ANTERIOR (ATYPICAL) CALLOSAL ABSENCE DUE TO CORTICAL DYSPLASIA AND SCHIZENCEPHALY

R. NURI SENER

Department of Radiology, Ege University Hospital, Bornova, Izmir, 35100, Turkey

SUMMARY

The corpus callosum develops from anterior to posterior, starting at the commissural plate of the lamina terminalis. We report a patient with cortical dysplasia and schizencephaly, which apparently interfered with the normal callosal development before 20 gestational weeks. The result was an atypical callosal dysgenesis in which the anterior parts (including anterior body, genu and rostrum) were absent while the remaining parts were developed. This finding suggests that the commissural plate may not be the only region where the corpus callosum starts to develop in some congenital brain malformations.


INTRODUCTION

The current theories suggest that the corpus callosum normally develops in an anterior to posterior direction, starting from the region called the commissural plate, and its genu forms first followed by posterior growth to form the body and splenium. This process is completed by approximately 20 weeks gestational age [1-4]. However, a recent report dealt with a different type of callosal dysgenesis in a patient with lobar holoprosencephaly, in whom there was anterior callosal absence while the remaining parts were developed, representing a different condition in contrast to current theories on callosal development [5]. In this communication we report another example of anterior callosal dysgenesis in a patient with cortical dysplasia and schizencephaly, and suggest that the commissural plate may not be the only region the corpus callosum starts to develop.

CASE REPORT

The present patient is a 30-year-old man with an epilepsy disorder (generalized seizures) since childhood. He was mentally normal except for occasional psychotic reactions. EEG studies revealed abnormal activities in the frontal lobes. An MR imaging examination was requested. This revealed striking findings. The posterior body and splenium of the corpus callosum were apparently developed, while the anterior body, genu, and rostrum were absent (fig. 1a, b). The anterior interhemispheric fissure extended to the lateral ventricles, and was surrounded by thickened cortices (cortical dysplasia) affecting the gyri cinguli (fig. 1c). In addition, there was a large schizencephalic cleft in the right frontal region, which was outlined by abnormal gray matter (fig. 1d).
Fig. 1a, and b. – Midsagittal (a), and left parasagittal (b) T1-weighted MR images show absence of the anterior body and genu of the corpus callosum (a), and resultant incomplete inversion of the cingulate gyri (b). The posterior body and splenium of the corpus callosum are developed (a). Note a large cleft connecting to the lateral ventricle (see fig. d).

Fig. 1c. – Axial, proton density-weighted MR image shows extension of the anterior interhemispheric fissure to the lateral ventricles, and the surrounding thickened cortices (cortical dysplasia) affecting the gyri cinguli.

Fig. 1d. – Coronal, T2-weighted MR image shows the large schizencephalic cleft in the right frontal region, which is outlined by abnormal gray matter, distinguishing it from a porencephalic cavity.

**DISCUSSION**

The corpus callosum normally starts to develop from the region called the comissural plate of the lamina terminalis at approximately 7 weeks gestational age. The growth is in an anterior to posterior direction; its genu forms first followed by posterior growth to form the body and splenium. This process is completed by approximately 20 weeks gestational age. The exception to this orderly anteroposterior development is the rostrum (most anterior small region) which forms latest of all. The corpus callosum may be totally absent due to agenesis of the comissural plate or due to persistence of the meninx primitiva in the comissural plate. When the earlier-formed segments (genu, body) are present, and the
later-formed segments (splenium-rostrum) are absent, the condition reflects callosal dysgenesis or partial absence. Besides this, callosal dysgenesis may manifest as diffuse thinning (hypoplasia) of the organ. Callosal agenesis or dysgenesis may be isolated or may be seen in a wide range of disorders including Chiari II malformation, Dandy-Walker malformation, sphenoethmoidal cephaloceles, septo-optic dysplasia, cleft lip and plate, acrocephalo-syndactyly, hypertelorism, coloboma, neuronal migrational disorders, dysmyelinating diseases, anomalies of the limbic system, and some others [1-4]. Recently, Barkovich and Quint [6] described a rare form of holoprosencephaly associated with midline interhemispheric fusion resulting in an atypical calloso dysgenesis: the genu and the splenium were present while the body was absent [6]. Following their observations, a report by Sener et al. [7] appeared dealing with similar brain changes in holoprosencephaly. This type of callosal dysgenesis (midline agenesis, and interhemispheric fusion) was attributed to faulty mesenchyme formation associated with maldevelopment of the interhemispheric fissure. Another recent report, documented anterior callosal absence (while the remaining parts were developed), again in a patient with holoprosencephaly [5]. Also, recent reports by Rubinstein et al. [8], and Kier and Truwitt [9] dealt with atypical types of callosal development. The current patient did not have holoprosencephaly, and the atypical callosal dysgenesis (absence of the anterior parts until the midbody) was associated with cortical dysplasia around the anterior interhemispheric fissure, and a large schizencephalic cleft was present in the right frontal region (fig. 1).

On the other hand, schizencephaly, and cortical dysplasia are neuronal migrational disorders. Schizencephaly refers to a full-thickness cleft (from ventricle to pia) in the cerebral hemisphere that is lined with polymicrogyric gray matter [10]. Cortical dysplasia consists of focal or diffuse cortical thickening (polymicrogyric cortex), and infoldings [11, 12]. Schizencephalic clefts may be unilateral or bilateral, and they most commonly are located near the precentral or postcentral gyri, a similar location for cortical dysplasias [1]. According to Barkovich and Kjos [10], schizencephaly is an extreme variant of cortical dysplasia, in which the infolding of cortex extends all the way into the lateral ventricle [10]. Van der Knaap and Valk [4] cited that the intrauterine time of onset for schizencephaly can be pinpointed approximately to 8 weeks, which is well before the development of the corpus callosum is completed (approximately 20 weeks). In addition, the time of onset for polymicrogyria is at approximately 20 weeks of gestation, a similar timing for completion of callosal development [4]. These data convincingly suggest that, in our patient the dysplastic cortex surrounding the anterior interhemispheric fissure as well as the large schizencephalic cleft affecting the right frontal region apparently interfered with the callosal development, which should normally start at the lamina terminalis (comissural plate). The result was an atypical callosal dysgenesis in which the anterior parts (including anterior body, genu and rostrum) was absent while the remaining parts were developed. This may suggest the comissural plate may not be the only region, where the corpus callosum starts to develop, at least in some of the congenital brain malformations.

REFERENCES