

Hemihydranencephaly; a Case Report

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Abstract

Background: Hemihydranencephaly is a rare disorder of the brain characterized by complete or almost complete unilateral absence of cerebral cortex with preservation of meninges, basal ganglia, pons, medulla, cerebellum, and falx.

Case Presentation: Thirteen year-old male child presented with left sided upper and lower limb weakness with facial asymmetry since the age of six months. His magnetic resonance imaging (MRI) scans demonstrated a nearly complete absence of the right cerebral hemisphere including basal ganglion, which was replaced by cerebrospinal fluid with a small residual rim of the occipital cortex. The imaging features were suggestive of right-sided hemihydranencephaly.

Conclusion: Patients with hemihydranencephaly provide an experiment of nature with potential implications for normal cognitive development and illustrate how much there is still to be learned about human development.

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Key Words: Hemihydranencephaly; Brain, Congenital anomalies; Carotid artery; Aplasia

Introduction

Hemihydranencephaly characterized by complete or almost complete unilateral absence of cerebral

cortex with preservation of meninges, basal ganglia, pons, medulla, cerebellum, and falx^[1] is a severe and extremely rare brain condition with only few case reports in the literature^[2-11]. We

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report one more case of right-sided hemihydranencephaly and review the relevant literature.

Case Presentation

13 year-old male child presented with left sided upper and lower limb weakness with facial asymmetry since the age of six months (Fig 1). The child was born at term from nonconsanguineous healthy parents with no complications. He is the eldest son among 2 otherwise healthy children. His sensorimotor and language development during childhood was not restricted and he is going to school, studying in fourth standard and is an average student. There was no history of seizures. Family history was unremarkable. His general and systemic examination was normal except for disuse atrophy and contractures on left face, upper and lower limbs (Fig 1). There was left upper motor neuron type of facial weakness. Tone was increased in left upper and lower limbs and plantar reflex was extensor on left side. Right upper and lower limbs were normal. His magnetic resonance imaging (MRI) scans

demonstrated a nearly complete absence of the right cerebral hemisphere including basal ganglion, which was replaced by cerebrospinal fluid. Only a small residual rim of the occipital cortex was seen and there cerebral peduncle was smaller on right side (Fig 2 and 3). The cerebellum and brain stem was apparently of normal size. MR angiography and venography demonstrated the absence of the right internal carotid artery and draining veins (Fig 4).

Routine laboratory examinations, including coagulation assessment, electrocardiogram, and renal ultrasound, were normal.

Discussion

Clinical features of hemihydranencephaly include contralateral hemiparesis, mild to severe mental retardation and seizures^[2-11]. As in present case normal motor functions in the initial few months after birth are consistent with the thinking that motor control at this age is subcortical^[11]. Occlusion of the ipsilateral carotid artery occurring before the last trimester of gestation (20th to 27th week of gestation) is proposed as the underlying mechanism for brain damage to

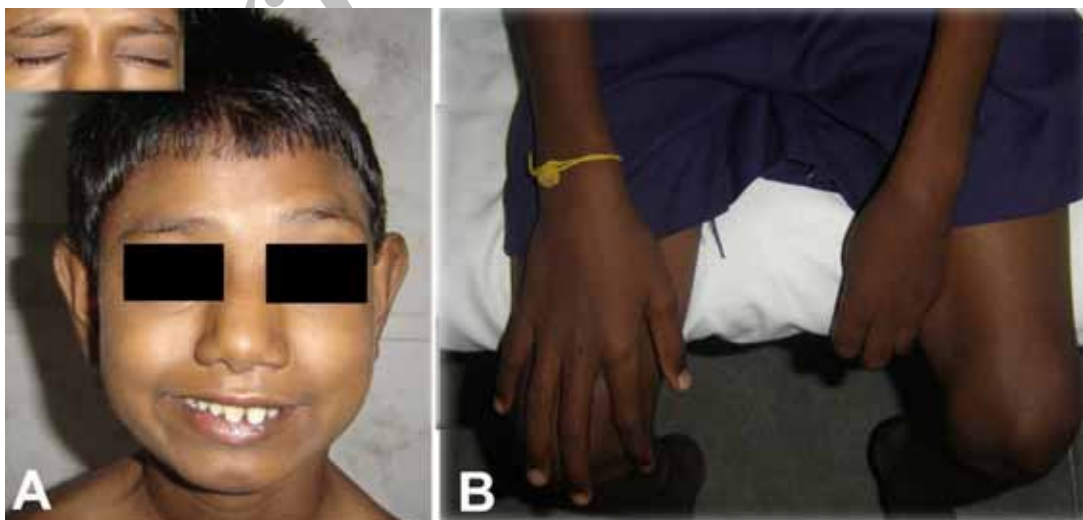


Fig 1: Clinical photograph showing the asymmetry of face, left upper motor neuron type of facial palsy (A) and contracture at the wrist (B)

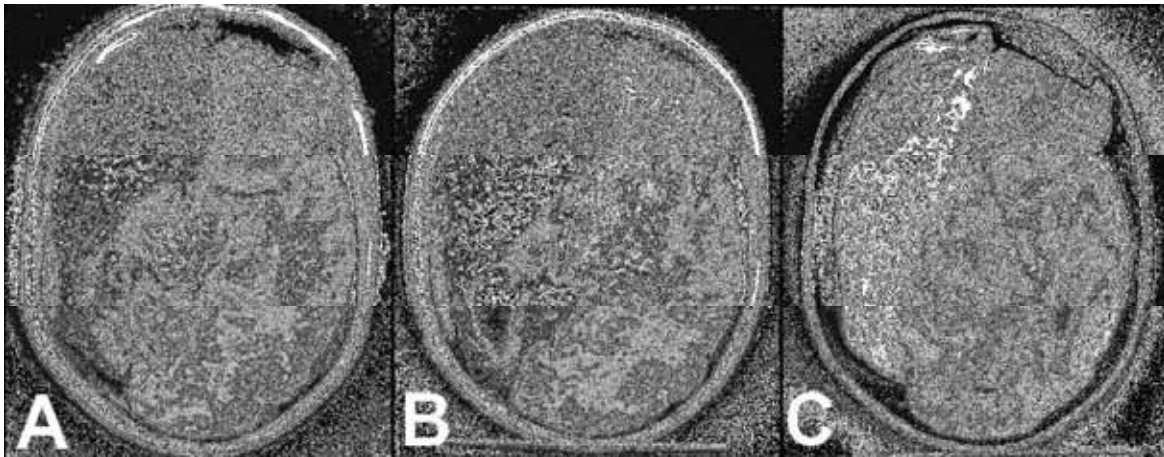


Fig 2: MRI axial image showing absence of right cerebral cortex and replace of the cavity with cerebrospinal fluid, the corpus callosum, basal ganglion and third ventricle are also not visualized, note the brain stem and the cerebellum are well preserved, the smaller cerebral peduncle on right side.

occur and which affects all of the supplied territories^[2,12]. It is largely unknown why hemihydranencephaly affects only one Occlusion of the ipsilateral carotid artery occurring before the last trimester of gestation (20th to 27th week

of gestation) is proposed as the underlying mechanism for brain damage to occur and which affects all of the supplied hemisphere and why the circle of Willis is not able to compensate for the demands in the developing brain^[10].

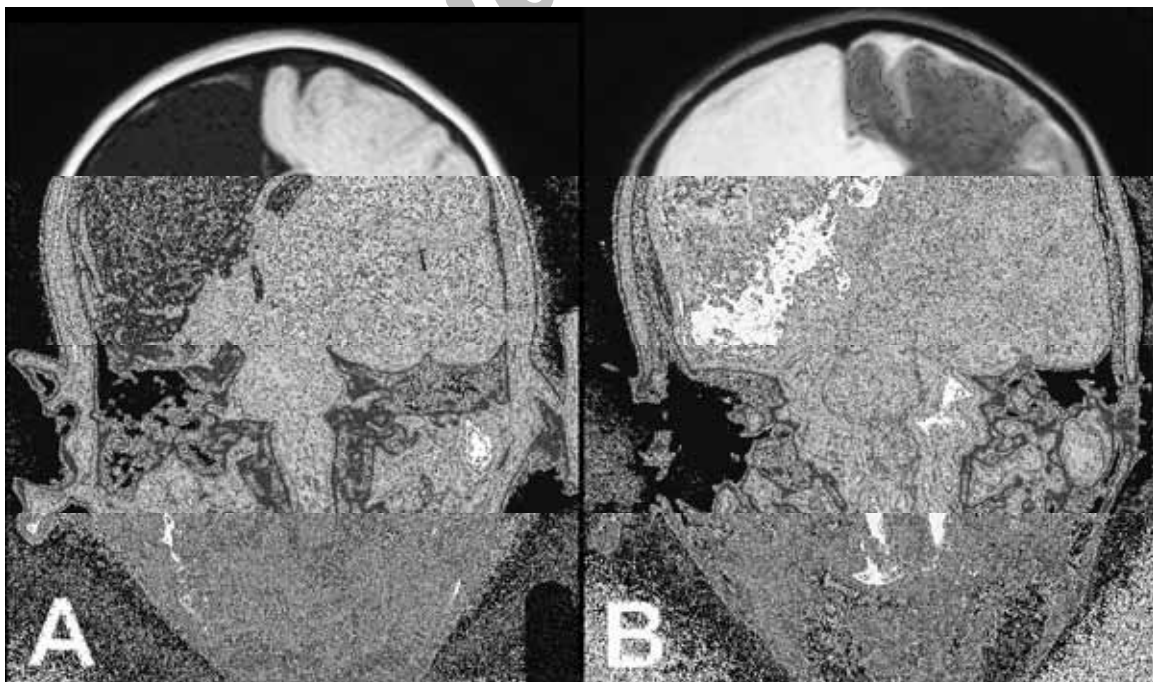


Fig 3: MRI coronal image showing absence of right cerebral cortex and replace of the cavity with cerebrospinal fluid, the corpus callosum, basal ganglion are also not visualized, note the smaller cerebral peduncle on right side.

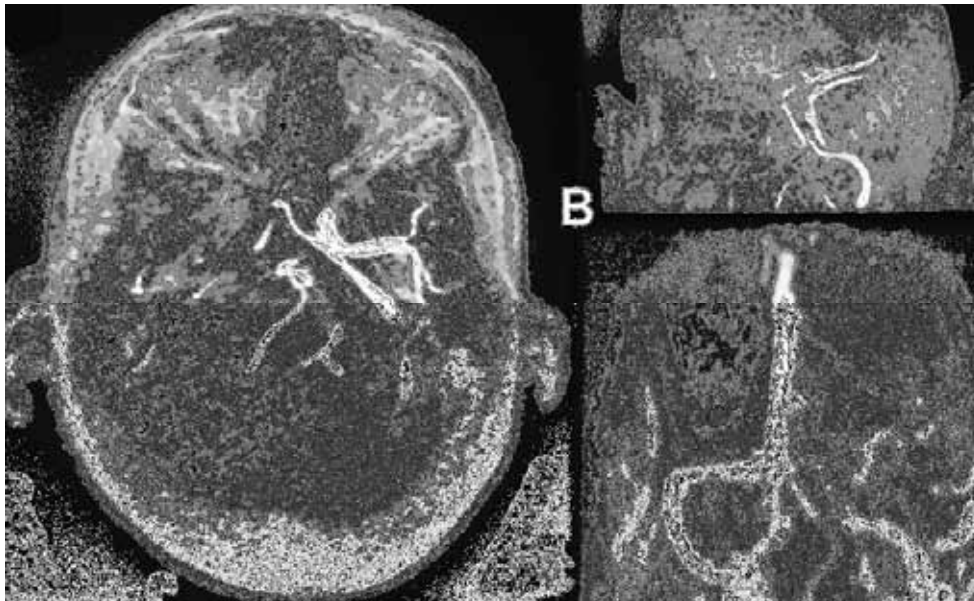


Fig 4: MR angiography showing the absence of the right internal carotid artery and veins on right side.

In patients with hemihydranencephaly the brain damage is assumed to occur after neural migration and before synaptogenesis^[13] and a number of predisposing conditions are implicated as the cause of damage, leading to vascular aplasia with vessel occlusion^[2-11,13]. In spite of the large parenchymal defects patients with hemihydranencephaly, carry a better prognosis than for most other destructive encephalopathies^[2-11]. The relatively good function in the preserved hemisphere in patients with hemihydranencephaly is attributed to neuronal plasticity^[2,9], as the immature brain may compensate for neuronal injury through cortical reorganization that is superior to such capacities in the adult brain^[14].

Conclusion

Patients with hemihydranencephaly provide an experiment of nature with potential implications for normal cognitive development^[9] and illustrate how much there is still to be learned about human development^[11].

References

1. Mori K. Anomalies of the central nervous system. In: Mori K (ed). *Neuroradiology and Neurosurgery*. New York, Thieme-Stratton. 1985; Pp:39-42.
2. van Doornik MC, Hennekam RC. Hemihydranencephaly with favourable outcome. *Dev Med Child Neurol*. 1992;34(5):454-8.
3. Quek YW, Su PH, Tsao TF, et al. Hydranencephaly associated with interruption of bilateral internal carotid arteries. *Pediatr Neonatol*. 2008;49(2):43-7.
4. Tsai JD, Kuo HT, Chou IC. Hydranencephaly in neonates. *Pediatr Neonatol*. 2008; 49(4):154-7.
5. Moser RP, Seljeskog EL. Unilateral hydranencephaly: Case report. *Neurosurgery*. 1981;9(6):703-5.
6. Suzuki M, Seki H, Yoshimoto T. Unilateral hydrocephalus combined with occlusion of the ipsilateral internal carotid artery. *Surg Neurol*. 1985;24(1):27-30.
7. Bae JS, Jang MU, Park SS. Prolonged survival to adulthood of an individual with

- hydranencephaly. *Clin Neurol Neurosurg.* 2008;110(3):307-9.
8. Porro G, Wittebol-Post D, de Graaf M, et al. Development of visual function in hemihydranencephaly. *Dev Med Child Neurol.* 1998;40(8):563-7.
9. Greco F, Finocchiaro M, Pavone P, et al. Hemihydranencephaly: case report and literature review. *J Child Neurol.* 2001;16(3): 218-21.
10. Ulmer S, Moeller F, Brockmann MA, et al. Living a normal life with the nondominant hemisphere: magnetic resonance imaging findings and clinical outcome for a patient with left-hemispheric hydranencephaly. *Pediatrics.* 2005;116(1):242-5.
11. Altschuler EL, Matsumura B, Capoor J, et al. Left-Hemispheric Hydran-encephaly with less favorable findings. *Pediatrics.* 2005;116(6);1603-4.
12. Myers RE. Cerebral ischemia in the developing primate fetus. *Biomed Biochim Acta.* 1989;48(2-3):S137-S142
13. Eyre JA, Miller S, Clowry GJ, et al. Functional corticospinal projections are established prenatally in the human foetus, permitting involvement in the development of spinal motor centres. *Brain.* 2000;123(pt 1): 51-64.
14. Kutzt-Buschbeck JP, Dreesmann M, Golge M, et al. Prenatal infarction of the left middle cerebral artery: a case report of excellent functional outcome. *Neuro Rehabilitation.* 2000;15(3):167-73.
15. Merker B. Life expectancy in hydranencephaly. *Clin Neurol Neurosurg.* 2008; 110(3):213-4.

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