ISSN-e 1984-8773

SOCIEDADE BRASILEIRA DE DERMATOLOGIA

Surgical & Cosmetic Dermatology

APOIO CIENTÍFICO:

www.surgicalcosmetic.org.br/

Piebaldism – portraits of hereditary character: a series of cases

Piebaldismo – retratos do caráter hereditário: uma série de casos

DOI: http://www.dx.doi.org/10.5935/scd1984-8773.2024160297

ABSTRACT

Piebaldism is a rare, autosomal dominant dyschromia characterized by circumscribed poliosis and triangular achromia in the frontal region in 90% of affected individuals, with no other systemic involvement. The incidence of dyschromia in this Dermatology Department, in a short space of time, and with plenty of images, motivated the presentation of this series of cases. We show two families with piebaldism, treated at the same Dermatology Department in 2021: the first family, with a mother and son presenting achromic macules on the trunk and poliosis in the frontal region since birth; the second family, with a grandmother, aunt, mother, and son showing the same characteristics described.

Keywords: Piebaldism; Pigmentation Disorders; Skin Diseases; Genetic

RESUMO

O piebaldismo é uma discromia rara, autossômica dominante, caracterizada por poliose circunscrita e acromia triangular na região frontal em 90% dos indivíduos acometidos, sem outros acometimentos sistêmicos. A incidência da discromia neste Serviço de Dermatologia, em curto espaço de tempo e com riqueza de imagens, motivou a exposição desta série de casos. Exibimos duas famílias com piebaldismo, atendidas no mesmo Serviço durante o ano de 2021: a primeira família, com mãe e filho apresentando máculas acrômicas pelo tronco e poliose na região frontal desde o nascimento; e a segunda família, com avó, tia, mãe e filho apresentando as mesmas características descritas.

Palavras-chave: *Piebaldismo; Transtornos da Pigmentação; Dermatopatias*

Letters to the Editor

Authors:

Melissa de Almeida Corrêa Alfredo[1]
Ingrid Rocha Meireles Holanda[1]
Ivanka Miranda Castro[1]
Gabriela Roncada Haddad[1]

[1] São Paulo State University (UNESP), School of Medicine, Department of Infectious Disease, Dermatology, Imaging Diagnosis and Radiotherapy, Botucatu (SP), Brazil.

Correspondence:
Gabriela Roncada Haddad
E-mail: gabriela.haddad@yahoo.com

Financial support: None.
Conflicts of interest: None.

Submitted on: 09/05/2023
Approved on: 10/04/2023

How to cite this article:
Alfredo MAC, Holanda IRM, Castro IM, Haddad GR. Piebaldism – portraits of hereditary character: a series of cases. Surg Cosmet Dermatol. 2023;15:e20230297.

INTRODUCTION

Piebaldism is a rare dyschromia characterized by circumscribed poliosis and triangular achromia in the frontal region in 90% of affected individuals. The incidence of dyschromia in the same Dermatology Department, within a short period of time and with plenty of images, motivated the presentation of this series of cases. We show two families with piebaldism, from different cities near the region, treated in the same department during 2021 (Figures 1 and 2).

Case series:

Case 1: One-year-old, male child. Referred from the BHU due to achromic macules on the body and poliosis in the frontal region since birth.

Mother with the same characteristics.

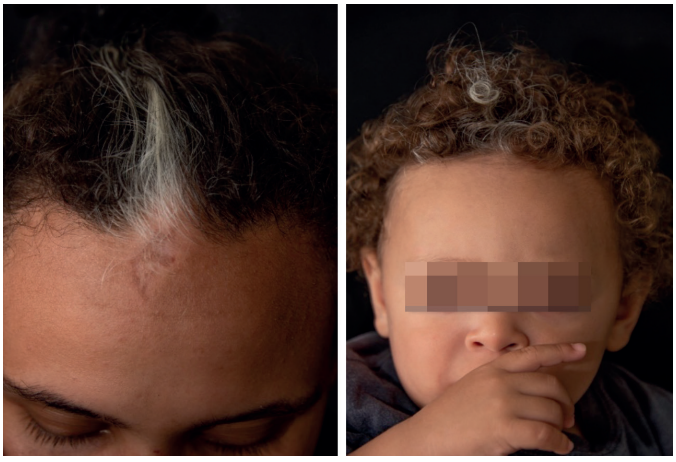

FIGURE 1: Poliosis in the frontal region of mother and child since birth

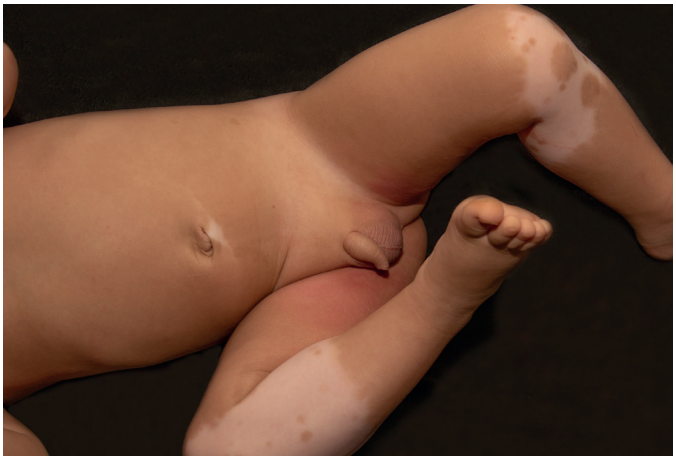

FIGURE 2: Achromic macules located bilaterally on the lower limbs

Case 2: Eight-month-old, male child. Referred from the Pediatrics Department due to achromic macules on the body and poliosis in the frontal region since birth (Figures 3 and 4).

Mother with the same characteristics. Maternal grandmother and aunt had the same phenotypic characteristics.

FIGURE 3: Poliosis in the frontal region of the maternal grandmother, mother, and son since birth

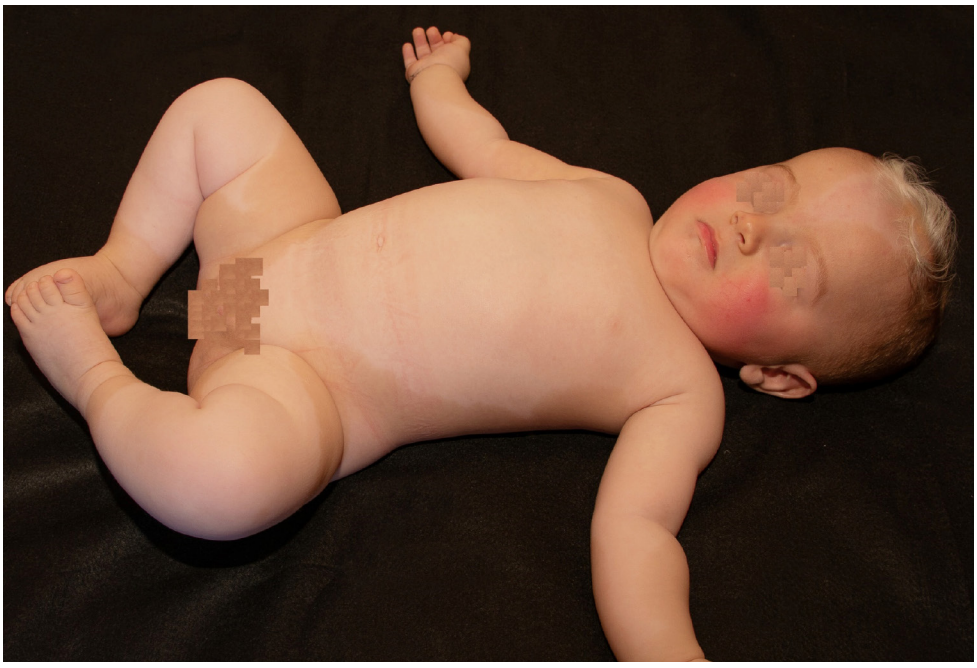

Figure 4: Achromic macules located bilaterally on the abdomen and lower limbs

DISCUSSION

Piebaldism is a rare dermatosis that affects around 1:20.000 individuals, regardless of sex or ethnicity. In 90% of cases, poliosis and areas of achromic skin are present, unrelated to systemic alterations.[1] This dyschromia has been described since Egyptian reports[2] and occurs due to mutations in the C-KIT 4q12 proto-oncogene, an autosomal dominant disorder, with abnormal migration of melanoblasts in the neural crest, resulting in body areas without melanocytic activity.[3]

Poliosis is typical of the disease, characterized by an area with no pigment in the frontal region of the scalp, which extends in a triangular shape to the forehead, converging towards the midline of the face. Achromic and hypochromic macules that affect the body are characteristically of a central pattern, located predominantly on the abdomen, the middle third of the upper and lower limbs, sparing the involvement of the hands, feet, and back.[1] The central pattern of dyschromia is explained because the mutation affects the neural crest. Affected individuals may present with hyperchromic macules and axillary ephelides.[3]

The diagnosis of piebaldism is clinical and, if it is suspected, a thorough clinical, ophthalmological, neurological, and gastrointestinal investigation is necessary, even in the neonatal period, to rule out differential diagnoses with possible systemic and deleterious symptoms, such as Waardenburg syndrome, which not only has achromic macules, but also iris heterochromia and sensorineural deafness.[1,3] Other differential diagnoses are vitiligo, Ito hypomelanosis, Wolf syndrome, and achromic nevus.[4]

As this is a benign condition, no treatment is required, but photoprotection guidelines are essential. Melanocyte transplants have been reported to reduce areas of achromia in patients with aesthetic complaints.[5]

CONCLUSION

Isolated piebaldism has no known systemic complications. As an autosomal dominant genodermatosis, it is necessary to question the family pattern when the diagnosis is suspected. ●

REFERENCES:

1. Shah M, Patton E, Zedek D. Piebaldism. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023. Available from: https://www.ncbi.nlm.nih.gov/books/NBK544238/
2. Huang A, Glick SA. Piebaldism in History—"The Zebra People." JAMA Dermatology. 2016; 152(11):1261.
3. Saleem MD. Biology of human melanocyte development, piebaldism, and waardenburg syndrome. Pediatr Dermatol. 2019;36(1):72-84.
4. Grob A, Grekin S. Piebaldism in children. Cutis. 2016 ;97(2):90-2.
5. Neves DR, Régis Júnior JR, Oliveira PJ, Zac RI, Silveira Kde S. Melanocyte transplant in piebaldism: case report. An Bras Dermatol. 2010;85(3):384-388.

AUTHOR'S CONTRIBUTION:

Melissa de Almeida Corrêa Alfredo ORCID 0000-0001-7423-4190
Approval of the final version of the manuscript; study design and planning; preparation and writing of the manuscript; data collection, analysis, and interpretation; active participation in research orientation; intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases; critical literature review; critical review of the manuscript.

Ingrid Rocha Meireles Holanda ORCID 0000-0002-2629-5244
Approval of the final version of the manuscript; study design and planning; preparation and writing of the manuscript; data collection, analysis, and interpretation; active participation in research orientation; intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases; critical literature review; critical review of the manuscript.

Ivanka Miranda Castro ORCID 0000-0002-3146-1892
Approval of the final version of the manuscript; study design and planning; preparation and writing of the manuscript; data collection, analysis, and interpretation; active participation in research orientation; intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases; critical literature review; critical review of the manuscript.

Gabriela Roncada Haddad ORCID 0000-0002-7516-9586
Approval of the final version of the manuscript; study design and planning; preparation and writing of the manuscript; data collection, analysis and interpretation; active participation in research orientation; intellectual participation in the propaedeutic and/or therapeutic conduct of the studied cases; critical literature review; critical review of the manuscript.