Neuroimaging findings in children with infantile spasms

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ABSTRACT

Objective: The aim of the study was to document the neuroimaging findings of children with infantile spasms (IS) seen over a 3-year period.

Methods: All children below the age of 4 years who presented to the Pediatric Department at the Northern Area Armed Forces Hospital, Hafr Al-Batin, Kingdom of Saudi Arabia from January 1, 1998 to December 31, 2000 with a history of seizures, atypical movements, psychomotor delay, flexor, extensor spasms or both were included in the study. Relevant birth, developmental and family history as well as information on the pattern of fits were documented. Investigations included complete blood count, serum electrolytes, liver function tests, screening for acquired and congenital metabolic disorders. The electroencephalogram, brain magnetic resonance imaging and computerized tomography scans were carried out routinely on all the children.

Results: There were a total of 30 Saudi children, 17 males and 13 females that fulfilled the criteria for evaluation of infantile spasms. The mean age was 10 months. The major causes of IS in this study were congenital brain lesions (40%) infections (20%), and birth trauma/asphyxia (16.7%). The etiology was unknown in 6 (20%) cases. The neuroimaging pattern was dysgenesis (30%), brain atrophy (23.7%), infarctions/hemorrhage (10%) and hydrocephaly (10%). In 8 cases (26.6%) the findings were normal.

Conclusions: The neuroimaging findings in this study are comparable with observations in other studies carried out under different clinical settings and environment.

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Infantile spasms (IS) are a unique form of the epileptic syndrome characterized by brisk flexion or extension of the neck, trunk and extremities during early infancy. They constitute approximately 2-3% of all childhood epileptic syndromes.[1] Originally, the entity was referred to as the West Syndrome, named after the English physician who vividly described the remarkable pattern of seizures in his son in 1841.[2] Apart from the characteristic myoclonic jerks, affected children frequently present with developmental retardation and an electroencephalographic (EEG) pattern referred to as hypsarrhythmia.[3] In a recent study in Saudi Arabia[4] approximately 5% of cerebral palsied children were found to have a hypsarrhythmic EEG pattern. In another review[5] 4.4% of Saudi children with corpus callosum agenesis had infantile spasms. The objective of this study was to demonstrate the neuroimaging findings in children with IS at the Northern Area Armed Forces Hospital, Hafr Al-Batin, Kingdom of Saudi Arabia from January 1, 1988 to December 31, 2000.

Methods. All children below the age of 4 years with a history of seizures and atypical movements with

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*Sadly, the first author died on 1st August 2003, condolences from the Editors and Editorial Board. As we failed to contact the second author, we acknowledge Dr. Okoroma who proof read the manuscript and can be contacted for any future correspondence.

Table 1 - Relevant clinical data of patients.

Case No.	Age of onset (months)	Sex	Etiology/clinical	Neuroimaging findings
1	4	F	Prenatal infection, CMV?, CHD/ cardiac surgery	Brain atrophy
2	20	M	Birth asphyxia	Cortical brain atrophy
3	6	F	Congenital microcephaly deafness	Normal
4	3	M	Etiology unknown	Normal
5	4	F	Birth asphyxia	Normal
6	10	F	Prenatal infection meningo encephalitis?	Hypodensity left parietal area (hemorrhage?)
7	7.5	M	Congenital	Corpus callosum agenesis
8	8	M	Prematurity and asphyxia	Moderate cerebral atrophy with dilated ventricles
9	8	M	Birth trauma	Infarction left temporal area
10	21	F	Idiopathic	Brain atrophy
11	3	M	Birth asphyxia and hyperbilirubinemia, congenital	Mega cisterna magna, corpus callosum agenesis
12	20	M	Hydrocephalus with meningomyelocele	Hydrocephalus with VP shunt
13	8	M	Congenital psychomotor delay	Lissencephaly with frontal macrogyria
14	7	M	Encephalopathy, post DPT?	Normal
15	24	M	Meningomyelocele with VP shunt	Hydrocephalus
16	12	M	Neonatal meningitis (Group B. strept.)	Hydrocephalus
17	7	F	Demyelinating disease (metabolic)	Normal
18	10	M	Congenital club foot, hyporeflexia	Brain atrophy with cavum septum pellucidum
19	6	F	Infantile seizures with intractable seizures gross motor delay	Cortical defect left hemisphere with ventricular dilatation
20	6	M	Psychomotor delay	Schizencephaly with corpus callosum agenesis
21	24	M	Congenital (dysgenetic?), delayed intellectual and motor milestones	Agenesis of the vermix and corpus callosum, Dandy Walker malformation
22	20	M	Birth asphyxia	Dilated ventricle and paraventricular infarction
23	11	F	Congenital microcephaly with mental retardation	Brain atrophy
24	8	M	Cerebral palsy	Isolated lissencephaly
25	4	F	Post meningitis, CP	Normal
26	12	F	Congenital hydrocephalus with VP shunt	Hydrocephalus with aqueduct stenosis
27	5	M	Neonatal sepsis (infection)	Normal
28	11	F	Tuberous sclerosis with café au lait patches	Multiple hypodense calcified areas
29	6	M	Hyperglycinemia (metabolic)	Brain atrophy
30	3	F	NSVD, post meningitis/sepsis	Normal

CMV - cytomegalovirus, CHD - coronary heart disease, DPT - diphtheria-pertussis-tetanus, Group. B. strept - Group B Streptococcus, CP - cerebral palsy, NSVD - normal spontaneous vaginal delivery, VP - ventriculoperitoneal, M - Male, F - Female

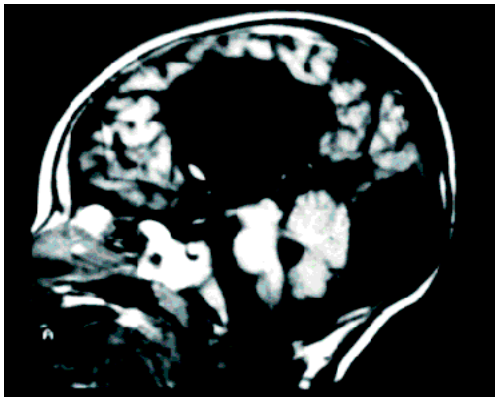

Figure 1

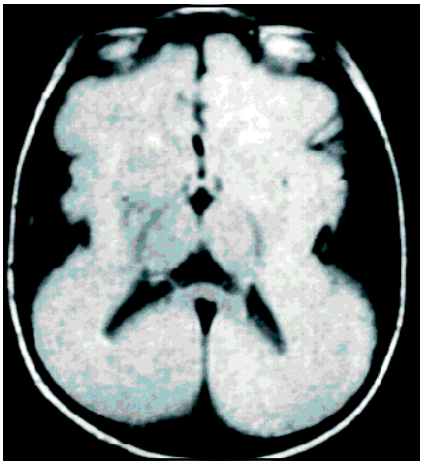

Figure 4

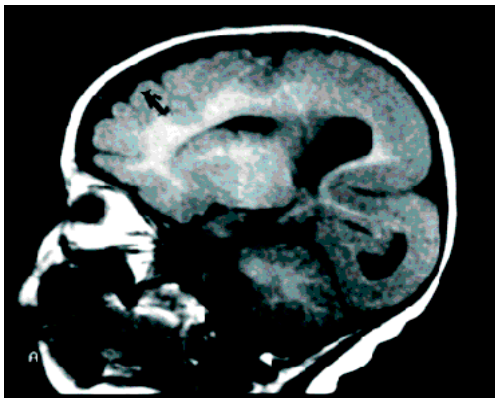

Figure 2

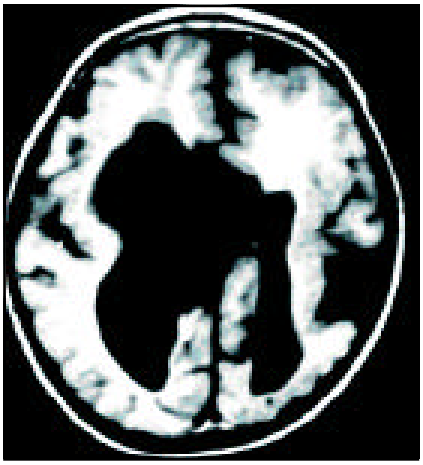

Figure 5

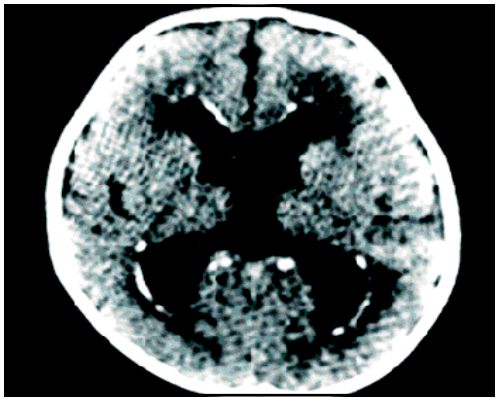

Figure 3

Figure 1 - Mega cisterna magna. Corpus callosum agenesis with interhemispheric cystic formation.

Figure 2 - Occipital and posterior parietal lissencephaly with frontal macrogyria.

Figure 3 - Tuberous sclerosis with subependymal and occipital calcified nodules.

Figure 4 - Isolated diffuse lissencephaly.

Figure 5 - Parietal parasagittal large “open lip” schizencephaly with corpus callosum agenesis.

or without psychomotor delay were included in this partly retrospective study. The patients were either identified in the pediatric ward or pediatric neurology clinic. Relevant birth, developmental and family history as well as information about fits, spasms or atypical movements with age of onset were obtained retrospectively. Laboratory investigations included the full blood count, serum electrolytes, liver function tests, and various tests for acquired and congenital metabolic disorders. Electroencephalographic studies and brain magnetic resonance imaging (MRI), computerized tomography (CT) scans or both were carried out routinely in all patients. The developmental status was assessed using the Denver Developmental Screening Test.

Results. There was a total of 30 Saudi patients (17 males and 13 females) who satisfied the clinical criteria of having infantile spasms. This gave a male to female ratio of 1.3 to 1. The age ranges varied from 2 to 36 months, with an overall mean age of 10 months. Twenty-one children (70%) had a seizure onset before the age of 12 months while 27 (90%) did so by 2 years. Intellectual impairment was documented in 97% of the children. Regarding the EEG patterns, 16 children (54.4%) presented with hypsarrhythmia, 6 patients (20%) had a suppression burst pattern and 4 (13.3%) showed dysrhythmic EEG features. **Table 1** shows the relevant clinical data on the patients. Probable causes of IS as seen in our patients are congenital lesions (12; 40%), infections (6; 20%), birth trauma/asphyxia (5; 16.7%), metabolic disorder (1; 3.3%), and idiopathic (6; 20%). The distribution of the neuroimaging findings were normal (8; 26%), dysgenetic lesions (9; 30%), brain atrophy (7; 23%), infarctions/hemorrhage (3; 10%), and hydrocephalus (3; 10%).

Discussion. In all 30 cases, the diagnosis of IS was confirmed by applying the clinical criteria of myoclonic jerks, hypsarrhythmia/modified hypsarrhythmia and in all but one case intellectual impairment. The sex distribution was within the limits of many large series.[6,7] Despite the small number of cases, the etiologic and neuroimaging findings are comparable to accounts in previous studies[8,9] with a predominance of cases in the secondary/symptomatic group. There were a total of 24 cases (80%) in our study. In this category, the sub group of dysgenetic lesions were quite prominent with lesions like mega cisterna magna with corpus callosum agenesis (**Figure 1**), lissencephaly with frontal macrogyria (**Figure 2**), cerebral calcifications in tuberous sclerosis (**Figure 3**), isolated lissencephaly (**Figure 4**) and schizencephaly with corpus callosum agenesis (**Figure 5**) among others. This compares favorably with the 20-30% in other international series.[10-12] In one study[13] 13 out of 32 autopsied cases of IS had malformations of the brain. Undoubtedly, dysgenetic lesions constitute a major cause of IS. In this respect, the superiority of the MRI over the brain CT scan as a neuroimaging procedure cannot be overemphasized.

From the varied etiologic factors and neuroimaging findings, it appears acceptable that the infantile spasm truly represents a non-specific reaction of the brain to various insults,[2,8] confirming the impression that it is more age related than a specific disease. What remains unexplainable is that not all injuries incurred on the immature brain will give rise to infantile spasms.[14]

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