

A Case Report of Idiopathic Eruptive Macular Pigmentation Disorder

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Abstract: Idiopathic eruptive macular pigmentation is a rare, etiology-unidentified benign acquired pigmentation disorder in clinical practice. It commonly occurs in children and adolescents, and is less common in adults. It is prone to being confused with and misdiagnosed as other pigmentation disorders. This article reports a case of a 34-year-old female patient who presented with progressive pigmentation on the trunk as the sole complaint. There was no clear trigger or history of preceding inflammatory disease. The skin lesions manifested as characteristic bluish-gray and brown patches, accompanied by mild itching and no other systemic discomfort. Our hospital, based on the patient's clinical manifestations and laboratory tests, excluded systemic, infectious, and drug-induced pigmentation disorders and made a clear diagnosis. After the patient was referred to a higher-level hospital, the diagnosis was confirmed to be consistent. Considering the characteristics of the disease, the patient voluntarily chose to discontinue treatment and opt for regular follow-up. This article combines authoritative standards and recent literature to analyze the diagnostic difficulties and special characteristics of adult cases, sort out the key points for differentiation, thereby improving the clinical standardized diagnosis and treatment level, reducing missed diagnoses of adult cases, and providing case references for the diagnosis and treatment of rare pigmentation disorders.

Keywords: Macular pigmentation disorder; Idiopathic; Eruptive

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1. Introduction

Idiopathic eruptive macular pigmentation (IEMP) is a rare clinical pigmentary disorder. It was first defined and reported by Degos *et al.* in 1978. Currently, it has five core diagnostic criteria in clinical practice ^[1]. There is no clear pre-existing skin inflammation, no special history of drug exposure, characteristic pigmented macules, typical histopathological changes, and a normal mast cell count. This disease is more common in children and adolescents under the age of 20, with the peak incidence in adolescence, and adult-onset cases account for less than 10% ^[2].

The pathogenesis of this disease is clear, mainly involving abnormal physiological functions of local melanocytes in the skin, which leads to increased melanin synthesis in the basal layer of the epidermis and the aggregation of a small number of pigment-destroying cells in the superficial layer of the dermis, without cell proliferation, malignancy, or persistent inflammatory damage^[3]. The main clinical manifestations are pigmented macules on the trunk and proximal parts of the limbs. There are no subjective symptoms, and only a few cases are accompanied by mild intermittent itching. The skin lesions progress slowly, have no risk of malignancy, and do not involve mucous membranes or internal organs. Due to the insufficient specificity of skin lesions, adult patients are prone to be confused with various pigmentary skin diseases^[4].

Skin microscopy and histopathological examination are the core diagnostic and differential diagnosis methods for this disease. Currently, there is no specific standardized treatment plan. Clinically, follow-up observation is the main approach, and symptomatic treatment is performed only for those with cosmetic needs. The diagnosis and treatment protocol of the adult female case reported in this article has high clinical reference value.

2. Case presentation

2.1. General information and clinical history

The patient is a 34-year-old female of Han ethnicity, a freelancer. On X month X year 2025, she visited the dermatology department due to “pigmentation spots on the trunk for 5 months, with aggravation and mild itching for 1 month.” Five months ago, without any clear cause (no exposure to sunlight, trauma, skin inflammation, history of special cosmetics or medication), scattered light brown and dark gray smooth macules appeared on the chest, abdomen, and waistback. There were no subjective symptoms, no desquamation or scabbing, and no related treatment was performed. Later, the number of lesions increased, the area expanded, some merged with each other, and the color deepened. One month before the visit, intermittent mild itching occurred at the lesion sites, without aggravation at night, without secondary damage such as blisters or exudation, and she came to the hospital for treatment.

The patient was previously in good health. She denied having liver, kidney, endocrine, or autoimmune diseases. She denied food and drug allergies. There was no history of special drug or toxic, photosensitive substance contact in the 3 months before the onset, and no irregular sleep patterns. Her personal history, menstrual history, and marital and childbearing history were normal. There was no history of pigmentation or hereditary skin diseases in the family. No genetic-related factors were considered for the time being.

2.2. Physical examination

After admission, the doctor conducted a comprehensive physical examination of the patient’s entire body system. The patient’s vital signs, including body temperature and blood pressure, were stable. The patient was conscious and in good mental condition. No enlarged superficial lymph nodes were found throughout the body, and no abnormalities were detected in the physical examinations of all systems. This ruled out the possibility that systemic diseases had affected the skin and caused pigmentation.

Special physical examination in the dermatology department: Multiple round and irregular-shaped bluish-gray and brown macules and patches were visible on the trunk, chest, abdomen, and waist. The size of the skin lesions varied, ranging from 0.5 to 3.0 cm. Some skin lesions merged into large areas, with unclear boundaries. The overall distribution was symmetrical and without regularity. The surfaces of all skin

lesions were smooth and flat, without secondary skin lesions such as scales. The texture of the skin lesions upon palpation was consistent with that of the surrounding normal skin, without infiltration, thickening, or hardening. The skin scratch test was negative. No similar pigmented skin lesions were found on the limbs, head and face, palms, soles, and mucous membranes (**Figure 1**).



Figure 1. Clinical gross skin lesion on the patient's back

2.3. Supplementary examination results

To make a clear diagnosis and conduct differential diagnosis, the doctor conducted comprehensive non-invasive imaging, histopathological, and serological examinations. The results were objective and clear. The dermoscopy showed typical skin lesions on the trunk, mainly consisting of uniform brown-gray and brown regular pigment networks, with local deep brown staining-like pigment deposition, and the pigment distribution was uniform; point-like, spherical, and linear superficial blood vessels could be seen, with a regular course, and no characteristic abnormal structures of pigmented nevus or malignant melanoma were observed (**Figure 2**).

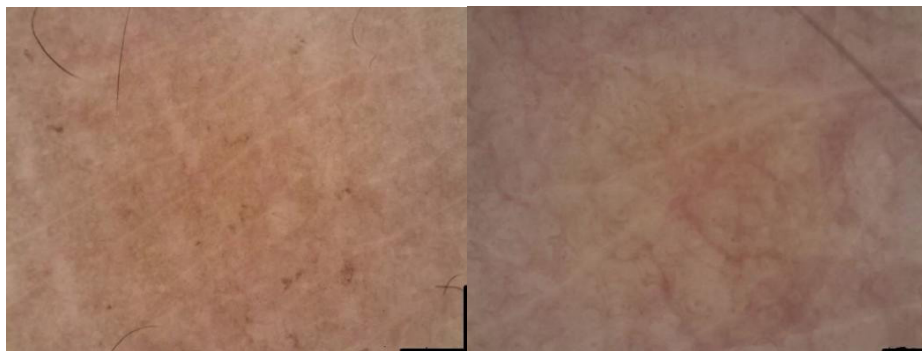


Figure 2. Dermoscopic manifestations of the skin lesion

Histopathological examination: After routine disinfection, tissue samples from typical skin lesions on the patient's trunk were taken for skin biopsy. After HE staining, microscopic observation was conducted. The structure of each layer of the epidermis was intact and clearly layered, with no excessive keratinization, incomplete keratinization, or hyphema changes; the uniformity of melanin granules in the basal layer of the epidermis increased, and there was no abnormal proliferation or aggregation of melanocytes. Scattered

pigment-infiltrating cells were observed in the dermal papillary layer and superficial layer, with uniform distribution and no large aggregations; a few scattered lymphocytes were observed infiltrating around the superficial dermal vessels, without band-like or lichenoid inflammatory infiltration, no liquefactive degeneration of basal cells, or interface dermatitis changes. No abnormal proliferation of mast cells was observed, no tumor cells or abnormal cells were found, and the collagen fiber structure of the dermis was normal (**Figure 3**).

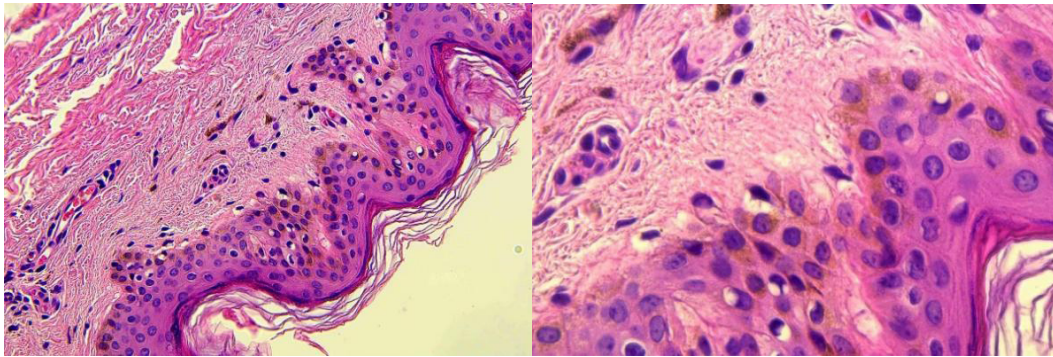


Figure 3. Histopathological examination of the lesion area under HE staining

Laboratory tests: Complete blood count, liver and kidney function tests, blood glucose test, thyroid function test, syphilis serological test, and human immunodeficiency virus antibody test were all conducted. All test indicators were within the normal reference range, completely ruling out skin pigmentation secondary to infectious diseases, liver and kidney metabolic abnormalities, endocrine diseases, autoimmune diseases, and other systemic diseases.

2.4. Clinical diagnosis and treatment process

In this study, the doctor combined the clinical manifestations of the patient's onset in adulthood, the characteristic imaging manifestations of regular pigmented networks and superficial regular blood vessels on dermoscopy, as well as the typical pathological changes such as increased basal layer pigmentation, superficial dermal infiltration of pigment-laden cells, and no interface dermatitis, along with laboratory tests to rule out various secondary pigmentation disorders. The preliminary diagnosis was made of idiopathic eruptive macular pigmentation syndrome.

The hospital doctor informed the patient in detail about the core conditions of this disease, such as the nature of the disease, its characteristics of onset, and the absence of malignancy risk. The doctor also informed the patient that there is no standardized and effective treatment plan for this disease, and the preferred intervention method in clinical practice is regular follow-up observation. For cosmetic needs, topical depigmenting agents and phototherapy can be selected as symptomatic measures, but the efficacy varies among individuals, and it cannot be completely cured. The patient voluntarily transferred to the dermatology department of a higher-level tertiary hospital for further diagnosis. The higher-level hospital, based on the complete clinical data of the patient, dermoscopy, and pathological reports, confirmed the diagnosis to be completely consistent with that of our hospital. After fully understanding the prognosis and current diagnosis of the disease, the patient, considering her own condition and wishes, voluntarily gave up all clinical intervention treatments and chose regular outpatient follow-up observation. Currently, the

patient's condition is stable, the skin lesions have not progressed further, and she is still under follow-up.

3. Discussion

3.1. Analysis of clinical manifestations and pathogenesis

The idiopathic eruptive macular pigmentation syndrome, based on the clinical research literature of this disease in the past decade both domestically and internationally, indicates that it is a sporadic disease worldwide, with more female patients, and adolescents are the absolute high-risk group. Cases of onset in adulthood are extremely rare^[5]. This patient, 34 years old, had onset in adulthood, which is a rare case of adult onset and shows a significant age difference from the classic adolescent-onset cases, being the core clinical feature of this case.

Currently, the academic community has a unified theory on the pathogenesis of this disease, with the core being local dysfunction of melanocytes and local microenvironment imbalance leading to melanin deposition, forming pigmented skin lesions, which are benign and reversible^[6,7]. Adolescent onset is often related to hormonal fluctuations, while adult onset mainly results from microenvironment imbalance in the skin. Most of these cases have no subjective symptoms, with only a few having mild itching. The skin lesions are commonly seen on the trunk and proximal limbs. The clinical manifestations of this patient fully conform to the core characteristics of this disease, with no triggering factors, a single lesion on the trunk, mild intermittent itching, progressive aggravation of the lesion, no systemic accompanying symptoms, and sufficient diagnostic basis and typical clinical features.

3.2. Diagnostic value of dermoscopy and histopathology

Dermoscopy examination, as an auxiliary diagnostic tool, is the core basis for clinical screening of this disease, effectively differentiating malignant pigmentation disorders^[8]. The microscopic appearance of this disease is mainly characterized by uniform pigmented networks, without pigment disorder, which can rule out melanoma and has a high specificity. The skin dermoscopy manifestations of this case are consistent with the literature characteristics, providing a basis for the initial diagnosis.

Histopathological examination is the gold standard for diagnosis of this disease and has absolute specificity. All diagnostic standards in domestic and foreign treatments uniformly stipulate the core pathological diagnostic points of this disease^[9]. Diagnostic points: increased basal layer pigmentation, superficial dermal infiltration of pigment-laden cells, no basilar cell liquefaction and degeneration, normal number of mast cells, and no tumor-like changes, which can distinguish it from other pigmentation diseases. The pathology of this case fully conforms to the above classic manifestations, without epidermal hyperplasia and other variations, excluding other diseases, supporting the final diagnosis.

3.3. Precise differential diagnosis analysis

Based on the skin lesion characteristics of this patient, the doctor strictly followed the diagnostic and treatment guidelines and conducted precise identification of all suspected pigmentation disorders one by one. All the identification points were based on authoritative dermatological diagnosis and treatment guidelines, without any subjective inference. Pigmented lichen planus: This disease belongs to inflammatory pigmented skin diseases. Most patients have clear precancerous inflammatory skin lesions of lichen planus, often accompanied by pigment damage of the oral mucosa. The pathological features are typical interface

dermatitis, liquefactive degeneration of basal cells, and band-like infiltration of lymphocytes in the superficial layer of the dermis. These characteristics are completely inconsistent with the clinical and pathological features of this patient, who has no history of inflammation, no mucosal damage, and no interface dermatitis. Therefore, it can be directly excluded.

Post-inflammatory hyperpigmentation: The prerequisite for this disease is a clear history of skin inflammatory conditions such as eczema, dermatitis, folliculitis, etc. The hyperpigmentation is strictly limited to the areas of the previous inflammatory lesions. The pathology only shows an increase in pigment in the basal layer, without aggregation of pigment-destroying cells in the superficial dermis. In this case, the patient had no pre-existing skin inflammatory history, and the distribution of the lesions did not follow the pattern of inflammation remnants. Pathologically, characteristic pigment-destroying cells could be observed, which can completely rule out this diagnosis.

Pigmented pityriasis rosea: This disease has a characteristic course of onset. In the early stage, multiple solitary mother lesions often appear. Subsequently, the skin lesions spread along the skin lines, and fine flaky scales can be seen on the surface of the lesions. Pathologically, acute inflammatory changes such as epidermal sponge edema can be observed. In this case, the patient had no mother lesions, no scales, and no pathological changes of epidermal inflammation, which allowed for a clear identification^[10].

Drug-induced pigmentation: This disease has a clear history of taking specific drugs. The appearance time of pigmentation is highly consistent with the medication period. After discontinuation of the medication, the pigmentation can slowly fade. In this case, the patient had no special medication history, and the onset was not related to the medication, which could be directly excluded. **Erythema multiforme:** This disease is more common in the face and neck area and is closely related to exposure to photosensitive substances and cosmetics. The skin lesions mainly present as reticular pigmentation and telangiectasia. Pathologically, there is epidermal atrophy and liquefaction of basal cells, which are significantly different from the lesion location, skin morphology, and pathological features of this case, allowing for thorough identification.

3.4. Treatment plan selection and prognosis evaluation

According to the *Chinese Guidelines for the Diagnosis and Treatment of Pigmentary Skin Diseases (2024 Edition)* and relevant international dermatological standards, idiopathic eruptive macular hyperpigmentation is a benign skin disease with no risk of malignancy and no systemic damage. The preferred clinical intervention strategy is regular clinical follow-up observation. No medication or surgical treatment is required. Clinically, only patients with a strong desire for cosmetic improvement are given the option to use topical hydroquinone creams, retinoid preparations, etc., for depigmentation, or undergo low-energy laser phototherapy. However, these symptomatic treatments can only slightly lighten the pigmentation and cannot completely cure the disease. There are individual differences in treatment efficacy, and there is no unified treatment standard.

After this patient was diagnosed, the doctor fully fulfilled the obligation to inform the patient, objectively and comprehensively explaining the benign prognosis of the disease, the limitations of the existing treatment methods, and informing the patient that there is no need for excessive treatment. Regular follow-up observation is sufficient. The patient, considering their own condition, recognized the benign prognosis of the disease and clearly refused all medication and physical treatments, choosing to undergo follow-up observation. This diagnosis and treatment decision conforms to the principles of standardized

diagnosis and treatment and avoids excessive medical care. Based on the follow-up data from the literature, the lesion regression cycle for patients with this disease is 1 to 3 years. The regression cycle for adult patients is slightly longer than that of adolescent patients, and the overall prognosis is good, without affecting the skin function and physical health of the patient.

3.5. Clinical value of the case and diagnostic insights

At present, the reports on idiopathic eruptive macular pigmentation disorders in both domestic and international literature mainly focus on cases in adolescents. The sample size of adult-onset cases is extremely small. Clinicians have insufficient knowledge of the diagnosis and treatment of adult cases, which easily leads to misdiagnosis. In this case, a 34-year-old adult female patient, with typical skin lesions, complete examination, and standardized diagnosis and treatment, has fully supplemented the clinical data of adult-onset cases and improved the clinical data of this disease at different age groups, thus having clear academic reference value.

Through the diagnosis and treatment of this case, clinical doctors have summarized clear clinical diagnosis and treatment inspirations: When diagnosing adult patients with unexplained pigmentation on the trunk, after excluding inflammatory, drug, and systemic disease causes, even if the patient is an adult-onset case, it is still necessary to include it in the differential diagnosis of idiopathic eruptive macular pigmentation disorder, and the disease should not be excluded due to the age factor; Skin microscopy combined with histopathological examination is an essential means for the diagnosis of this disease, and both are indispensable; In clinical diagnosis and treatment, strict adherence to the principles of benign skin disease diagnosis and treatment is required, avoiding excessive treatment, fully respecting the patient's treatment wishes, and achieving standardized and individualized diagnosis and treatment. At the same time, this case further confirms that there is no absolute age limit for the onset of this disease. Although adult-onset cases are rare, the clinical features and pathological standards are exactly the same as those of adolescent cases, and the diagnostic norms can be universally applied.

4. Conclusion

In summary, idiopathic macular pigmentation disorder is a rare benign pigmentation disorder with a favorable prognosis. It mainly affects adolescents and is relatively rare in adults. The disease is prone to misdiagnosis. The core clinical manifestations of this disorder are symmetrical and progressive grayish or brown pigmented macules on the trunk. It may be accompanied by mild intermittent itching, without systemic damage. Skin microscopy reveals characteristic regular pigment networks and superficial vascular structures. The tissue pathology has specific diagnostic criteria, which is the core basis for diagnosing this disease. This disease does not require special treatment. Regular follow-up observation is the preferred intervention plan. Symptomatic treatment is only applicable to patients with cosmetic needs. Clinicians need to broaden their diagnostic thinking. For adult patients with unexplained pigmentation on the trunk, timely complete skin microscopy and tissue pathology examinations should be conducted to accurately differentiate and diagnose the disease, avoiding misdiagnosis and excessive medical treatment. At the same time, continuous accumulation of adult-onset case data is needed to further improve the clinical diagnosis and treatment system and research on the pathogenesis of this disease.

Disclosure statement

The authors declare no conflict of interest.

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