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Case Report

Agenesis of the Corpus Callosum and Neuropsychiatric Manifestations: About a Case Partial Anterior Agenesis and Review of Literature

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Abstract

Agenesis of the corpus callosum is a cerebral malformation frequently associated with neurological and psychiatric manifestations as well as with intellectual disorders. Partial agenesis concerns in almost all cases the posterior part and exceptionally the rostrum and the knee. Fetal ultrasound as well as MRI plays an important role in the positive diagnosis and detects associated malformations and brain anomaly.

We report in this paper a case of partial anterior agenesis of the corpus callosum, documented in our hospital by reviewing the data of the literature.

Keywords: *Corpus Callosum; Partial Agenesis; Epilepsy; Cognitive Impairment; Psychosis; MRI; Foetal Ultrasound*

Introduction

The corpus callosum is the largest connective structure in the brain. Its agenesis results from a defect in the development of interhemispheric axons [1]. It can be of primary or secondary cause, total or partial, the latter rarely involve the anterior part [2]. The repercussions of agenesis of the corpus callosum can alter the intellectual development of the child and lead to neurological and psychiatric manifestations.

Case Report

A 53-year-old man treated for a psychiatric disorder on neuroleptics for 15 years, presented with an abrupt onset right hemiparesis. A CT scan showed white matter hypodensities of different ages in the subcortical and periventricular hemispheres as well as in the brainstem.

MRI confirmed the presence of ischemic lesions, with trace of vascular leukopathy, the most recent of which in the left para-ventricular region corresponded clinically to the right hemiparesis (Figure 1). The study also showed partial agenesis of the rostrum, the knee and the anterior body of the corpus callosum (Figure 2); this partial agenesis of corpus callosum was most likely to be the cause of the psychiatric manifestations in our patient.

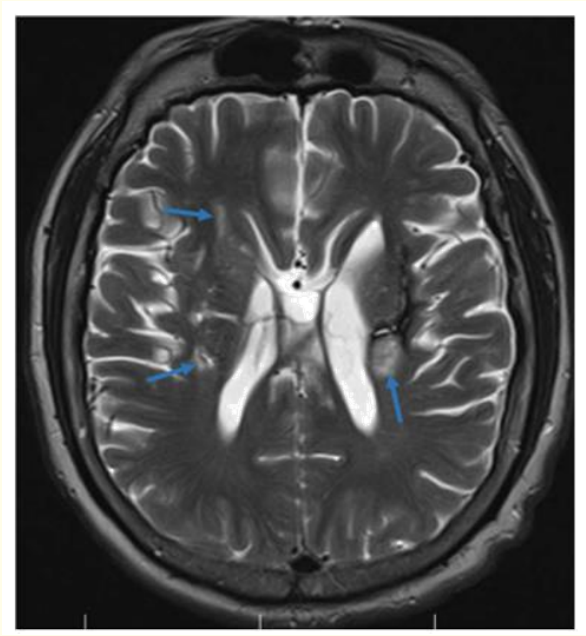

Figure 1: *Axial T2 weighted image on brain MRI showing nodular and punctiform hyperintense lesions in the white matter (⟹).*

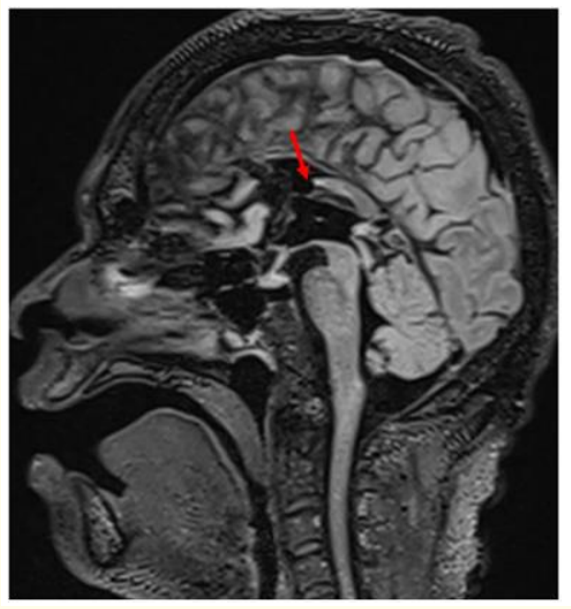

Figure 2: *Midsagittal T1 weighted image on brain MRI showing the absence of the rostrum, the genu and the anterior part of corpus callosum.*

Discussion

The incidence of agenesis of the corpus callosum is estimated at 1/4000 births and is increasing with the increase of use of MRI [1]. It is slightly predominant in males and can be discovered incidentally by imaging [3]; in fact, intellectual development can be considered normal upon discovery of agenesis of the corpus callosum, probably due to the formation of new nerve pathways [3].

The development in thickness and length of the nerve fibers of the corpus callosum follows a rostro-caudal direction, from the knee to the splenium. The rostrum forms last and fuses with the knee [2].

Agenesis of the corpus callosum can be primary where genetic abnormalities (demyelinating diseases, metabolic diseases) and microcephaly cause a defect in myelination and migration of nerve fibers, while secondary agenesis (which is rarer) is related, in addition to genetic abnormalities, to environmental factors (fetal alcohol syndrome, neonatal distress, severe neonatal hypoglycemia, HIV encephalopathy...) [4,14]. It can be isolated or associated with cerebral anomalies (interhemispheric cysts, hydrocephalus, Dandy-Walker malformations...) and be integrated in various syndromes (Aicardi, Andermann, Apert...) [13].

In children, agenesis is often primary and the diagnosis is made following intellectual impairment, epilepsy or psychiatric diseases; some studies have reported partial agenesis of the corpus callosum in early schizophrenia [5]. Epilepsy does not seem to be a direct consequence of partial or total agenesis but is a frequent circumstance of discovery in children [3,13].

In adults, agenesis of the corpus callosum is most often discovered following a psychotic episode or in the setting of depression or dementia [3]. The studies of Arnone., *et al*. have shown hypotrophy of the corpus callosum in patients suffering from bipolar disorder or schizophrenia and have suggested that the defect in the transmission of information between the two cerebral hemispheres in these patients may be responsible for the psychiatric manifestations [7].

In MRI, different evaluation methods have been proposed for the measurement of the corpus callosum, which should be evaluated by the thickness or volume of each part on a strict midsagittal plane. Anomalies of the corpus callosum are divided into three types: hypoplasia (total or partial), dysplasia (with or without hypoplasia) and total agenesis (Figure 3); in this subdivision, partial agenesis corresponds to the apple core appearance (hypoplasia of the corpus and agenesis of the splenium) or to an anterior remnant vestige of the corpus callosum (agenesis of the corpus and the splenium) [15]. The rostro-caudal developmental direction of the corpus callosum explains the predominance of hypoplasia on the posterior part [4].

MRI also looks for associated brain abnormalities: interhemispheric cysts and lipomas may be present at the time of diagnosis [16]. Developmental abnormalities of the anterior white commissure, fornix, and optic chiasma as well as polymicrogyria are frequently associated on imaging [2,14].

In the fetus, ultrasound (performed after the 20th week of amenorrhea) may suspect total agenesis: in transverse section it shows the absence of the septum pellucidum, the presence of colpoclephaly and an enlargement of the interhemispheric space. In coronal section, the 3rd ventricle is high situated and the occipital horns of the lateral ventricles make a bull horn appearance. In midsagittal section, the corpus callosum and the pericallosal artery are not visualized [14]. These abnormalities may be encountered to a lesser degree in partial agenesis, and only a broad and short aspect of the interhemispheric sulcus may suspect agenesis [14]. These signs are not specific and a fetal MRI should complement the ultrasound scan as it allows the diagnosis to be made with greater sensitivity [11,12].

Anterior partial agenesis has been rarely reported in the literature. Studies have shown that the existence of an extrinsic factor, such as extrinsic compression or ischemia of the corpus callosum, could be the cause of anterior partial agenesis [2]. Schaefer., *et al*. reported two cases of anterior partial agenesis of the corpus callosum on MRI in which post-mortem anatomical studies revealed their presence in an ectopic position by fusion of the frontal lobes on the midline [2].

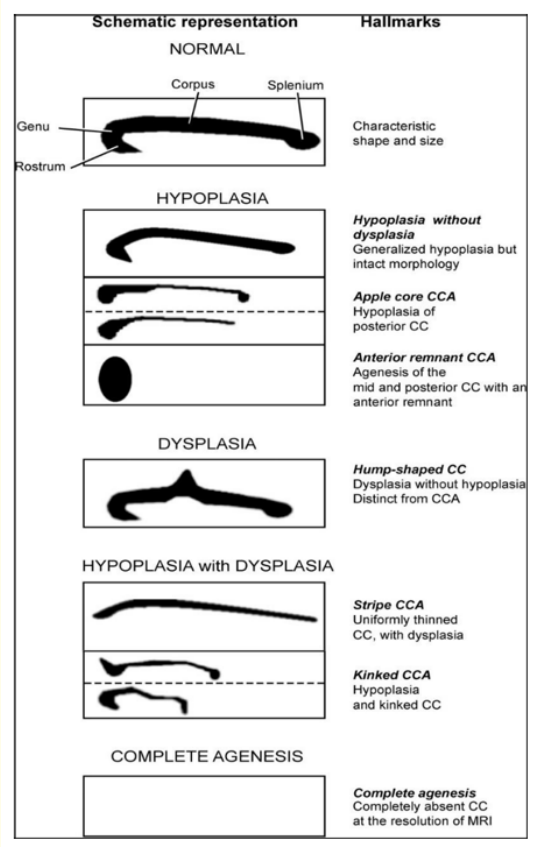

Figure 3: *Schematic representation of morphological anomalies of the corpus callosum [15].*

In our case, MRI did not reveal an obvious cause, namely the presence of a cerebral mass syndrome. The clinical history of psychiatric disease since the age of 38 years in our patient does not seem to be a cause of this agenesis: in fact, partial agenesis of the corpus callosum encountered in schizophrenia [5], depression [6], or bipolar disorders [7] do not have a regional predominance. Finally, the hypothesis of an ischemic origin of this anterior agenesis in our patient cannot be eliminated and remains probable: Lyoo., *et al*. and Yamanouchi., *et al*. have shown an atrophy of the corpus callosum in patients suffering from vascular dementia; in this case, the damage occurs preferentially in the anterior part of the corpus callosum [8-10]. The prognosis of agenesis of the corpus callosum is poor when associated with cerebral malformations or genetic abnormalities, while it remains favorable if the agenesis is isolated [13,14].

Conclusion

Partial anterior agenesis of the corpus callosum occurs exceptionally and its presence should lead to search for local destruction of nerve fibers by a mass or an infarct. Neurological and psychiatric manifestations are frequent in this condition and can be revealing.

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