family history of similar disease.
Physical examination	**Consciousness**: Compo's mentis cooperative **General State**: Moderate illness **Vital Sign:** Pulse Rate: 100 x/minute Respiratory Rate: 28 x/minute Temperature: 36.5°C Height: 53.0 cm Weight: 8.2 kg; Body Mass Index: Z-score +2 s/d -2 (normoweight) **Head**: Within normal limit **Eye**: Conjungtiva anemis -/-, sclera icteric -/- **Ear, Nose, and Throat**: Within normal limit **Teeth and mouth**: Within normal limit **Lymph Nodes**: No enlargement on cervical, axillaries, or inguinal lymph nodes **Thoraks**: Within normal limit **Pulmo:** Inspection: Symmectrical right and left; Palpation: Fremitus right = left; Percussion: Sonor; Auscultation: Vesicular, rhonchi (-/-), wheezing (-/-) **Cor:** Inspection: Ictus cordis is not visible Palpation: Ictus cordis is palpable one finger LMCS RIC V Percussion: Right border is LSD, left border is one finger medial of LMCS, top border is RIC II Auscultation: Regular rhythm, murmur and gallop are negative **Abdomen:** Inspection: Not distended; Palpation: Hepar and lien are not palpable; Percussion: tymphani; Auscultation: Bowel sounds is normal; **Extremities**: edema (-/-)
Dermatological Examination	Multiple erythematous macules on the face, hyperpigmented macules on the chest, abdomen, and right arm, hyperpigmented plaques on the legs, and a nodule on the back. Positive Darier's sign (swelling and redness induced by rubbing of the lesion after 2 minutes). No systemic involvement.
Laboratory evaluation	Hb: 11.4 gr/dl (N (Normal): 9.6 - 15.6 gr/dl) Leukocyte: 19.03 /mm^3 (N: 4000 - 12.000 /mm^3) Thrombocyte: 508.000 /mm^3 (N: 150.000 - 450.000 /mm^3) Hematocrit: 37 % (N: 34 - 48%) PT: 9.8 seconds (N: 9.70 - 12.93 seconds) APTT: 28.6 seconds (N: 21.41 - 28.81 seconds) SGOT: 43 u/l (N: < 33 u/l) SGPT: 20 u/l (N: < 43 u/l) Ureum: 26 mg/dl (N: 20-50 mg/dl) Creatinine: 0.3 mg/dL (N: 0.6-1.2 mg/dl) GDS: 93 mg/dL (N: 50-300 mg/dL) Absolute eosinophil: 1.332. **Conclusion**: Leucosytosis with eosinophilia, trombositosis.
Dermoscopy evaluation	Central pigment network and a light-brownish peripheral structure.
Histopathological evaluation	Histopathological features are suspicion of urticaria pigmentosa (cutaneous mastocytosis). Increased infiltration of mast cells in the dermis. **Conclusion**: Histopathological features are urticaria pigmentosa (cutaneous mastocytosis)
Diagnosis	Urticaria pigmentosa

Table 2. Treatment and follow up.

Treatment	Follow up
General Treatment: • Explained to the patient's parents regarding the disease (etiology of urticaria pigmentosa, prognosis, and complications). • Explained that the disease is benign and requires only symptomatic treatment. • Explained the possibility of spontaneous resolution before puberty. • Educated the parents about avoiding trigger factors such as certain drugs, food, local or systemic anesthetics, heat, and friction. • Educated the parents about the possibility of systemic involvement symptoms such as whole-body flushing, shortness of breath, and diarrhea. • Advised regular follow-up to detect systemic involvement. **Specific Treatment:** **Systemic Therapy:** • Cetirizine 2 mg once daily (powder). **Topical Therapy:** • Hydrocortisone 2.5% cream applied twice a day on red-brown patches.	**Anamnesis:** • No new reddish patches or bumps that feel itchy. • Patient had no fever. • No diarrhea, nausea, or vomiting. • The brownish patches reduced and became faded. **Physical examination:** • Pulse Rate: 100 x/minute • Respiratory Rate: 28 x/minute • Temperature: 36.5°C **Dermatology State:** • Location: Neck, face, chest, stomach, back, both arms, legs. • Distribution: Regional. • Shape and arrangement: Round-unspecific/symmetrical. • Border: Defined. • Size: Lenticular-plaque. • Efflorescence: Erythematous macules, hyperpigmentation macules (on chest, right arm, both thighs), hyperpigmentation plaques (on face and back). **Treatment:** • Cetirizine 2 mg once daily (powder). • Hydrocortisone 2.5% cream applied twice a day on red-brown patches.

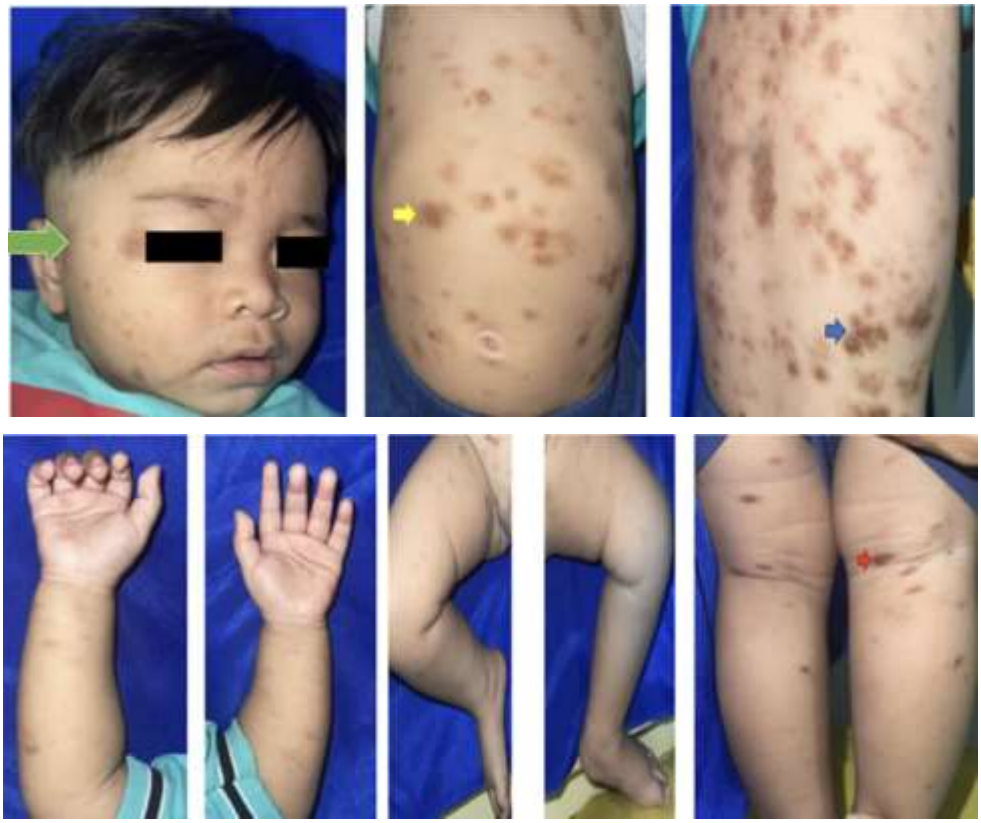

Figure 1. Erythematous macules (on face), hyperpigmented macules (on chest, abdomen, on right arm, both of thigh), hyperpigmented plaques (on legs), nodule (on back).

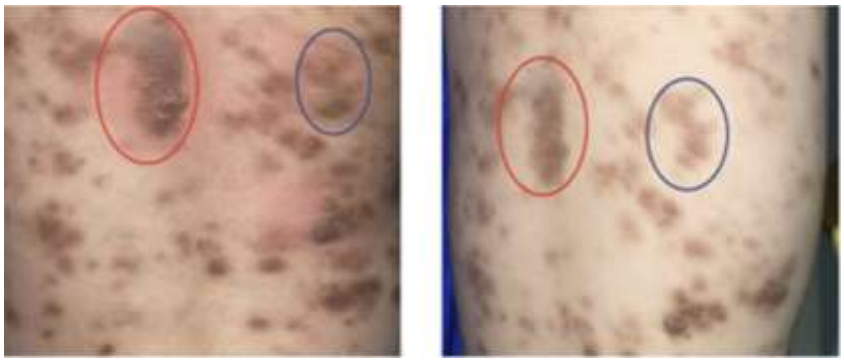

Figure 2. Swelling and redness induced by rubbing of the lesion within 2 minutes signifying a positive Darier's sign.

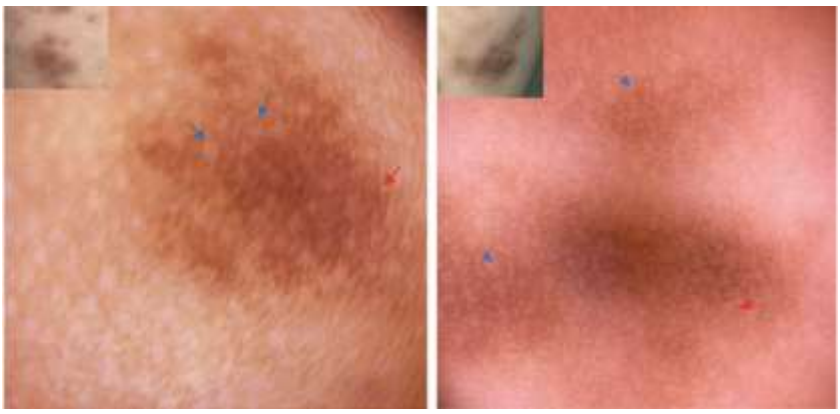

Figure 3. Central pigment network (blue arrows) and a light-brownish peripheral structure (red arrows).

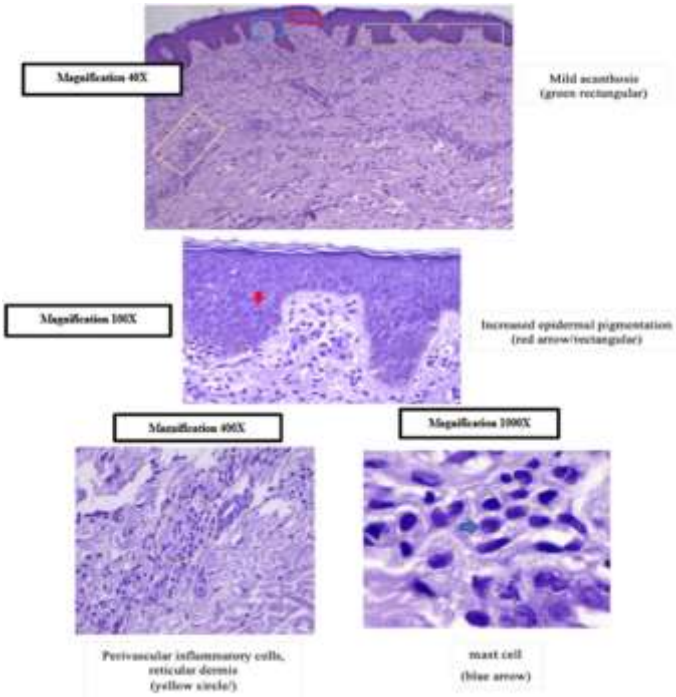

Figure 4. Histopathological examination with Giemsa. There were pieces of skin tissue with a epidermal skin with increased epidermal basal pigmentation, dermal papillae partially dilated, containing cells with oval to spindle-shaped nuclei, granular cytoplasm, resembling sal mast. Cells are scattered around the blood vessels in the papillary dermis and reticular dermis.

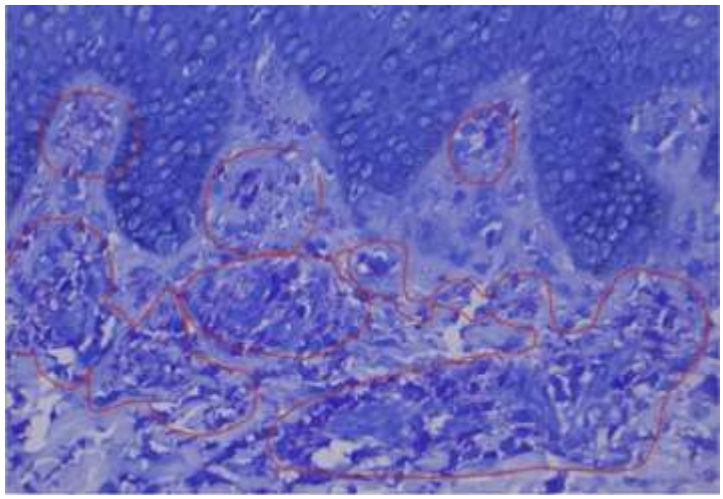

Figure 5. Histopathological examination with Ziehl Nielsen, increased infiltration diffuse mast cells in the dermis (red circle). Magnification 100x.

3. Discussion

The diagnosis of urticaria pigmentosa (UP) in this infant, as described in this case report, serves as an excellent example of the multifaceted approach required to accurately diagnose this condition, particularly in infants. It highlights how a systematic evaluation, integrating clinical findings, dermoscopy, and histopathology, can lead to a confident and timely diagnosis. This is particularly important in infants, where the presentation of UP can be varied and may mimic other common skin conditions. The diagnostic journey began with a thorough clinical evaluation, starting with eliciting a detailed history from the infant's mother. This step proved invaluable in understanding the timeline and evolution of the skin lesions. The mother's observation of the initial appearance of red bumps shortly after birth, a hallmark of UP, immediately raised suspicion. The subsequent transformation of these bumps into brownish patches further supported this initial impression. This emphasizes the importance of actively listening to parental concerns and obtaining a detailed history, as parents often provide crucial observations about the onset, progression, and characteristics of skin lesions. Next, a comprehensive physical examination was conducted. This involved a systematic assessment of the infant's entire skin surface, noting the morphology, distribution, and characteristics of the lesions. The presence of multiple hyperpigmented macules and plaques, in addition to a solitary nodule, all exhibiting the classic Darier's sign (urtication or wheal formation upon rubbing), significantly strengthened the suspicion of UP. The Darier's sign is a pathognomonic finding for mastocytosis, and its presence is a strong indicator of UP. Dermoscopy, a non-invasive technique using a handheld device to magnify and illuminate the skin, played a crucial role in refining the diagnostic process. It allowed for a more detailed visualization of the skin lesions, revealing subtle features that may not be apparent to the naked eye. In this case, dermoscopy revealed a central pigment network and a light brownish peripheral structure, both of which are characteristic dermoscopic features of UP. These findings, in conjunction with the clinical presentation and positive Darier's sign, significantly increased the confidence in the diagnosis of UP. Dermoscopy is increasingly recognized as a valuable tool in the evaluation of pigmented skin lesions, including those associated with UP. It can help differentiate UP from other skin conditions with similar clinical presentations, such as melanocytic nevi, lentigines, and café-au-lait macules. While clinical and dermoscopic findings strongly suggested UP, histopathological examination of a skin biopsy provided the definitive confirmation. This involved

obtaining a small tissue sample from one of the lesions and examining it under a microscope. The presence of an increased number of mast cells in the dermis, a defining feature of UP, unequivocally confirmed the diagnosis. This step is crucial not only for confirming the diagnosis but also for excluding other conditions that may mimic UP clinically or dermoscopically. Histopathology remains the gold standard for diagnosing UP and other forms of cutaneous mastocytosis. It allows for the visualization of the characteristic mast cell infiltrates and helps differentiate UP from other skin conditions with similar presentations, such as mast cell activation syndrome or other forms of cutaneous mastocytosis. This case vividly illustrates the importance of a comprehensive and integrated approach to diagnosing UP in infants. Relying solely on clinical presentation may lead to misdiagnosis or delayed diagnosis, as the presentation of UP in infants can be subtle and overlap with other common skin conditions. By combining clinical findings with dermoscopy and histopathology, clinicians can achieve a more accurate and timely diagnosis. Early diagnosis allows for the prompt initiation of appropriate management strategies, including symptomatic relief and parental education about potential triggers and complications. A definitive diagnosis allows for effective communication with parents, providing them with a clear understanding of the condition, its prognosis, and potential complications. This can alleviate parental anxiety and empower them to actively participate in their child's care. Although rare, UP can be associated with systemic mastocytosis, which involves mast cell infiltration in multiple organs. Early diagnosis enables close monitoring for signs and symptoms of systemic involvement, facilitating prompt intervention if necessary. While the clinical and dermoscopic findings in this case strongly pointed towards UP, it is crucial to consider other skin conditions that can mimic UP in infants. Transient Neonatal Pustular Melanosis (TNPM), this benign condition, more common in infants with darker skin tones, presents with pustules and hyperpigmented macules at birth. However, unlike UP, TNPM lesions typically resolve spontaneously within a few weeks without scarring. Infantile acne, this condition, characterized by erythematous papules and pustules, can occur in infants. However, it usually lacks the characteristic Darier's sign and the distinct dermoscopic features of UP. Miliaria, also known as "prickly heat," miliaria results from sweat gland obstruction and presents with small, erythematous papules and vesicles. It typically occurs in hot and humid conditions and lacks the hyperpigmentation and Darier's sign characteristic of UP. Eczema, this common inflammatory skin condition can present with erythema, scaling, and pruritus. However, it usually lacks the characteristic hyperpigmentation and Darier's sign of UP. Histopathological examination plays a crucial role in differentiating UP from these other conditions by confirming the presence of mast cell infiltration in the dermis.[11-13]

The remarkably early onset of urticaria pigmentosa (UP) in this infant, with lesions appearing a mere two days after birth, is a striking aspect of this case and carries profound clinical implications. While UP commonly manifests within the first two years of life, such an early presentation is quite unusual and underscores the critical importance of including UP in the differential diagnosis of cutaneous lesions even in newborns. Early recognition and accurate diagnosis are paramount, as they pave the way for prompt and appropriate management, potentially mitigating the disease burden and significantly improving the quality of life for affected infants and their families. Diagnosing UP in newborns presents unique challenges due to the diverse array of neonatal skin conditions that can mimic its presentation. Newborns frequently exhibit various transient skin eruptions, including erythema toxicum neonatorum (often referred to as "newborn rash"), transient neonatal pustular melanosis (characterized by pustules and hyperpigmented macules), and miliaria, commonly known as "prickly heat." These conditions often share overlapping clinical features with UP, such as erythematous macules, papules, and even pustules.

Differentiating UP from these benign, self-limiting conditions based solely on visual inspection can be exceedingly difficult, especially for clinicians who may be less familiar with the subtle nuances of neonatal dermatology. In this particular case, the initial presentation of red bumps on the infant's back shortly after birth could have easily been mistaken for one of the more prevalent neonatal skin conditions. However, the persistence and progression of the lesions, coupled with the development of new lesions on other areas of the body, raised a red flag and prompted a more thorough investigation. This underscores the critical importance of meticulous monitoring and close follow-up of any skin lesions in newborns, even those that initially appear benign or inconsequential. A comprehensive and detailed clinical history, encompassing the precise timeline of lesion development, any associated symptoms (such as itching or irritability), and any relevant family history, is indispensable. Moreover, a thorough and systematic physical examination, with particular attention to the morphology, distribution, and specific characteristics of the lesions, is essential. A hallmark clinical finding in UP is Darier's sign, which refers to the characteristic urtication or wheal formation that occurs upon rubbing or stroking the lesions. Eliciting Darier's sign requires a gentle but deliberate stroking or rubbing of the lesions, and it may not always be readily apparent. Therefore, maintaining a high index of suspicion for UP and employing a careful and methodical examination technique are crucial for detecting this pivotal clinical finding. Early-onset UP may have distinct clinical implications compared to UP that develops later in childhood. Emerging evidence suggests that early-onset UP may be associated with an increased risk of systemic involvement and a more persistent disease course. Systemic mastocytosis, characterized by the infiltration of mast cells in various organs beyond the skin, can manifest with a wide range of symptoms, including flushing, diarrhea, abdominal pain, bone pain, and even life-threatening anaphylaxis. While systemic involvement is relatively less common in children with UP, it represents a serious complication that necessitates prompt recognition and aggressive management. Although the infant in this case report did not exhibit any overt signs or symptoms suggestive of systemic involvement at the time of diagnosis, the remarkably early onset of his condition warrants vigilant monitoring and regular follow-up assessments. This proactive approach is vital to ensure the early detection and timely intervention should systemic involvement occur. Regular follow-up appointments should encompass a thorough review of the infant's symptoms, a comprehensive physical examination, and potentially laboratory investigations, such as a complete blood count with differential, liver function tests, and tryptase levels, to assess for any subtle indicators of systemic involvement. While the majority of children with UP experience spontaneous resolution of their skin lesions by adolescence, a subset of individuals may encounter persistent or recurrent symptoms that extend into adulthood. Consequently, long-term follow-up is of paramount importance to meticulously monitor disease progression, assess the response to therapeutic interventions, and provide ongoing support and education to families navigating the challenges of this chronic condition. Regular follow-up allows clinicians to diligently track the course of the disease, identify any new developments or potential complications, and judiciously adjust treatment strategies as needed. In this particular case, the infant demonstrated a favorable response to treatment with oral antihistamines (cetirizine) and topical corticosteroids (hydrocortisone), experiencing a significant regression of the lesions and a marked improvement in symptoms. However, continued follow-up is essential to ensure the long-term control of his symptoms and to remain vigilant for any insidious signs of systemic involvement. This may entail periodic clinical evaluations, dermoscopic examinations to assess lesional changes, and potentially repeat skin biopsies to evaluate disease activity at the histopathological level. Furthermore, long-term follow-up provides an invaluable

opportunity to educate and reassure parents about the natural history of UP, potential triggers that can exacerbate symptoms, and effective management strategies. Parents should be thoroughly counseled on how to identify and avoid potential triggers, such as certain medications (e.g., nonsteroidal anti-inflammatory drugs), specific foods (e.g., shellfish, nuts), and physical stimuli (e.g., friction, heat, cold) that can provoke mast cell degranulation and worsen symptoms. They should also be informed about the constellation of signs and symptoms that may herald systemic involvement and instructed to seek prompt medical attention should these occur.[14-16]

The management of urticaria pigmentosa (UP) in this infant, as detailed in this case report, exemplifies a patient-centered approach that prioritizes alleviating the troublesome symptoms and enhancing the child's overall quality of life. This approach is consistent with the current understanding of UP, which recognizes that while it is a chronic condition, its symptoms can often be effectively managed to minimize discomfort and optimize the well-being of affected individuals. The cornerstone of symptomatic treatment for UP is the use of oral antihistamines. These medications work by blocking the action of histamine, a chemical released by mast cells that plays a central role in the development of itching, flushing, and the formation of wheals, which are characteristic features of UP. Second-generation antihistamines, such as cetirizine, are generally favored due to their advantageous side effect profile, including a lower risk of sedation compared to first-generation antihistamines. In this case, the infant was prescribed cetirizine, which led to a significant reduction in itching and a noticeable improvement in his overall comfort. Topical corticosteroids, such as hydrocortisone, can be applied directly to the affected skin areas to provide localized anti-inflammatory and anti-pruritic effects. These medications help to reduce the redness, swelling, and itching associated with UP lesions. The infant in this case was treated with hydrocortisone cream, which further contributed to the resolution of his skin lesions and the alleviation of his symptoms. In addition to pharmacological interventions, a variety of non-pharmacological measures can be employed to effectively manage UP and enhance the quality of life for affected individuals. These measures encompass a holistic approach that addresses not only the physical symptoms but also the psychosocial impact of the condition. Identifying and diligently avoiding potential triggers that can exacerbate UP symptoms is of paramount importance. Common triggers include certain medications (such as non-steroidal anti-inflammatory drugs, which can provoke mast cell degranulation), specific foods (such as shellfish and nuts, which can elicit allergic reactions), physical stimuli (such as friction, heat, and cold, which can trigger mast cell activation), and emotional stress, which can modulate immune responses and influence disease activity. Educating parents and caregivers about these triggers and empowering them to minimize the child's exposure to them can significantly improve symptom control and reduce the frequency and severity of flares. Gentle and meticulous skin care practices can help to soothe and protect the affected skin, promoting healing and preventing further irritation. Using mild, fragrance-free cleansers and moisturizers can help to maintain skin hydration and preserve the skin's natural barrier function. Avoiding harsh soaps, excessively hot water, and vigorous rubbing or scratching can also minimize skin discomfort and reduce the risk of secondary infections. UP, particularly in its more extensive or severe forms, can have a profound impact on the quality of life of affected individuals and their families. The visible skin lesions, the unpredictable nature of symptoms, and the potential for systemic involvement can lead to emotional distress, social isolation, and anxiety. Providing comprehensive psychosocial support, including counseling, support groups, and educational resources, can help individuals and families cope with the challenges of living with UP, fostering resilience and promoting overall well-being. The prognosis of UP in children is generally considered favorable, with the majority of cases resolving spontaneously by adolescence. However, a subset of

individuals may experience persistent or recurrent symptoms that necessitate long-term management. The factors that influence the disease course and prognosis of UP are not fully elucidated, but they may include the age of onset, the extent and severity of skin involvement, the presence of systemic involvement, and the individual's genetic predisposition. Regular follow-up is essential for monitoring disease progression, assessing the response to treatment, and providing ongoing support and education to families. Follow-up visits allow clinicians to meticulously track the course of the disease, identify any new developments or potential complications, and adjust treatment strategies as needed. These visits also provide a valuable opportunity to reinforce trigger avoidance strategies, address any concerns or questions that families may have, and offer psychosocial support to help them navigate the challenges of living with UP. In this case, the infant's positive response to treatment and the generally favorable prognosis of UP in children are encouraging signs. However, continued follow-up is crucial to ensure the long-term control of his symptoms and to remain vigilant for any signs of systemic involvement. This may involve periodic clinical evaluations, dermoscopic examinations to assess lesional changes, and potentially repeat skin biopsies to evaluate disease activity at the histopathological level.[17,18]

Comprehensive parental education is not merely an adjunct to the medical management of urticaria pigmentosa (UP) in infants, but rather an indispensable cornerstone. Equipping parents with a thorough understanding of the disease, its natural history, potential triggers, available treatment options, and effective coping strategies empowers them to actively participate in their child's care, fostering a sense of control and agency in navigating this chronic condition. A clear and concise explanation of UP, its underlying causes, and its potential impact on their child is paramount. Parents need to understand that UP is a chronic condition characterized by an abnormal proliferation of mast cells in the skin. These mast cells, which are normally involved in immune responses, release histamine and other inflammatory mediators that trigger the characteristic symptoms of UP, such as itching, redness, swelling, and the formation of wheals (urticaria). Providing parents with this foundational knowledge helps them grasp the nature of their child's condition and dispel any misconceptions or fears they may have. Parents should be informed about the typical trajectory of UP, including the possibility of spontaneous remission, which is often observed in children. It is crucial to emphasize that the majority of children with UP have a favorable prognosis and that their condition is unlikely to significantly impact their long-term health or development. However, it is equally important to acknowledge the potential for persistent or recurrent symptoms in some cases and the rare possibility of systemic involvement, which requires vigilance and prompt medical attention. A comprehensive overview of the available treatment options for managing UP symptoms is essential. This includes educating parents about the appropriate use of medications, such as oral antihistamines (e.g., cetirizine) to control itching and topical corticosteroids (e.g., hydrocortisone) to reduce inflammation and skin irritation. Non-pharmacological measures, such as trigger avoidance and gentle skin care practices, should also be emphasized. Parents should be informed about the potential side effects of medications and when to seek medical advice if their child experiences any adverse reactions or worsening of symptoms. A critical aspect of parental education is identifying and understanding potential triggers that can exacerbate UP symptoms. Common triggers include certain medications (such as non-steroidal anti-inflammatory drugs, or NSAIDs), specific foods (such as shellfish and nuts, which are common allergens), physical stimuli (such as friction, heat, cold, and pressure), and emotional stress, which can modulate immune responses and influence disease activity. Parents should be equipped with the knowledge and skills to recognize these triggers and implement strategies to minimize their child's exposure, thereby reducing the frequency and severity

of UP flares. While systemic involvement is relatively rare in children with UP, it is essential for parents to be aware of this possibility. Systemic mastocytosis, characterized by the infiltration of mast cells in various organs beyond the skin, can manifest with a range of symptoms, including flushing, diarrhea, abdominal pain, bone pain, and even life-threatening anaphylaxis. Parents should be educated about these potential signs and symptoms and instructed to seek immediate medical attention if they suspect their child may be experiencing systemic involvement. UP can be a challenging condition for both children and their families, and the emotional and psychosocial impact should not be underestimated. Providing parents with effective coping strategies and psychosocial support can help them navigate the challenges, reduce stress, and improve their overall quality of life. This may involve connecting them with support groups where they can share experiences and learn from others facing similar challenges, providing access to educational resources and online communities, and offering counseling services to address any emotional distress or anxiety related to their child's condition. When parents have a clear understanding of the nature of UP, the rationale for treatment, and the potential benefits and risks of different interventions, they are more likely to adhere to the prescribed treatment plan. This can lead to better symptom control, reduced frequency and severity of flares, and improved outcomes for their child. Knowledge is a powerful antidote to fear and anxiety. By providing parents with comprehensive and accurate information about UP, clinicians can help alleviate their anxieties and empower them to take an active role in their child's care. Understanding the condition and its management can instill a sense of confidence and control, enabling parents to make informed decisions and advocate for their child's needs. Educating parents about potential complications, such as systemic involvement, equips them with the knowledge to recognize warning signs early on. This can facilitate prompt medical intervention, potentially preventing serious health issues and improving long-term outcomes. By understanding how to manage UP symptoms, avoid triggers, and implement effective coping strategies, parents can play a pivotal role in helping their child live a full and active life. This can enhance the child's overall quality of life, reduce the impact of the condition on their daily activities and social interactions, and foster a sense of normalcy and well-being.[19,20]

4. Conclusion

This case of early-onset urticaria pigmentosa (UP) in a 7-month-old infant underscores the importance of a comprehensive approach to diagnosis, incorporating clinical findings, dermoscopy, and histopathology. The early onset of lesions, characteristic morphology, positive Darier's sign, and histopathological confirmation of mast cell infiltration collectively ensured an accurate diagnosis. Effective management of UP in this infant involved a two-pronged approach parental education and symptomatic treatment. Educating parents about the disease, its natural history, potential triggers, and treatment options empowered them to actively participate in their child's care. Symptomatic treatment with antihistamines and topical corticosteroids effectively controlled the infant's symptoms and improved his quality of life. This case highlights the need for early diagnosis and appropriate management of UP in infants to alleviate symptoms and improve their quality of life. It also emphasizes the importance of close follow-up to monitor disease progression and detect any signs of systemic involvement. The favorable outcome in this case demonstrates the effectiveness of this combined approach in managing UP in infants.

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