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CASE REPORT

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Corpus callosum dysgenesis: a report of four different cases

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Summary

Background: Callosal anomalies account for approximately 2% of all CNS congenital malformations. Dysgenesis of the corpus callosum (complete or partial agenesis) can be an isolated CNS malformation or coexist with other pathologies.

Case report: In all four cases corpus callosum dysgenesis was revealed by MRI (1.5T). The presented cases show examples of wide spectrum of all callosal anomalies – complete agenesis in case 1 and different sort of partial agenesis in cases 2, 3 and 4. In all the presented cases the callosal defect was found incidentally.

Conclusions: Diagnostics of the corpus callosum malformations is based on ultrasound scans (pre- and postnatal), CT and most of all, the MR. Except for major callosal defects described in our report, MRI quite often reveals tiny anomalies of callosal shape. Corpus callosum defects can not only be congenital, but also caused by acquired disorders (traumatic, vascular, inflammatory and neoplastic).

Key words: corpus callosum • dysgenesis • MRI

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Background

Corpus callosum develops from the posterior part of the primitive lamina terminalis. This process begins with eight weeks' gestation, with development of callosal precursors and fibres from the isocortical plates of the hemispheric vesicles. The genu of corpus callosum is formed approximately in the 11th week, and then in chronological order the body (trunk), splenium and rostrum appear until 18–20th week of gestation [1].

Anomalies of the corpus callosum are relatively frequent – about 2% of all CNS (central nervous system) congenital malformations. Dysgenesis of the corpus callosum can be divided into two groups: total callosal defect – complete agenesis (case 1) or different partial callosal defects – partial agenesis (hypogenesis, hypoplasia) (cases 2, 3 and 4) [1, 2].

Jinkins and co-authors divide callosal anomalies into three types:

- type I – callosal agenesis (case 1),
- type II – absence of the posterior part of corpus callosum caused by the presence of lipoma or an unknown reason (case 2),
- type III – corpus callosum hypoplasia, which is accompanied by other brain malformations (case 3) [3].

Dysgenesis of the corpus callosum may be an isolated CNS malformation, may coexist with telencephalic dysgenesis (eg. cortical dysplasia, periventricular heterotopia), can be associated with genetic syndrome (Aicardi, Mowat-Wilson, Nijmegen) or even posterior fossa malformations (Dandy-Walker syndrome, Chiari malformations, arachnoid cyst). Other possible accompanying abnormalities are the Probst bundles (thickened bundles of white matter parallel to the lateral ventricles) and midline tumours (interhemispheral cyst lipoma, meningioma) [4, 5].

In patients, who underwent surgical callosal section, "split-brain" syndromes are observed. However, in most cases those deficits do not occur in patients with congenital malformations of the corpus callosum. The most common clinical manifestations of dysgenesis is a mild mental delay (school related difficulties, behavioral problems), neuropsychological deficits related to tasks requiring interhemispheric communication; obsessive-compulsive disorders were also reported. Epilepsy inducted by coexisting grey matter heterotopia may occur. However, in general, corpus callosum dysgenesis is asymptomatic, if not associated with other anomalies.

Case 1

A 21-year-old man, complaining of headaches for two months, with no neurological deficits – medical history irrelevant. Psychological examination disclosed no mental delay, however, the patient admitted having school difficulties. MR scans revealed complete callosal agenesis with dilatation of the third ventricle (Fig. 1) accompanied by asymmetry of frontal lobes, mild focal pachygyria of left frontal lobe and dilated posterior horns of lateral ventricles (Fig. 2) – type I of callosal anomaly according to Jinkins.

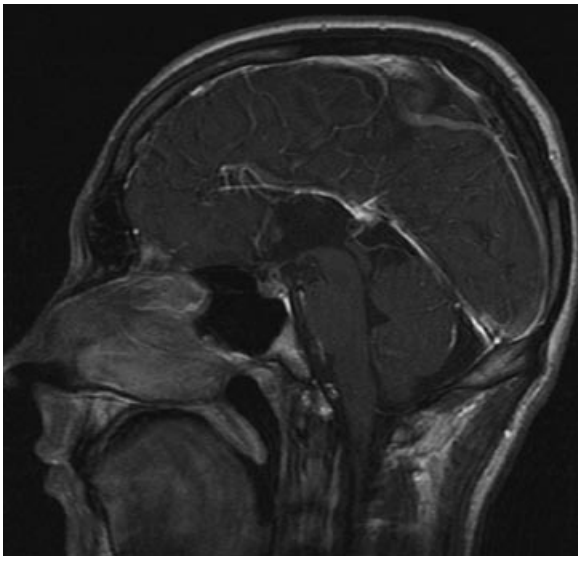

Figure 1. MRI, T1-weighted image, sagittal projection: agenesis of the corpus callosum; commisura anterior visible in front of dilated third ventricle.

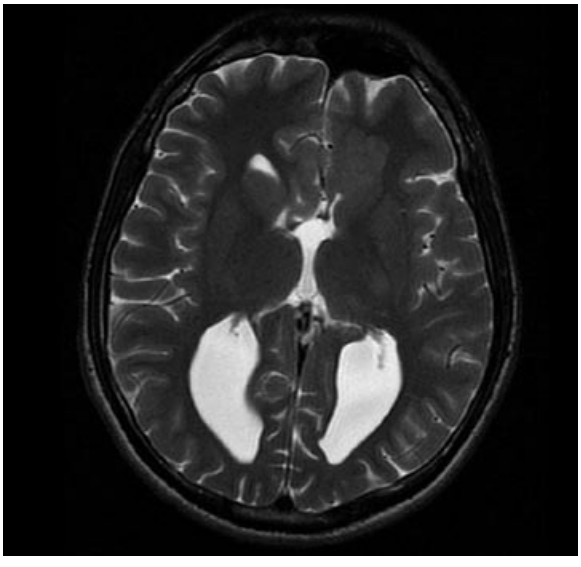

Figure 2. MRI, T2-weighted image, axial projection: typically dilated posterior horns of lateral ventricles and left frontal lobe focal pachygyria.

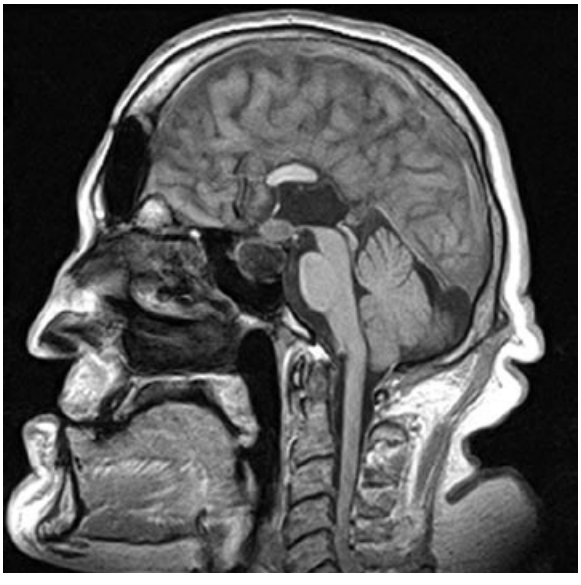

Figure 3. MRI, T1-weighted image, sagittal projection: partial callosal agenesis with the absence of rostrum, splenium and posterior part of body; deepened sella turcica filled with liquid pushing pituitary gland upwards; enlarged frontal sinuses and mandible.

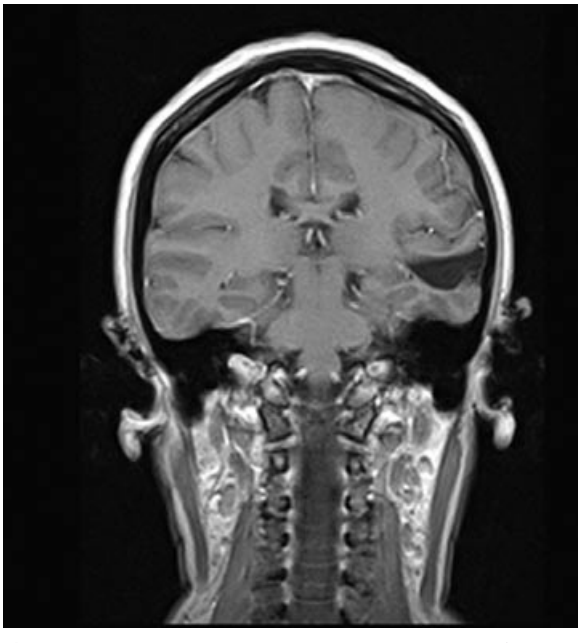

Figure 4. MRI, T1-weighted image, coronal projection: closed fissure in left temporal lobe.

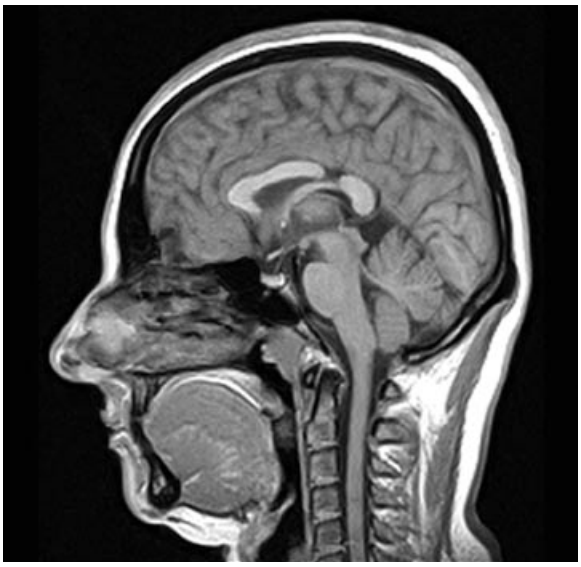

Figure 5. MRI, T1-weighted image, sagittal projection: corpus callosum broken in posterior part of its body at the length of 1 cm.

Case 2

A 44-year-old man with previously diagnosed (CT) and operated pituitary macroadenoma (clinically: acromegalia) undergoing postoperative substantional hormonotherapy was referred to MR Unit for a routine examination. The patient showed no psychological or mental disabilities. MR sagittal scans revealed partial callosal agenesis with absence of rostrum, splenium and posterior part of the body. Besides, it showed a deepened sella turcica which was filled with liquid pushing the pituitary gland upwards. Additionally, characteristic features of acromegalia, such as enlarged frontal sinuses and mandible, were seen (Fig. 3).

Case 3

A 38-year-old woman complaining of left side paraesthesia for a short time, with the history of epilepsy of many years (partial complex seizures with 'deja vu') was sent to MR Unit. Clinicians considered such pathologies as congenital malformation (including arteriovenous malformation) and tumor. MR revealed the presence of closed fissure in left temporal lobe (Fig. 4). Moreover, a quite rare corpus callosum hypoplasia was found – corpus callosum was broken at the length of 1 cm (Fig. 5).

Case 4

A 22-year old woman with suspicion of pure gonadal dysgenesia and obesity (110kg of body mass) was referred to MR scanning for pituitary gland evaluation. There were no mental disorders found in psychological examination. The MR scans did not show any pathology in the pituitary gland. However, a callosal anomaly in form of absence of rostrum and splenium, thickening and shortening of body and genu was found.

Discussion

The diagnostics of the corpus callosum malformations is based on ultrasound scans (prenatal or transcranial), CT, but most of all the MRI (prenatal and postnatal) [6, 7].

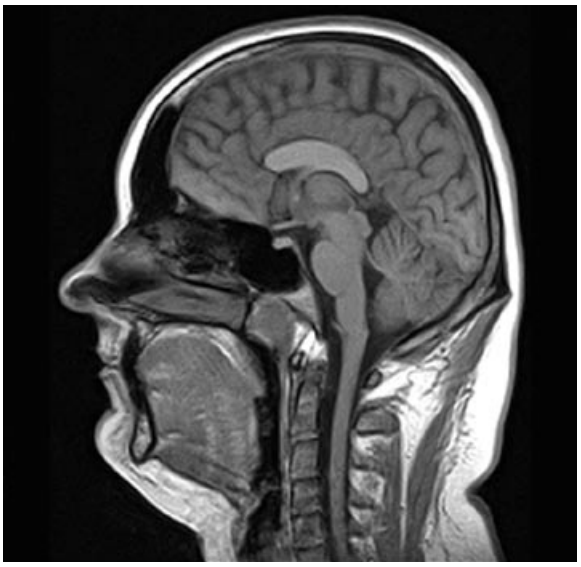

Figure 6. MRI, T1-weighted image, sagittal projection: absence of rostrum and splenium, thickened and shortened callosal body and genu.

In cases of complete callosal agenesis all modalities present splaying of narrow and pointed frontal horns, colpocephaly (medial widening of the lateral ventricles), lateral displacement of the medial walls of the anterior and posterior horns and elevation of the dilated third ventricle. MR and ultrasound scans in sagittal and coronal projections evaluate the absence (complete agenesis) or hypoplasia (partial agenesis) of the corpus callosum.

In all presented cases, radiological diagnosis of callosal anomalies was made incidentally, as in the majority of patients they do not show any specific symptoms, and are most often asymptomatic. Except for these major callosal defects described in our report, MRI quite often reveals tiny, subtle anomalies of callosal shape, i.a. focal thinning of posterior part of callosal body (Fig. 7).

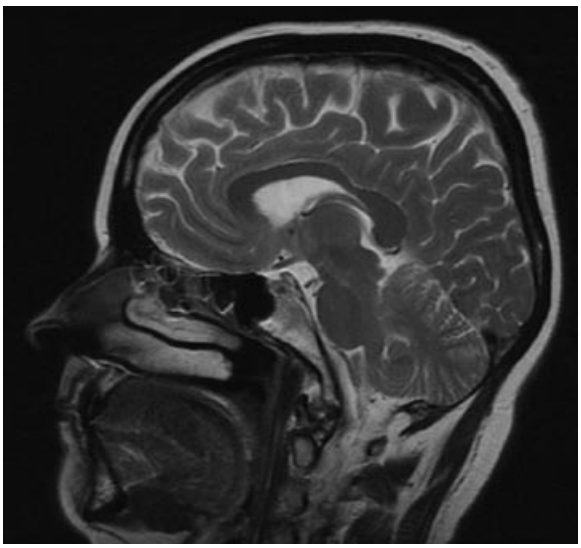

Figure 7. MRI, T2-weighted image, sagittal projection: tiny anomaly of callosal shape – thinning of posterior part of callosal body.

The cases described in this study show the congenital malformations of the corpus callosum. However, it must be remembered, that this anatomical structure can also be affected by acquired disorders, such as traumatic, vascular, inflammatory and neoplastic pathologies. Corpus collosum dysgenesis should also be differentiated from callosal atrophy.

Conclusions

With the development of diagnostic imaging (especially MR) the number of diagnosed callosal malformations has greatly increased. In general, they are found incidentally. Corpus callosum dysgenesis is asymptomatic in most cases, although among children it is often accompanied by other CNS defects.

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