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Right cerebral hemiatrophy: Neurocognitive and electroclinical features

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ABSTRACT

The purpose of this study was to retrospectively evaluate the cognitive and electroclinical characteristics of right cerebral hemiatrophy (Dyke–Davidoff–Masson syndrome [DDMS]). Cognitive assessments with a particular emphasis on visuospatial functions, electroclinical features, and neuroimaging characteristics were analyzed for five patients with a clinically and neuroradiologically confirmed diagnosis of right-sided DDMS. Intelligence tests revealed mental retardation in all but one. Neuropsychological assessments demonstrated consistent impairments in tasks that have a spatial component (spatial processing and orientation discrimination), whereas attention, executive functions and verbal memory domains were variably impaired. Electroclinically, the main seizure types were simple partial motor, complex partial, and secondarily generalized seizures. Interictal EEG delineated lower amplitudes and slow background activity in the affected hemisphere. Overall, the cognitive performance of patients with DDMS encompasses a broad spectrum of impairments affecting multiple domains. Our findings support the concept that dorsal visual pathways responsible for spatial processing may be lateralized to the right hemisphere.

1. Introduction

Cerebral hemiatrophy, or Dyke–Davidoff–Masson syndrome (DDMS), is caused by developmental hypoplasia or acquired atrophy of one cerebral hemisphere and leads to diverse clinicopathological entities. Primary radiological characteristics of this syndrome comprise unilateral loss of cerebral volume and secondary compensatory alterations in the calvarium, which include mainly unilateral calvarial thickening, overdevelopment of the paranasal sinuses and petromastoid air cells, and elevation of the sphenoid wing and petrous ridge [1–3]. Atrophy of brainstem and thalamus may also be present; accompanying crossed cerebellar atrophy has occasionally been reported [4,5]. Clinical presentations show divergence according to the degree of brain injury and always include contralateral hemiparesis, facial asymmetry, and epileptic seizures by definition [1,2]. Cerebral hemiatrophy results mainly from vascular etiology, infections, trauma, or prolonged febrile convulsions during the early stages of life.

The distinctive neuroradiological characteristics of this syndrome have drawn eminent attention, and several consecutive reports have lately expanded the neuroimaging findings of the syndrome [6–11]. However, although epilepsy is one of the primary features of the syndrome, its characteristics, seizure types, and electroencephalographic features remain incompletely explored and comprise only individual case reports thus far [2,4,12]. Furthermore, given the lateralized hemispheric organization of the human brain, long-term cognitive disorders resulting from monohemispheric atrophy are especially intriguing in this childhood-onset syndrome. Impairments in language and visuospatial orientation have previously been pointed out in individual cases and need to be further investigated according to the cerebral hemispheric lateralization [2,13].

In this study, we retrospectively report five cases of DDMS with right cerebral hemiatrophy in an attempt to highlight the neurocognitive features of the syndrome affecting the right hemisphere. We predicted that these patients would show robust impairments in specific tasks that demand visuospatial processing. Furthermore, we describe the electroencephalographic characteristics of this syndrome and our clinical experience with the management of seizures resulting from this condition.

2. Methods

All patients followed by our epilepsy outpatient unit between 1993 and 2008 with a clinically and neuroradiologically confirmed diagnosis of right hemiatrophy (DDMS) were included and retrospectively evaluated in this study. Personal history, cognitive and neurological examinations, clinical presentation, seizure characteristics, EEG features, antiepileptic medications, response to treat-

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ments, and neuroradiological findings were analyzed. The study was approved by the local ethics committee.

General intellectual functioning was evaluated using the Wechsler Adult Intelligence Scale (WAIS-R) or Wechsler Intelligence Scale for Children (WISC-R). IQ scores were categorized into three groups: impaired (<70), borderline (70–84), and normal (⩾85). A concise neuropsychological assessment included the following tests [14]: (1) Digit Span subtest (Forward and Backward), (2) Verbal Fluency (Category Naming), (3) Go–No-Go Paradigm, (4) Luria Grapho-Motor Tasks, (5) Boston Naming Test (BNT) (30 items, Turkish modification), (6) California Verbal Learning Test, (7) Benton's Line Orientation Test (BLO), (8) Benton's Facial Recognition Test (BFR), (9) Visual Target Cancellation Task (VTC), (10) clock drawing, (11) copying two-dimensional figures (a diamond within a square, intersecting pentagons [15]), (12) copying three-dimensional figures (cube house), and (13) drawing ordinary objects from memory (flower, human figure). Mood was assessed with Hamilton Depression Scale (HDS) [16].

Ictal symptomatologies were obtained from the patients' own descriptions and those of their first-degree relatives who had observed their seizures. Seizures were classified according to the Commission of the International League Against Epilepsy [17]. EEG evaluation included scalp recordings using the International 10–20 system with 16–21 channel recordings using paperless EEG machines (Medelec DG Compact 2000). Brain MR images were obtained with 1.5-T Picker-Eclipse MRI equipment and comprised T 1-weighted, T 2-weighted, and FLAIR techniques in the axial, coronal, and sagittal planes.

3. 3. Results

3.1. Patient demographics, mental and neurological examination

Five patients (four females) met the requirements for this study. Patient demographics and neurological examination findings are summarized in Table 1. General educational level of the group was subnormal, and learning difficulties were reported in four. Intellectual functioning varied from borderline to impaired (Table 1). All patients manifested variable degrees of impairment in attention, executive functions, and memory. Speech was fluent and grammatical without paraphasias. Comprehension and repetition were normal in all patients; naming, as assessed with the BNT, was subnormal in two. Impairments in visuospatial abilities were remarkable in the whole group. BLO was impaired in all individuals, whereas BFR was normal in two. Only one patient was successful in two-dimensional and three-dimensional figure copying; the rest were found to be impaired in the visuoconstructional tasks. Major impairments included failure of intersection or missing number of angles in pentagon copying, and loss of perspective and disorganized piecemeal approach in three-dimensional copying. When patients were asked to draw a human being from memory, two patients drew the figure without a trunk; extremities stemmed directly from the head and the key face details were omitted (Fig. 1). VTC tasks did not detect hemineglect and HDS did not indicate depression in any of the patients.

3.2. Seizure types, EEG findings, and treatment

Interictal EEG recordings demonstrated primarily lower amplitudes and slow background activity in the affected hemispheres (Fig. 2). A summary of the findings is provided in Table 1. Epilepsy was considered drug resistant in one patient who had daily focal motor seizures leading to secondary generalization during her menstrual periods. Of importance, this patient had the earliest seizure onset in the whole group.

3.3. MRI findings

All patients had right-sided cerebral hemiatrophy on neuroimaging. Additional neuroimaging findings included ipsilateral bony structural changes ($N = 5$), prominent cortical sulci ($N = 4$), thalamic atrophy ($N = 4$), brainstem atrophy ($N = 3$), callosal atrophy ($N = 2$), midline shift ($N = 2$), porencephaly ($N = 1$), and contralateral cerebellar atrophy ($N = 1$) (Fig. 3).

4. Discussion

In this study, we searched for the pattern of cognitive impairment in patients with right hemiatrophy due to DDMS. Our findings demonstrated consistent impairments in orientation discrimination and spatial processing, whereas several cognitive domains including attention, executive, and memory functions

Table 1
Clinical and electroencephalographic features of the patients with right hemiatrophy.

Case	Age	Sex	DDM[a] type	Assumed etiology	IQ	NE[b]	Aura	Age at seizure onset	Seizure type	EEG	AED	AE response	Follow-up period (years)
1	28	M	Postnatal	Intracranial infection (3 months)	Impaired	HP	None	3 months	SPM, SGS	RH ⇓ amp RH theta	DPH	Good	14
2	26	F	Postnatal	Intracranial infecton (1 year)	Borderline	HP, AM	Sparkling red light	1 year	SPM	RH ⇓ amp RH theta RT hs	CBZ	Good	13
3	12	F	Postnatal	Prolonged febrile convulsions (1 year)	Impaired	HP, HE	Staring, oral automatism, epigastric sensation	1 year	SPM, CP	RH ⇓ amp D theta RT is/w	VAL	Good	5.5
4	30	F	Perinatal	Birth trauma, hypoxia	Impaired	HP, HE	None	3 days	SPM, SGS	RH ⇓ amp RH theta	CBZ, PB	Drug resistant	4
5	31	F	Congenital	Vascular	Impaired	HP	Epigastric sensation, staring, oral automatisms	2 years	CP, SGS[c]	RH ⇓ amp RH theta RT is/w RTO hs	CBZ, PB	Good	7

[a] DDM, Dyke–Davidoff–Masson syndrome; NE, neurological examination; HP, hemiparesis; AM, hand athetoid movement; HE, hemihypoesthesia; SPM, simple partial motor seizures; CP, complex partial seizures; SGC, secondarily generalized seizures; RH, right hemisphere; ⇓ amp, decreased amplitude (low voltage); D, diffuse; theta, theta wave activity; is/w, isolated spike and wave; RT, right temporal; hs, hypersynchrony; RTO, right temporo-occipital; DPH, diphenylhydantoin; CBZ, carbamazepine; VAL, sodium valproate; PB, phenobarbital.
[b] Total hemiatrophy comprising the face, body, and extremities was present in all patients.
[c] Secondary generalization was added on partial seizures several years after the onset of seizures.