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(LETTER TO EDITOR)

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Fahr's syndrome with dermatological manifestation

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Abstract

Fahr's syndrome is a rare condition characterized by the presence of calcifications in the basal ganglia, typically secondary to a disorder of phosphocalcic metabolism, primarily hypoparathyroidism. It is generally challenging to clinically suspect as it can remain asymptomatic or manifest with diverse symptoms not corresponding to any specific clinical pattern. The dermatological expression of this syndrome is exceptional and is linked to hypoparathyroidism and hypocalcemia. In a hypocalcemic environment, keratinocytes exhibit significant proliferative activity, which may explain the pustular rash in our patients. Diagnosis relies on the presence of bilateral and symmetrical calcifications in the basal ganglia. Treatment involves calcium and vitamin D supplementation. Therefore, it is important to consider this syndrome in the presence of any aseptic pustulosis associated with disorders of phosphocalcic metabolism, particularly in cases with associated neuropsychiatric pathologies. We retrospectively report observations from six patients with Fahr's syndrome revealed through dermatological manifestations over a five-year period.

Keywords: Fahr's syndrome; Amicrobial pustulosis; Neuropsychiatric symptoms; Hypoparathyroidism; Central gray nuclei calcification

1. Introduction

Fahr's syndrome is a rare anatomoclinical entity characterized by the presence of bilateral and symmetrical intracerebral calcifications in the central gray nuclei. This condition is usually associated with disorders of phosphocalcic metabolism. However, dermatological expression is very rare.

2. Letter to the editor

Amicrobial pustulosis corresponds to a group of dermatoses that can be triggered by various factors, including hypocalcemia, and represents an exceptional cutaneous manifestation of Fahr's syndrome. We report 6 cases of amicrobial pustulosis revealing Fahr's syndrome.

This is a retrospective study collecting cases of Fahr's syndrome hospitalized in the dermatology department over a 5-year period, from January 2018 to January 2023.

A total of 6 patients were included (figure1,2), consisting of 5 women and 1 man. The average age of the patients was 36.66 years. A history of thyroidectomy was found in 2 cases. Neuropsychiatric disorders were present in all patients: epilepsy (4 cases), schizophrenia (1 case), behavioral disorders (1 case), and altered consciousness (1 case). All patients were admitted with a febrile generalized pustulosis. Skin biopsy revealed a non-specific subcorneal pustulosis in our patients. Bacteriological and mycological cultures from the pustules were sterile. Phosphocalcic balance was disturbed in all cases, showing hypocalcemia, hyperphosphatemia, and low PTH levels. Brain CT scans revealed bilateral calcifications in the central gray nuclei, confirming Fahr's syndrome. Treatment involved calcium and vitamin D

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supplementation, along with local care. The clinical outcome was marked by skin whitening concurrent with correction of calcium levels.

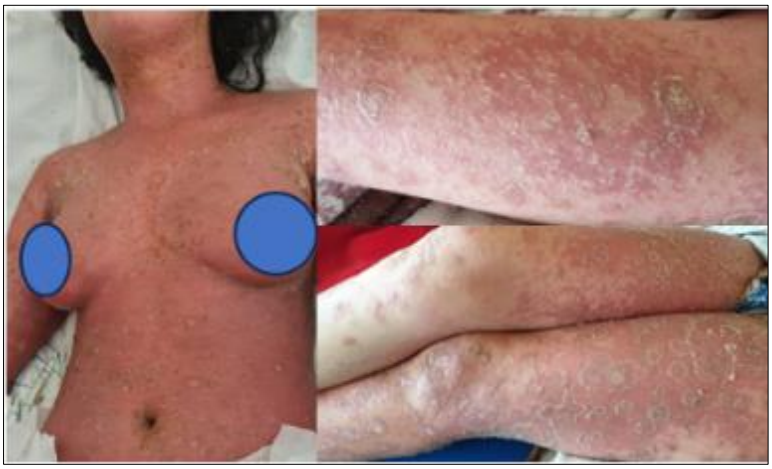

Figure 1 A 28-year-old female patient with schizophrenia past, was admitted with generalized febrile pustulosis

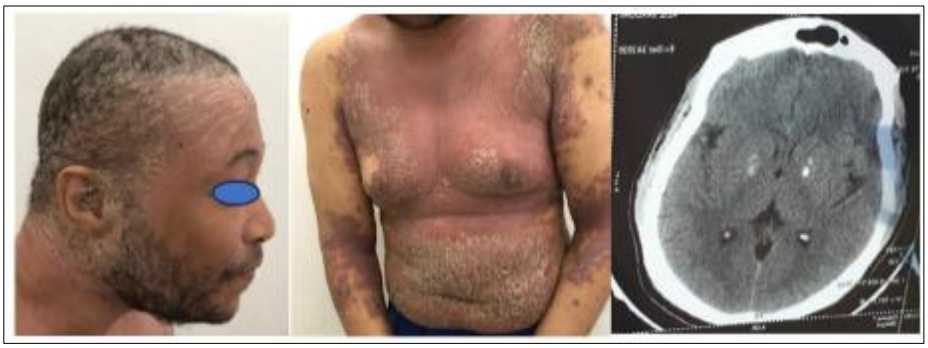

Figure 2 A 34-year-old patient treated for generalized tonic-clonic crises under phenobarbital and behavior disorders under neuroleptics for 14 years was admitted for erythematosquamous lesions with pustules evolving in the context of fever, with bilateral calcifications in the central gray nucleus

Fahr's syndrome, or bilateral striopallidodentate calcinosis, is a rare anatomoclinical entity characterized by bilateral and symmetrical intracerebral calcifications, typically in the central gray nuclei (1,2).

Fahr's triad is defined by the association of symmetrical calcifications of the central gray nuclei, neuropsychiatric symptoms, and hypoparathyroidism found in our patients (3).

The dermatological expression of this syndrome remains exceptional and is linked to hypoparathyroidism and hypocalcemia, as essential cadherins for keratinocyte adhesion are calcium-dependent molecules (4,5).

Diagnosis is challenging due to the nonspecific clinical polymorphism. In our patients, cutaneous manifestations in the form of pustulosis led to the discovery of idiopathic hypoparathyroidism, responsible for an unrecognized Fahr's syndrome.

Treatment relies on correcting phosphocalcic disorders, leading to a spectacular clinical improvement (6), emphasizing the importance of systematically investigating Fahr's syndrome in any amicrobial pustulosis associated with neuropsychiatric manifestations.

3. Conclusion

The interest of our work is to raise awareness among practitioners about a rare entity, which is Fahr's syndrome, as well as its exceptional dermatological expression. We encourage clinicians to consider it in the presence of any aseptic pustulosis associated with neuropsychiatric manifestations.

Compliance with ethical standards

Acknowledgments

The researchers acknowledge and appreciate the patients included in this case report.

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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