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Fahr Syndrome Secondary to Hypoparathyroidism with Dermatological Revelation

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ABSTRACT

Fahr syndrome is a rare anatomoclinical entity, with a polymorphic clinical presentation, skin involvement can be revealing. Vitamin D and calcium supplementation have contributed to the stabilization of the disease. Fahr's syndrome is likewise referred to as Fahr's disease, also referred to as idiopathic basal ganglia calcification. Fahr ailment is the mixture of encephalopathy and revolutionary calcification of the basal ganglia. Fahr's syndrome is familial and inherited, with autosomal dominant instances making up 60% of diagnoses. The gene that is accountable for Fahr syndrome has been mapped to chromosome 14. Fahr's ailment is neurological sickness characterized by means of odd calcified deposits in basal ganglia and cerebral cortex. We report a case.

Keywords: Syndrome de Fahr; Erythroderma; Rare; Fortuitous discovery

INTRODUCTION

Fahr syndrome is a rare neurodegenerative disorder determined by the presence of bilateral and symmetrical intracerebral calcifications of the basal ganglia. This condition is usually associated with phosphocalcic metabolism disorders, mainly secondary to hypoparathyroidism. It can be sporadic or familial. Chance discovery or during neuropsychic disorders, however, skin involvement can be revealing. The diagnostic confirmation exam of choice is the brain scanner. The prognosis for Fahr syndrome is good because the clinical and neuropsychic signs regress after the correction of the phosphocalcic disturbances. We report the case of a 38-year-old patient who presented with erythroderma revealing Fahr syndrome.

OBSERVATION

A 38-year-old patient, with no specific history, including no known mental retardation or psychiatric history, was hospitalized for the management of pruritic erythroderma, progressing for 1 month. Upon admission, the patient was confused and presented with a behavioral disorder, all evolving in a context of apyrexia and preservation of the general state. The dermatological examination showed a dry erythroderma covering 80% of the cutaneous surface (Figure 1), the remainder of the examination showed a shell of the scalp, the mucous membranes were free. Several diagnoses were proposed such as psoriasis, pemphigus foliaceous, taxidermy and chronic eczema. The psychiatric examination revealed a delirious systemic delusional syndrome, with significant confusion, without perceptual disturbances, the patient's judgment was slightly impaired. The day after her hospitalization, the patient installed pustules in the trunk and forearms. A complete biological assessment was carried out with the discovery of hypoalbuminemia at 26 g/l, corrected calcemia was reduced to 35.5 mg/l vitamin D deficiency at 9 ng/ml, hyperphosphatemia at 48 mg/l, a rate very low parathyroid hormone at 0.8 pg/ml, a collapsed TSH at 0.010 micro UI/ml, the rest were normal, in particular, the renal, hepatic balance and viral serologies. The patient was treated with a loading dose of calcium gluconate with a maintenance dose. Histological examination of a skin biopsy had shown an appearance compatible with pustular psoriasis. A cerebral CT was carried out objectifying bilateral and symmetrical intracerebral calcifications evocative of a syndrome of Fahr (Figure 2). The evolution was favorable after the correction of the phosphocalcic disorders associated with a symptomatic treatment and total improvement on the cutaneous level (Figure 3). The decline in six months. The erythroderma-like skin manifestation revealed idiopathic hypoparathyroidism, responsible for an unrecognized Fahr syndrome. The patient was referred for consultation endocrinology for additional support.

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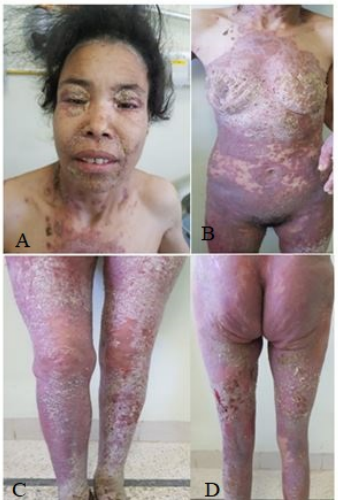

Figure 1: Erythroderma covering 80% of the cutaneous surface affecting the face (A); the trunk (B); the anterior (C) and the posterior (D) limbs.

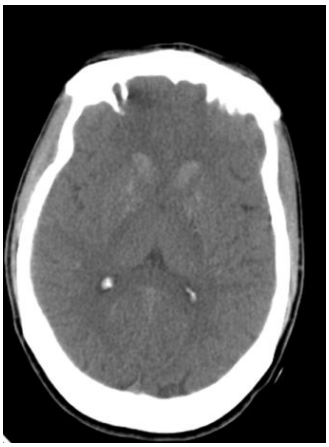

Figure 2: Bilateral calcifications of the central gray nuclei, the semi-oval center, and the cerebellum.

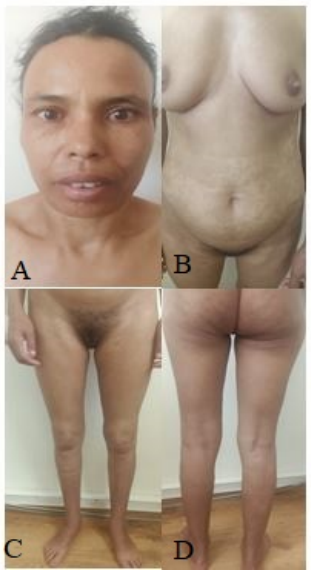

Figure 3: Total skin improvement at the level of the face (A); the trunk (B); the anterior (C) and the posterior (D) limbs.

DISCUSSION

Fahr syndrome is a rare entity whose causes are poorly understood. Its pathophysiology is not fully understood. In 1930, a German pathologist, Karl Theodor Fahr, described a case of a man with symmetrical calcifications of the basal ganglia and cerebral cortex [1]. Some reports describe the inheritance of Fahr syndrome, mainly in an autosomal dominant way [2]. However, in the majority of patients, the syndrome does not have a genetic background. Bilateral basal ganglia calcifications can be observed in disorders of calcium and phosphorus metabolism, especially in hypoparathyroidism. However, the frequency of their occurrence is low [3,4]. It is generally difficult to suspect because of its clinical polymorphism with a predominance of neuropsychiatric manifestations: behavioral disorders, confusional or delusional syndrome, and less usual cognitive disorders, intellectual deterioration, mental retardation, extrapyramidal attack, generalized or partial seizures, and more rarely pyramidal syndrome and intracranial hypertension [5,6] and the existence of dermatological lesions linked to parathyroid hormone deficiency [7]. Different cutaneous signs are attributed to the parathyroid deficit: scaly dry skin, eczematiform rashes, hyperkeratotic maculopapular rashes, pellagroid hyperpigmentations of Addisonian or melasma type, exfoliative erythroderma.

the data in the literature are very limited or even exceptional: an observation identical to ours was reported by Beurey et al. associating amicrobial pustulosis, hypoparathyroidism and comitiality [8]. Another recent observation of Fahr syndrome associated with extensive psoriasis has also been reported [9].

It is most often associated with dysparathyroidism: hypoparathyroidism, primary or postoperative, is the most classic anomaly associating hypocalcemia, hyperphosphoremia, hypocalciuria, hypophosphaturia and decrease in the serum parathormone level.

X-rays of the skull may show bilateral suprasellar opacities. Currently, the diagnosis is based on the brain scanner [7,10]. The evocative images are characterized by bilateral and symmetrical calcifications of the central gray nuclei and serrated nuclei of the cerebellum. In magnetic resonance imaging, calcifications appear as a hyposignal in T1 and T2 [7,11].

The peculiarity of our observation is its occurrence at an early age, the mode of onset was in the form of a cutaneous manifestation with a type of erythroderma which made it possible to discover idiopathic hypoparathyroidism responsible for an unknown Fahr syndrome.

Hypocalcemia seems to play a role in their training since early therapeutic management could prevent their constitution.

CONCLUSION

Skin involvement in Fahr syndrome can be revealing as in the case of our patient. The dermatological expression of Fahr syndrome is unknown. Our observation illustrates the important relationship between the hypoparathyroidism calcium vitamin D and skin axis, Indeed, the administration of calcium and vitamin D made simultaneously disappear the

dermatological and neurological problems in our observation. In front of phosphocalcic metabolism disorders, and in particular, in the case of associated neuropsychiatric or endocrine signs, intracerebral calcifications must be sought in order to detect Fahr syndrome and thus adopt the most appropriate therapeutic measures. In addition, new research should focus on the genetic study.

CONFLICTS OF INTEREST

Authors declare that there is no conflict of interest.

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