

Congenital cerebral hemiatrophy: a case study and review of the literature

Abstract

Background: Congenital malformations of the central nervous system encompass a set of developmental anomalies. Cerebral hemiatrophy (CHA) is a rare radiological finding.

Case presentation: We report the case of a 3-year-old girl admitted for progressive right-sided hemiparesis, associated with psychomotor delay and seizures. Brain CT scan revealed severe right hemispheric atrophy with extensive parieto-temporal schizencephaly. Management consisted of antiepileptic therapy, motor rehabilitation, and regular neurological follow-up.

Conclusion: Congenital CHA, often related to late neuronal migration disorders, requires an etiological diagnosis to optimize management and prognosis.

Keywords: cerebral hemiatrophy, schizencephaly, congenital malformations, hemiparesis

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Introduction

Congenital malformations of the central nervous system (CNS) are a major cause of neurological morbidity in children. Cerebral hemiatrophy (CHA) is a radiological sign characterized by a loss of volume in one cerebral hemisphere, which may result from agenesis, perinatal destruction, or a cortical developmental disorder such as schizencephaly.¹ We report a case of congenital HAC revealed by extensive schizencephaly and discuss the diagnostic and therapeutic aspects.

Clinical observation

A 3-year-old girl from a consanguineous marriage was admitted for progressive motor deficit on the right side of her body. The pregnancy was not monitored, and delivery was at term by vaginal delivery after prolonged labor. Clinical examination revealed. The patient presented with contralateral spastic hemiparesis 4/5, predominantly affecting the upper limb, with pyramidal hypertonia, hyperreflexia, and a positive Babinski sign. Segmental amyotrophy of the contralateral upper limb was noted. Neuropsychological examination revealed moderate psychomotor delay and language impairment characterized by slowness and poverty of speech. Cranial nerve examination showed central facial palsy contralateral to the cerebral atrophy. She also experienced recurrent focal seizures, partially controlled by antiepileptic treatment (MRC). Paraclinical assessment: Brain MRI was not available due to limited resources, A brain scan showed right hemispheric atrophy, dilation of the right lateral ventricle communicating with the subarachnoid spaces, and a right parieto-temporal cortical cleft consistent with type II schizencephaly. In our patient the treatment consisted of medical management combining an antiepileptic drug to control seizures, a motor rehabilitation program, and regular neurological follow-up (Figure 1).

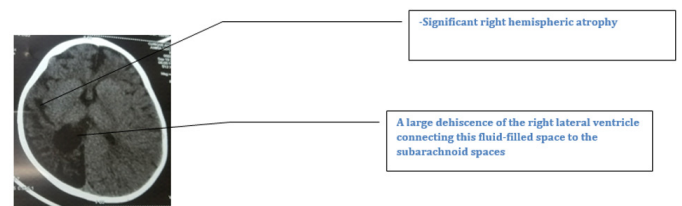


Figure 1 Axial CT scan showing morphological and structural changes in the cerebral parenchyma:

Discussion

HAC can be congenital or acquired. Congenital forms are often linked to early neuronal migration disorders, including schizencephaly.² Risk factors: genetic abnormalities, intrauterine infections, consanguinity.³ Diagnosis is based on brain imaging, with MRI being the gold standard.⁴ Treatment is symptomatic: antiepileptic drugs, motor rehabilitation, functional surgery in cases of refractory epilepsy.^{5,6} Differential diagnoses include Dyke-Davidoff-Masson syndrome (characterized by calvarial thickening and sinus hyperpneumatization), Rasmussen's encephalitis (progressive unilateral hemispheric atrophy with drug-resistant epilepsy), sequelae of neonatal or infantile stroke, cortical malformations (schizencephaly, polymicrogyria), and post-infectious sequelae.⁷⁻⁹

In our observation, the therapeutic strategy was based on an exclusively medical approach. The introduction of appropriate antiepileptic treatment reduced the frequency of seizures. Early motor rehabilitation aimed to limit functional sequelae, in accordance with the recommendations reported in the African literature.^{10,11} Sow et al.³ emphasize the importance of early multidisciplinary management to

improve the functional prognosis of brain malformations. Similarly, Diop et al.⁶ stress the role of regular neurological follow-up in the detection and prevention of long-term complications. Management is mainly symptomatic: antiepileptic drugs, motor rehabilitation, and speech therapy. In severe or pharmaco-resistant cases, functional neurosurgery such as hemispherectomy or hemispherotomy may be considered.¹²

Conclusion

Congenital HAC is rare but debilitating. Early diagnosis, appropriate symptomatic management, and rigorous prenatal monitoring are essential to improve prognosis.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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