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The Racing Car sign and Devil's Horn appearance: important radiological signs of Corpus Callosum agenesis

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ABSTRACT

Agenesis of corpus callosum (ACC) is a common developmental anomaly of the brain. Often, the diagnosis is delayed due to misinterpretation of the radiological appearances. We here present a case of ACC with typical radiological signs. These radiological signs can be diagnosed very easily and will help to signpost the child to proper care at an early age.

INTRODUCTION

The corpus callosum is the largest interhemispheric association fibre bundle in the brain and is the largest fibre tract in the central nervous system [1]. Agenesis of corpus callosum (ACC) is one of the most frequent congenital malformations of the brain [1]. It may occur on its own or may be associated with other congenital malformations of the central nervous system. Also, it may be associated with developmental and behavioural problems in children.

ACC can be easily diagnosed by its typical radiological appearances. Since this is quite common a condition, clinicians of all branches should be aware of the radiological appearances of ACC. We here present such a case with some of the typical radiological appearances.

THE CASE REPORT

A 15 year old boy was admitted to our hospital with fever. While examining him, we noticed slight dysmorphic appearance of the face (Figure 1) with frontal bossing. His body weight was 35 kg and height 148 cm (BMI: 16). There was no other skeletal abnormality or skin changes. Developmental history revealed that the boy had delayed motor milestones and delayed speech development. He had studied till fourth standard at school after which he had to leave school as he had difficulty in memorizing his lessons. However, his communication skills were normal. He was a loner in personal life and preferred to stay indoors with very few friends. He was presently working as a labourer which did not require much specialized skills. He was good at following commands. The boy was completely independent in activities of daily living. He had never had seizures or syncope.

CT scan of the brain (figure 2) revealed complete agenesis of corpus callosum with parallel non-converging lateral ventricles in axial image. Also, there was presence of racing car sign and devil's ear sign.

The boy was referred to neuropsychiatry department for evaluation and counselling.

DISCUSSION

Agenesis of corpus callosum (ACC) has certain typical radiological signs. The “racing car” sign is found in axial image, CT scan or MRI scan. It is actually like a formula one racing car seen from above, as illustrated in figure 3. The two lateral ventricles are parallel, instead of being converging and they resemble tyres of a racing car [2]. The third ventricle is dilated and migrated superiorly and it represents the body of the racing car. The devil's horn sign is the typical appearance (as seen here) of occipital horns. The horns are dilated posteriorly with anterior end ending in a sharp angulation due to impingement of Probst bundles [3]. This is also called “tear drop” sign [2]. There are also additional radiological signs of ACC like absence of cingulate gyrus and a third ventricle which opens superiorly to interhemispheric fissure [3]. But interpretation of these signs usually require presence of a radiologist. But the racing car sign and devil's horn sign can be diagnosed even by basic physicians

Fig. 1 : Facial appearance of the patient with frontal bossing and facial muscle atrophy

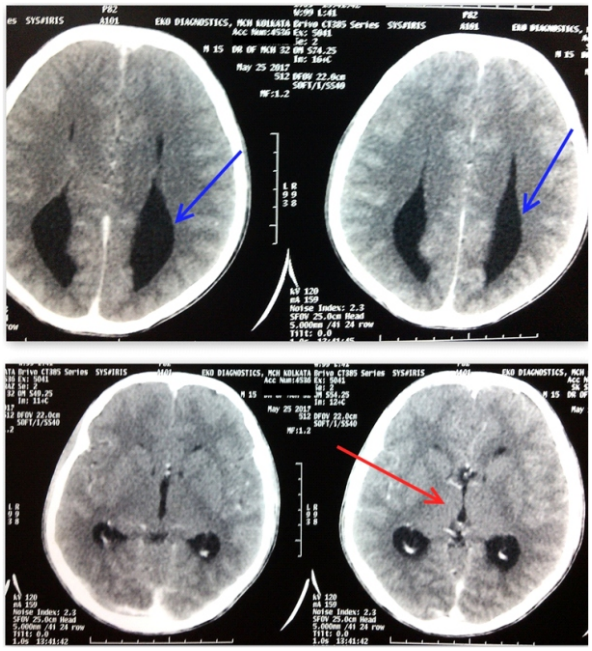

Fig. 2 : axial CT scan showing devil's horn sign (blue arrow), also called tear drop sign or colpocephaly and racing car sign (red arrow).

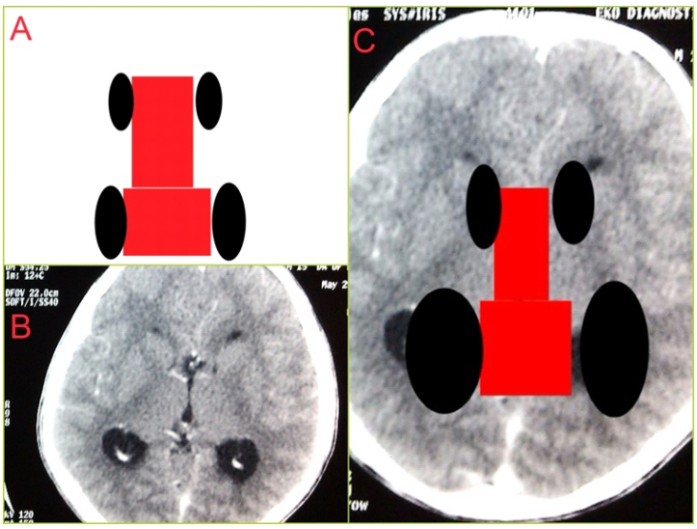

Fig. 3 : schematic diagram of a racing car (A), the Axial CT scan of the brain (B) and the schematic racing car diagram superimposed on the Axial CT scan to demonstrate "racing car" sign

and hence, will be of use in rural remote health centres. Proper radiological diagnosis can help to avoid unnecessary treatment for hydrocephalous, with which this is often confused, or other cerebral malformations. ACC can also be diagnosed by antenatal ultrasound study [2].

CONCLUSION

Clinicians should be aware of radiological appearance of ACC. This short article aims to sensitize general clinicians about these typical radiological signs.

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