DERMATOPATHOLOGY

FAHR SYNDROME: A NEW OBSERVATION

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Background: Fahr syndrome is a rare anatomoclinic entity determined by the presence of bilateral and symmetrical intracerebral calcification of basal ganglia. This condition is usually associated with phosphocalcic metabolic disorders. We report a new observation of Fahr syndrome.

Observation: A 36-year-old woman with a history of epilepsy teated with phenobarbital and sodium valproate, a cataract surgery in her left eye,was hospitalized in our formation for aseptic pustulosis. The clinical examination revealed a dysmorphic syndrome with a facial hypertelorism, infiltrated skin, gingival hypertrophy, short and wide neck, pudgy fingers, and a stunting delay. She had also speech problems and mental retardation. Dermatological examination revealed pustular lesions in the neck, large folds and trunk. Biological assessment found deep hypocalcemia at 27mg / l, phosphorus at 47mg / l, parathyroid hormone at 36.4 pg / ml, hypothyroidism and hypogonadism. Computed tomography showed bilateral and symmetrical intracerebral calcifications. Cerebral magnetic resonance imaging showed hypersignals on T2-weighted sequences.The diagnosis of Fahr syndrome was retained. Calcium and vitaminotherapy D had allowed a favorable evolution.

Key message: Phosphocalcic metabolism disorders, particulary while associated with neurological or endocrine symptoms, should insit us to do imaging to look for intracerebral calcifications.