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Case Report

Coexistence of pigmentary demarcation line type B and congenital vitiligo: A unique case report and exploration of potential association

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ABSTRACT

This case report describes a rare occurrence of coexisting Pigmentary Demarcation Line (PDL) type B and congenital vitiligo in a 4-year-old girl. The patient presented with linear band of hyperpigmentation along the blaschko line on the inner sides of bilateral thighs and lower abdomen, accompanied by depigmented patches on the knees, since birth. The clinical history, examination findings, and histopathological analysis supported the diagnosis. The case sheds light on a potential association between PDL and congenital vitiligo, which is not well-documented in the existing literature. Further research and long-term follow-up are recommended to investigate the underlying mechanisms and potential genetic predisposition, providing a deeper understanding of association between these conditions.

1. Introduction

Pigmentary Demarcation Lines (PDL) and congenital vitiligo are dermatological conditions characterized by distinct patterns of pigmentation alterations in the skin. PDL refers to lines of transition from hyperpigmented to hypopigmented or normal skin, while congenital vitiligo presents as depigmented patches on the skin since birth. This case report presents a rare occurrence of coexisting PDL type B and congenital vitiligo in a 4-year-old girl. This highlights a potential association between these conditions, emphasizing the need for further research and genetic evaluation to explore the underlying mechanisms and genetic predisposition.

2. Case Report

A 4-year-old girl, born through a non-consanguineous marriage with a normal perinatal phase, presented with linear pigmentation over the inner side of bilateral thighs and lower abdomen, accompanied by two depigmented patches over bilateral knees since birth. The mother reported that the extent of pigmentation increased proportionately with the growth of the child and there was a history of accentuation of pigmentation after sun exposure. Furthermore, a family history of a similar pattern of hyperpigmented and depigmented lesions was noted in her maternal uncle and one of her cousins (Figure 1).

Cutaneous examination revealed hyperpigmented macules arranged in a blaschkoid distribution over the anteromedial aspect of bilateral thighs, the lateral aspect of the left thigh, and the right lateral aspect of the abdomen. The hyperpigmented macules had well-defined lateral borders and ill-defined medial borders blending into the surrounding normal skin (Figure 2). Additionally, there were two well-defined depigmented macules sized between 3x3 cm and 5x6 cm, with irregular borders present over bilateral knees (Figure 2). These depigmented lesions did not exhibit accentuation under Woods lamp examination. Mucosal and systemic examinations were within normal limits. Histopathological examination

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Table 1: Summary of associations ofpigmentary demarcation line

Types	Site	Clinical association
A	Lateral aspects of upper anterior portion of the arms extending over the pectoral area	Heterochromia iridis and wooly hair [1]
B	Posteromedial portion of the lower limbs, extending from the perineum to the ankles	Congenita vitiligo (present case)
C	Hypopigmented lines in pre- and parasternal areas	Heterochromia iridis and wooly hair [1]
D	Posteromedial area of the spine	None
E	Bilateral hypopigmented bands on the chest in the zone between the mid-third of the clavicle and the periareolar skin	None
F	V-shaped patch on the lateral cheeks	Periorbital melanosis [2]
G	W-shaped patch on the lateral cheeks	None
H	Linear bands extending from the oral commissures to the lateral aspects of the chin	None

Table 2: Review of literature of congenital vitiligo

S.No.	Studies	Age	Race	Site of vitiligo	Associations	Family history
1.	Lerner and Nordlund [3] (3 cases) 1978	Not mentioned	Not mentioned	Not mentioned	Not mentioned	Not mentioned
2.	Chandra et al [4] 1992	2 months	Indian	Bilateral knees, right forearm, left hypochondrium	None	Positive in a third degree relative
3.	Jain et al [5] 1997	22 years/ F	Indian	Hands, feet	None	Positive in first- and second-degree relatives (AD)
4.	Kedward et al [6] 2008	71 years	-	Elbows, knees, hands, feet	None	Positive in first degree relatives
5.	Kambhampati et al [7] 2016	4 months/ M	Indian	Hands	None	Not present
6.	Barro et al [8] 2017	2 months/ F	African	Abdomen, chest, philtrum, left knee, both arms white forelock	Child born to HIV positive mother and father on ART	Not present
7.	Casey et al. [9] 2017	23/F	Caucasian	Neck, mons pubis, perianal region	None	Not present
8.	Pande et al. [10]	27days/ F	Indian	Generalised	None	Present in mother
9.	The present case	6month/ F	Indian	Knee	Pigmentary demarcation line	Present in maternal uncle and cousin.

of both the hyperpigmented and depigmented lesions revealed increased basal melanin and loss of melanocytes, respectively.

Based on the clinical history and examination findings, a diagnosis of Pigmentary Demarcation Line (PDL) (type B) with congenital vitiligo was considered. Genetic counselling was conducted, and the patient and her family were informed about the course of the disease. Treatment plans included Q-switched NdYAG laser therapy for the PDL lesions and the prescription of topical corticosteroids for the vitiligo lesions.

3. Discussion

PDLs were initially described by Matzumoto in the Japanese population in 1913 as lines of transition from hyperpigmented to hypopigmented or normal skin. [11] James et al. observed that PDLs tend to appear at an earlier age in black children and adults. [12] In the literature, there are eight types of PDLs (A-H), with PDL type B being the most prevalent in black female neonates, [12] consistent with the findings in the present case.

The etiology of PDLs is believed to involve genetic and hormonal influences, neurogenic inflammation, peripheral nerve compression, and pigmentary mosaicism along the

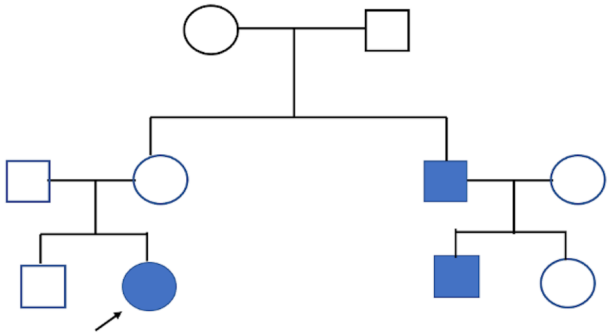

Fig. 1: Pedigree chart of a 4-year-old female with pigmentary demarcation line with congenital vitiligo

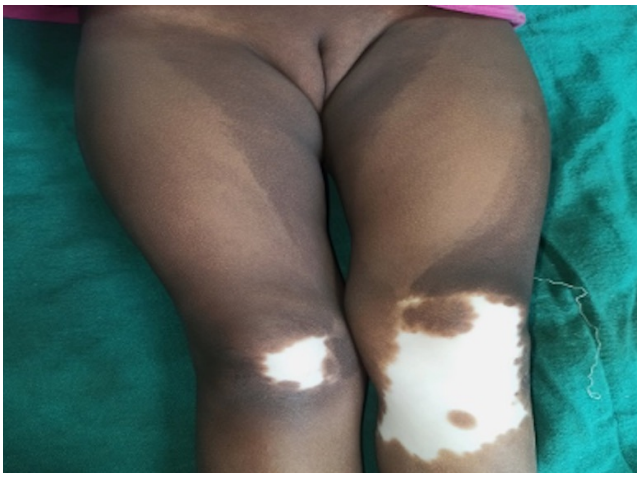

Fig. 2: Well-definedhyperpigmented macules located symmetrically over the anteromedial aspect of bilateral thigh and lateral surface of the left thigh with two well-defined depigmented macules over bilateral knees with irregular borders.

blaschko lines.[12] The linear distribution of lesions along the blaschko lines in our case further supports the hypothesis of pigmentary mosaicism.

While the association of PDLs with other dermatological conditions is uncommon, anecdotal reports have described links between PDLs and heterochromia iridis, woolly hair, and periorbital melanosis (Table 1).[1,2] Notably, there has been no documented association between congenital vitiligo and PDLs in the existing literature.

Congenital vitiligo itself is a rare condition with an unknown etiology.[3] However, considering the site, symmetry, and progression of the lesions in the absence of a white forelock and poliosis, the diagnosis of congenital vitiligo was favored in this case. Only eight case reports of congenital vitiligo have been published (Table 2), with four of them indicating a positive family history, suggesting a potential genetic predisposition.[4] However, the role of inheritance in the pathogenesis of PDLs and congenital vitiligo remains uncertain. Some individual reports have proposed an autosomal dominant pattern of inheritance in PDLs and congenital vitiligo.[5,13]

Further research is required to determine whether the coexistence of PDLs with congenital vitiligo in this case is coincidental or represents a true association. We recommend conducting a detailed genetic evaluation and implementing long-term follow-up to gain a deeper understanding of these conditions and their potential interplay.

4. Conclusion

This case report presents a unique coexistence of Pigmentary demarcation line (PDL) type B and congenital vitiligo in a 4-year-old girl. The combination of pigmentation along the line of blaschko suggestive of PDL and depigmented patches characteristic of vitiligo suggests a possible association between these conditions. Given the limited existing literature on the subject, further investigation is necessary to explore the underlying mechanisms and potential genetic predisposition. A comprehensive genetic evaluation and long-term follow-up are recommended to enhance our understanding of the relationship between PDL and congenital vitiligo.

5. Source of Funding

None.

6. Conflict of Interest

None.

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