The Corpus Callosum Agenesis: A Case Report

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Abstract 
Agenesis of corpus callosum is the most frequent cerebral malformations. Diagnosis is based on brain imaging. It is most often asymptomatic. Hyperhidrosis can be part of his clinical picture. We present the case of a 49 year old patient, unemployed, with a history of frontal lobe epilepsy and extra pyramidal syndrome. He consulted for a left hemiparesis with sweating attacks. Physical examination showed facial dysmorphism, psychomotor retardation, hemiparesis predominantly left brachiocephalic and bilateral pyramidal syndrome. Brain imaging showed, in addition to cerebral ischemia, a corpus callosum agenesis. This observation helped to highlight a rare disease whose clinical presentation is nonspecific. This malformation is to seek at prenatal, postnatal and even at the adulthood. Hyperhidrosis attacks without obvious underlying pathology should suggest this diagnosis.

Keywords: corpus callosum agenesis, hyperhidrosis, brain malformations, epilepsy, brain imaging


1. Introduction

The corpus callosum (CC), the largest commissural in the brain, is essential for inter-hemispheric integration of sensory, motor, and higher-order cognitive information.

Corpus callosum agenesis (CCA) is the most frequent cerebral malformations. It can be symptomatic or asymptomatic. Diagnosis is based on imaging brain. Clinical signs are variable, they may include mental retardation, more or less moderate, seizures, “out control” behavior, facial dysmorphism and hyperhidrosis. Treatment is purely symptomatic.

We report the case of CCA revealed at adulthood by hyperhidrosis attacks.

2. Observation

A 49 year old man, unemployed, having the history of frontal lobe epilepsy, an extra-pyramidal syndrome consulted for a left hemiparesis installed three days ago with concept of sweating attacks. Physical examination objectified a broad forehead, hypertelorism, protruding tip of the nose, low-setears, psychomotor retardation, left hemiparesis and bilateral pyramidal syndrome. Brains canner objectified several old bilateral is chemicgaps, ischemia semi-recent at the territory of the right middle cerebral artery and total agenesis of the corpus callosum (Figure 1). We have not found other malformations. The karyotype was normal. Biological examination showed only hypercholesterolemia (8 mmol/ l) and hypertriglyceridemia (1.93 mmol/ l).The patient was treated with antiepileptics, antiplatelet agents and statins.

The evolution was marked by the disappearance of epileptic seizures, improvement of muscle strength but persistence of sweating attack.

3. Comments

Corpus callosum agenesis is a brain malformation characterized by the absence of the main inter hemispheric commissure. Development of the corpus callosum (CC) occurs during weeks 6–20 of gestation, and development defects, observed in 0.3%–0.7% of individuals undergoing brain imaging, are among the most common of brain malformations [1]. It must be differentiated from other malformations (Dandy Walker disorder gyration, microcephaly, holoprosencephaly ...), abnormality chromosomal, metabolic disease (pyruvate deficit dehydrogenase), viral embryofetopathy and a product outlet toxic (drugs, alcohol) or drug (valproate ...). In our case, the diagnosis was confirmed by brain scanner which eliminated all other diagnosis. Karioyte didn’t show abnormalities.

The CCA can be isolatedor belong malformation syndrome. Indeed, there are many syndromes with an "associated" CCA as Aicardisindrome, orofacial digital, Anderman, Shapiro. This latter includes hyperhidrosis attacks and hypothermia with CCA [2]. Our case associate also CCA and hyperhidrosis.

Hypothalamic thermostat disorderis the cause of hypothermia. The mechanism that lead to isolated agenesis of the corpus callosum still unknown. An anomaly of the commissural plate, defect migration callous axons or an abnormality of the corpus callosum neurons have been suggested [3]. Isolated CCA can be
sporadic, X-linked recessive, autosomal dominant or recessive [4]. If several chromosomes seem involved in the development and maintenance of the corpus callosum (1, 8, 13, 15, 18, 21, X ...), no gene has been identified for these isolated agenesis.

In Tunisia a genetic study was performed on 8 families has concluded to heterogeneity of genetic profile of subjects having a thin CC [5]. In 50% of cases, the CCA is isolated. It may be symptomatic or asymptomatic. Our patient has an isolated and symptomatic form. Indeed, we found no associated malformation. The main clinical signs was epilepsy. Cerebral ischemia was anepi-phenomenon. The patient had several cardiovascular risk factors. The mental retardation is usually detected at the time of schooling. The intelligence quotient (IQ) tests normal in 2/3 of cases, there may be a moderate to severe intellectual deficit in 1/3 of cases, there may be a moderate to severe intellectual deficit. The diagnosis of CCA was made on the occasion of seizures, psychomotor support, speech therapy, psychotherapy if psychomotor disorder [9]. Our patient remained stable for a long time thanks to anti-epileptics. The diagnosis of CCA was made on the occasion of hyperhidrosis attacks and cerebral ischemia.

4. Conclusion
The CCA is a common malformation. It may be symptomatic or revealed by a highly polymorphic and non-specific clinical feature. It should be researched in prenatal, postnatal and even in adulthood. Hyperhidrosis attacks without obvious underlying pathology should consider this diagnosis.

References