Pattern of congenital brain malformations at a referral hospital in Saudi Arabia: An MRI study

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BACKGROUND: More than 2000 different congenital cerebral malformations have been described in the literature, for which several classification systems have been proposed. With the help of these classification systems, it is now possible, with neuroimaging, to time neuroembryologic events. Magnetic resonance imaging (MRI), in particular, is useful in studying these malformations. This study evaluated the pattern of congenital brain malformations in a university referral hospital setting.

PATIENTS AND METHODS: The records of all MRI brain examinations at our hospital over a period of 3 years for children younger than 15 years of age were reviewed. Cases of congenital cerebral malformations were analyzed by sex, age at presentation, type of congenital cerebral malformation, and other associated congenital cerebral malformations.

RESULTS: Of the 808 MR examinations of different parts of the body for children in the study period, 719 (89%), on 581 patients, were of the brain. Eighty-six children (14.8%) were found to have single or multiple congenital brain malformations. In these children, 114 congenital brain malformations were identified, the commonest being cortical migrational defects (25 patients, 22%), neural tube closure defects (22 patients, 19%), and corpus callosum dysgenesis (22 patients,19%). The least common was vascular malformation. Sixteen patients (18.6%) had more than one congenital brain malformation.

CONCLUSION: Neural tube closer defects, cortical migrational abnormalities, and corpus callosum anomalies were the commonest congenital brain malformations, while vascular malformations were the least common. Most of the identified malformations demonstrated the usual pattern, but a few showed unusual patterns and associations.

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Congenital brain malformations are extremely heterogeneous, and often more than one malformation exists in the same patient. Traditionally, these malformations were studied and classified based on pathologic examination. However, the development of magnetic resonance imaging (MRI) has resulted in a much better understanding of the entire spectrum of congenital brain malformations, as investigation by means of autopsy was limited to the most severe end of the spectrum. Furthermore, MRI has allowed us to observe the evolution of these malformations as the brain undergoes the process of maturation in infancy and childhood.[1] The aim of this study was to evaluate the pattern of congenital brain malformations in a referral university hospital in Saudi Arabia based on MRI findings.

Materials and Methods

All MRI studies of the brain for children younger than 15 years of age at our hospital over a 3-year period were retrospectively reviewed. We analyzed cases with congenital cerebral malformations by sex, age at presentation, type of congenital cerebral malformation, and other associated congenital cerebral malformations. The minimum imaging requirement was the standard MRI sequences of sagittal T1-wieghted images (T1WI), axial T2WI and fluid attenuation inversion recovery (FLAIR) images. Other sequences such as high-resolution brainstem T2WI, 3-dimentional spoiled gradient-echo (3D SPGR), short-tau inversion recovery (STIR), and post-gadolinium images were reviewed when available. Cases with inadequate or technically suboptimal examination were excluded from this study. All patients older than 15 years of age at the time of initial MRI examination were also excluded.

Results

MRI examinations of different parts of the body were performed on 808 pediatric patients at our hospital over the 3-year period using the 1.5T system. Seven hundred and nineteen (89%) exams on 581 patients were of the brain. Congenital cerebral malformations were found in 86 (14.8%) patients. The total number of cerebral malformations identified in these patients was 114 (Table 1). The age of patients at first MRI imaging ranged from 3 days to 15 years (mean, 3 years and 9 months). There were 44 girls (51.2%) and 42 boys (48.8%). The presence of two or more congenital cerebral malformations was seen in 16 (18.6%) patients. The most common malformations occurring in combination were corpus callosum dysgenesis, cortical dysplasia, lissencephaly (agyria/pachygyria complex), and gray matter heterotopia. Table 2 shows the frequency of the different cerebral malformations in the study group.

Corpus callosum dysgenesis

Corpus callosum dysgenesis was seen in 22 (25.6%) patients. Table 3 shows the clinical data, type of corpus callosum malformation, and the other associated congenital cerebral malformations in these 22 patients. In 10 (45%) patients the entire corpus callosum was absent (agenetic). In 9 (41.5%) patients the callosal agenesis partially affected the splenium and posterior half of the body, and in 1 (4.5%) patient there was global hypoplasia of the corpus callosum with no signs of agenesis. In 2 (9%) patients part of the body of corpus callosum was not developed, fit-

Table 1. Distribution of 114 cerebral malformations in 86 patients.

Neural tube closure defects	
Chiari I	1
Chiari II	12
Chiari III	1
Meningo-encephalocele	4
Dermoid cyst	4
Cortical migrational defects	
Lissencephaly/pachygyria complex	19
Heterotopia	6
Cortical organizational defects	
Focal cortical dysplasia	6
Polymicrogyria (PMG)	3
Schizencephaly	1
Vascular Malformations	
AVM	1
Venous angioma	1
Neurocutanous syndromes	
Neurofibromatosis I	9
Sturge-Weber syndrome	4
Tuberous sclerosis	1
Posterior fossa malformations	
Joubert syndrome	3
Dandy-Walker malformation	3
Cerebellar malformation	4
Brainstem malformation	1
Corpus callosum dysgenesis	
Partial	11
Complete	10
Global hypoplasia	1
Others	
Holoprosencephaly	3
Intracranial lipoma	5
TOTAL	**114**

Table 2. Description of 16 patients with multiple congenital cerebral malformations.

Age	Sex	Clinical history	Malformations
5 y	M	Developmental delay	Complete corpus callosum agenesis Cortical dysplasia Periventricular heterotopia Right cerebellar hypoplasia
7 y	M	Familial mental retardation	Partial lissencephaly Partial corpus callosum agenesis
4 y	M	Previous repair of myelomeningocele	Chiari II Bilateral occipital polymicrogyria Partial corpus callosum agenesis
2 y	F	Seizures	Periventricular heterotopia Partial lissencephaly
14 y	M	Seizures	Complete corpus callosum agenesis Tubulonodular interhemispheric lipoma
3 y	M	Cerebral palsy Family history of epilepsy	Chiari II Bilateral occipital polymicrogyria Partial corpus callosum agenesis
2 y	M	Previous repair of myelomeningocele	Chiari II Schizencephaly-occipital Polymicrogyria/Pachygyria Periventricular heterotopia
3 d	F	Frontal swelling	Frontal meningocele Holoprosencephaly Heterotopia
14 mo	M	Seizures Developmental delay	Partial corpus callosum agenesis Dandy-Walker
1 mo	M	Dysmorphic	Partial lissencephaly Complete corpus callosum agenesis
1 y	M	Hypotonia Seizures	Partial corpus callosum agenesis Cortical dysplasia Periventricular heterotopia
3 y	F	Hypotonia Seizures Dysmorphic features	Partial lissencephaly Partial corpus callosum agenesis
6 y	M	Developmental delay	Partial lissencephaly Complete corpus callosum agenesis
5 mo	F	Occipital swelling	Occipital myelomeningocele Dandy-Walker
7 mo	F	Seizures Developmental delay	Periventricular heterotopia Polymicrogyria-frontal & occipital Complete corpus callosum agenesis
19 mo	F	Seizures	Complete corpus callosum agenesis Partial lissencephaly

Table 3. Description of 22 patients with corpus callosum malformation.

Age	Sex	Clinical history	Type of corpus callosum malformation	Other associated malformations
5 y	M	Developmental delay	Complete agenesis	Cortical dysplasia Periventricular heterotopia Right cerebellar hypoplasia
19 mo	F	Seizures	Complete agenesis	Partial lissencephaly
6 y	M	Developmental delay	Complete agenesis	Partial lissencephaly Interhemispheric cyst
7 mo	F	Seizures Developmental delay	Complete agenesis	Periventricular heterotopia Polymicrogyria
1 mo	M	Dysmorphic	Complete agenesis	Partial lissencephaly
14 y	M	Seizures	Complete agenesis	Interhemispheric lipoma
3 y	M	Developmental delay	Partial agenesis of body	Middle interhemispheric fusion
9 y	F	Developmental delay	Partial agenesis of body	Middle interhemispheric fusion
14 mo	M	Seizures Developmental delay	Partial agenesis of posterior ½ and rostrum	Dandy-Walker
4 y	M	Previous repair of myelomeningocele	Partial agenesis of posterior ½ and rostrum	Chiari II Polymicrogyria
3 y	M	Cerebral palsy Family history of epilepsy	Partial agenesis of posterior ½ and rostrum	Chiari II Polymicrogyria
1 y	M	Hypotonia Seizures	Partial agenesis of posterior ½ and rostrum	Cortical dysplasia Periventricular heterotopia
3 y	F	Hypotonia Seizures Dysmorphic features	Partial agenesis of posterior ½ and rostrum	Partial lissencephaly
7 y	M	Familial mental retardation	Partial agenesis of posterior ½ and rostrum	Partial lissencephaly
7 y	F	Seizures	Complete agenesis	-
2 mo	M	Seizures Developmental delay Dysmorphic features	Complete agenesis	-
2 y	F	Developmental delay	Complete agenesis	-
8 mo	M	Hypomelanosis of etto	Complete agenesis	-
2 y	M	Hypotonia Large head	Global hypoplasia	-
13 y	F	Seizures Multiple café au lait spots	Partial agenesis of posterior ½ and rostrum	-
1 y	F	Cerebral palsy	Partial agenesis of posterior ½ and rostrum	-
8 y	F	Developmental delay	Partial agenesis of posterior ½ and rostrum	-

Table 4. Types of corpus callosum malformation in 22 patients and frequency of the associated malformations.

Sex	N (%)
Male	13 (59)
Female	9 (41)
Type of corpus callosum malformation	
Complete agenesis	10 (45)
Partial agenesis of posterior 1/2 and rostrum	9 (41)
Partial agenesis of body	2 (9)
Global hypoplasia	1 (5)
Associated malformation	
Partial lissencephaly	5 (23)
Periventricular heterotopia	3 (14)
Polymicrogyria	3 (14)
Chiari II	2 (9)
Middle interhemispheric fusion	2 (9)
Cortical dysplasia	2 (9)
Interhemispheric lipoma	1 (5)
Dandy-Walker	1 (5)
Cerebellar hypoplasia	1 (5)

ting into the rare entity of middle interhemispheric fusion (syntelencephaly), which is a variant of holoprosencephaly. Of the 22 patients with corpus callosum malformation, 8 (36%) patients had the anomaly with no other associated cerebral malformations, while 14 (64%) patients had other associated malformations. In these 14 patients, the most commonly encountered cerebral malformations were partial lissencephaly (36%, n=5), periventricular heterotopia (21%, n=3), and polymicrogyria (21%, n=3) (Table 4). Only one patient with complete corpus callosum agenesis had an associated interhemispheric cyst and partial lissencephaly affecting the temporal lobes. Only one patient with corpus callosum malformation had an associated interhemispheric lipoma. Interhemispheric lipoma was seen in 4 other children, who otherwise had a completely normal corpus callosum. Two of these children were siblings with frontonasal dysplasia and both had two separate pericallosal lipomas in almost identical locations. One patient with a interhemispheric lipoma also had a normal corpus callosum and the full picture of Pai syndrome.

Neural tube closure defects

Chiari I malformation was seen in only one patient in association with Apert's syndrome. All patients with Chiari II malformation (12 patients) were imaged after repair of myelomeningocele and at an age older than 1 year in most of the cases. In 3 (25%) patients with Chiari II there was an associated cortical malformation in the form of polymicrogyria (PMG) in the region of the occipital lobes. Two of these patients had also dysgenetic corpus callosum. Only one patient with Chiari III was identified. Two of the four patients with dermoid cyst presented with meningitis. In three patients the dermoid was occipital in location. All four patients had an extracranial dermoid cyst with extension intracranially through a calvarial defect. Myelomeningocele was seen in 4 patients (2 frontal and 2 occipital). One patient with frontal myelomeningocele had also holoprosencephaly and gray matter heterotopia, and another patient with occipital myelomeningocele had the full picture of Chiari III malformation.

Cortical migrational defects

Cortical migrational defects were found in 23 (26.7%) patients, 17 of whom had lissencephaly and the remaining 4 gray matter heterotopia. Two patients had both lissencephaly and gray matter heterotopia. Table 5 shows the frequency of different types of lissencephaly with the partial lissencephaly being the most common (74%). Five (26%) patients of 19 with lissencephaly had associated corpus callosum dysgenesis of variable degree and one had a hypoplastic cerebellum and brainstem. All the six patients with gray matter heterotopia had one or more other associated cerebral malformations, which are summarized in Table 6.

Cortical organizational defects

Three patients were found to have disorders of cortical organization in the form of polymicrogyria (PMG). In only one of these patients was there also schizencephaly in association with cortical migrational defects (partial lissencephaly and periventricular heterotopia) as well as Chiari II malformation. The other two patients had an associated dysgenetic corpus callosum. All the six patients with non-balloon cell focal cortical dysplasia presented clinically with seizures. In four of these patients, the dysplastic cortex was in the temporal lobe with no other associated malformations. In the other two, there was an associated corpus callosum dysgenesis and nodular periventricular heterotopia.

Neurocutaneous syndromes

Among the nine patients with neurofibromatosis, three had optic pathway glioma and one had plexiform neurofibroma involving the neck, face, and tongue. Seven of the 9 patients demonstrated areas of high signal intensity on T2WIs, the so-called unidentified bright objects (UBOs), in the cerebellum, brainstem, and basal ganglia representing myelin vaculization. Four patients had Sturge-Weber syndrome; two also had the clinical features of Klippel-Traunany syndrome. All these four patients were males. Tuberous sclerosis was seen in only one patient.

Posterior fossa malformations

Of the three patients with Joubert syndrome, two were siblings. All the three patients presented with abnormal eye movement, hypotonia, and episodic apnea-hyperpnea. On imaging, all three patients demonstrated thick and horizontal superior cerebellar peduncles, rostrally deviated fastigium causing an abnormal shape of the 4th ventricle, absent superior cerebellar peduncle decussation in the midbrain, a shallow pontomesencephalic junction with a positive molar tooth sign, and vermian agenesis. In one of the siblings, there was a retrocerebellar cyst with no communication with the fourth ventricle. Four patients had cerebellar hypoplasia and one patient had dorsal brainstem malformation. Dandy-Walker malformation was seen in three patients.

Holoprosencephaly

Two of the three patients with holoprosencephaly had middle interhemispheric fusion (syntelencephaly). In these two patients part of the body of the corpus callosum was absent while other portions of corpus callosum were normal. The third patient with holoprosencephaly had the semilobar type associated with frontal meningocele.

Table 5. Types of lissencephaly in 19 patients.

Types of lissencephaly	Number of patients (%)
Classical-Complete	3 (16%)
Classical-Partial	14 (74%)
Cobblestone	1 (5%)
Bilateral perisylvian	1 (5%)

Table 6. Cerebral malformations seen in 6 patients with gray matter heterotopia.

Cerebral malformation	Number of patients (%)*
Corpus callosum dysgenesis	3 (50%)
Cortical dysplasia	2 (33%)
Polymicrogyria	2 (33%)
Lissencephaly	2 (33%)
Chiari II	1 (17%)
Schizencephaly	1 (17%)
Cerebellar hypoplasia	1 (17%)
myelomeningocele	1 (17%)
Holoprosencephaly	1 (17%)

* Some patients had more than one cerebral malformation associated with gray matter heterotopia.

Discussion

Magnetic resonance imaging (MRI) has had an important impact on the study and understanding of congenital cerebral malformations. While autopsy and pathologic analysis reveals a lot about severe cerebral malformations, MRI allows for study of the entire spectrum of such malformations, from mild to severe.[1] Moreover, MRI permits multiple cuts in multiple planes at multiple occasions, providing a better understanding of the temporal evolution of these diseases.[1] Because most brain structures develop at about the same time during fetal life, it is common to see multiple anomalies in association.[2] Hence, one case with multiple anomalies may fit into many classes of cerebral malformations.

Holoprosencephaly is a rare cerebral malformation (forebrain dysgenesis) in which there is lack of separation of the cerebral hemispheres due to failure of induction of the basal forebrain and middle part of the face.[3,4] Holoprosencephaly has been traditionally classified into lobar, semilobar, and alobar types according to severity, with the alobar type the most severe. Lack of separation of thalami and subsequent lack of formation of the third ventricle, absent falx and corpus callosum, and fusion of basal ganglia are the imaging signs of holoprosencephaly with the severity and conspicuity according to the severity of the disease. Patients with this anomaly have no specific clinical signs; however, maternal diabetes is a known risk factor. There was only one patient with semilobar holoprosencephaly identified in this study with associated frontal meningocele and gray matter heterotopia. Association with meningocele is rare. The reason for the small number of cases of holoprosencephaly is probably that most of these patients

are diagnosed on CT and shunted without performing MRI. The middle interhemispheric variant of holoprosencephaly (MIH), sometimes referred to as syntelencephaly, was recognized in two patients in this study. MIH was first described in 1993[3] and is considered a very rare anomaly characterized by an abnormal midline connection of the cerebral hemispheres in the parietal and posterior frontal regions, with interhemispheric separation in the basal forebrain, occipital, and anterior frontal lobes.[4] In this type of anomaly, there is an unusual callosal dysgenesis in the form of an absent body and preserved genu and splenium.[5]

Intracranial lipomas result from abnormal differentiation of the meninx primitiva into fat. They reside in the subarachnoid space and the most common location is the interhemispheric fissure (Figure 1).[2] Two patients with intracranial lipoma identified in this study were siblings with complex facial anomalies in the form of frontonasal dysplasia for which they underwent several maxillofacial surgeries. Each one of these two patients had two separate pericallosal lipomas in identical locations, the first to be reported in the literature.[6] Another patient in this series with pericallosal lipoma had the full clinical picture of Pai syndrome, which has also been reported as the fifth case in the world literature.[7] In addition to the intracranial lipoma, patients with Pai syndrome also have a median cleft in the upper lip and cutaneous polyps.

Joubert syndrome is a non-progressive familial autosomal recessive disease characterized by an abnormal respiratory pattern, abnormal eye movement, ataxia, and developmental delay. Patients have brainstem and vermian malformation. Because of its autosomal recessive inheritance, Joubert syndrome is more common in consanguineous marriages.[8] Two of three patients in this study were siblings. The clinical diagnosis of this syndrome may be at times difficult since it shares features with several other conditions.[9] The radiological diagnosis requires a high index of suspicion and scrutiny in assessment of posterior fossa structures. The clinical and radiological presentations of the three patients with Joubert syndrome in this study were similar to those noticed in previously reported cases from Saudi Arabia.[10,11] The molar tooth sign is the hallmark of Joubert syndrome on imaging and results from a combination of three malformations: 1) an abnormally deep and wide interpeduncular cistern, 2) thickened and horizontally superior cerebellar peduncles, 3) and hypoplasia of the vermis.[12] The conspicuity of the sign on neuroimaging depends on the severity of these three anomalies. In a group of 45 patients, molar tooth sign was present in 82% of cases and was the only intracranial abnormality in 66%.[12] Molar tooth sign was present in all three patients in this study.

The posterior half of the body and the splenium are usually absent in cases of partial corpus callosum agenesis because the corpus callosum does not form simultaneously. The initial axons of corpus callosum cross the midline at a point on the line joining the anterior commissure and mamillary bodies, then the axons anterior and posterior to this point cross.[13] Atypical cases of corpus callosum agenesis not following this sequence are usually related to holoprosencephaly (see the discussion later). Anomalies of the corpus callosum are often associated with other cerebral malformations;[14] the commonest in this study were anomalies of neuronal migration (lissencephaly, 23% and heterotopia, 14%) and disorders of cortical organization (polymicrogyria, 14%) (Table 4, Figure 2). The entity of corpus callosum agenesis with interhemispheric cyst, which is most commonly seen in boys,[2] was found in only one patient in this series. This patient was a 6-year-old boy and he had a migrational anomaly in the form of partial lissencephaly affecting both temporal lobes. Although corpus callosum dysgenesis might be an incidental finding on imaging, all the patients with this anomaly in the study group had neurological symptoms. Asymptomatic corpus callosum anomalies are more frequent in adult patients.

The neurocutaneous syndromes are probably underrepresented in this study for two reasons: first, some of these syndromes present at an age older than 15 years; examples are neurofibromatosis and tuberous sclerosis, and they were excluded from this study. Second, other neurocutaneous syndromes may be diagnosed by CT only, for example Sturge-Weber syndrome (Figure 3) and tuberous sclerosis, and excluded from this study if no MRI has been done for patients with these diseases. Neurofibromatosis type 1 (NF1) is an autosomally dominant disease initially described by von Ricklinghausen in 1882. The gene defect in this disease is in the long arm of chromosome 17.[2] Optic pathway glioma is the most common tumor complicating this disease, with an incidence as high a 15%, but about half are asymptomatic.[14] These gliomas are most commonly low grade and can affect both optic nerves. The bright T2 foci in the white matter seen in these patients is the typical myelin vacuolization, which is seen after the age of 2 years in patients with NF1 and disap-

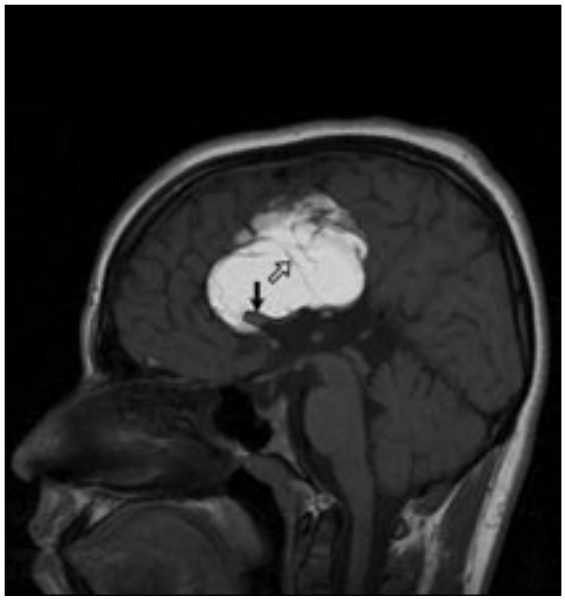

Figure 1. Sagittal T1WI of the brain of a 14-year-old boy with refractory seizures demonstrating large interhemispheric lipoma of the tubulonodular type associated with almost complete corpus callosum agenesis. Only the genu and part of the anterior body of corpus callosum are seen (arrow). Typical vascular structures coursing through the lipoma (open arrow) indicate its origin from the subarachnoid space.

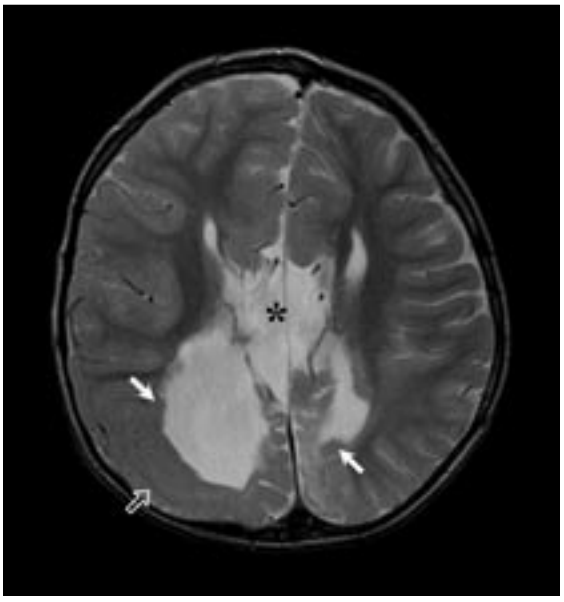

Figure 2. Axial T2WI of the brain of a 5-year-old boy with developmental delay showing the absence of the corpus callosum allowing the third ventricle to have a superior extension (asterisk) and continuation with the interhemispheric fissure, and showing parallel orientation of the lateral ventricles. Nodular gray matter heterotopia is seen in the periventricular area (arrows). The right occipitoparietal area shows a thick cortex and lack of a sulci, indicating cortical dysplasia (open arrow).

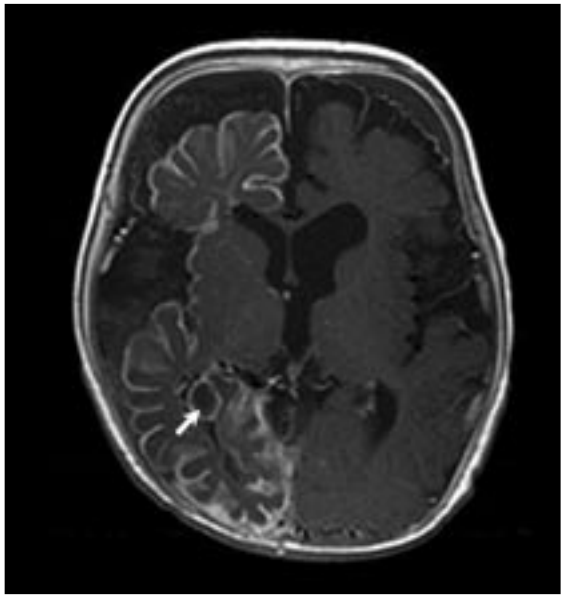

Figure 3. 10-month-old boy with body and face nevus and seizures. Enhanced axial T1WI of the brain showing remarkable atrophy of both cerebral hemispheres and gyriform enhancement over the entire right hemisphere. Note the enlargement of the ipsilateral choriod plexus (arrow) that is typically seen in cases of Sturge-Weber syndrome.

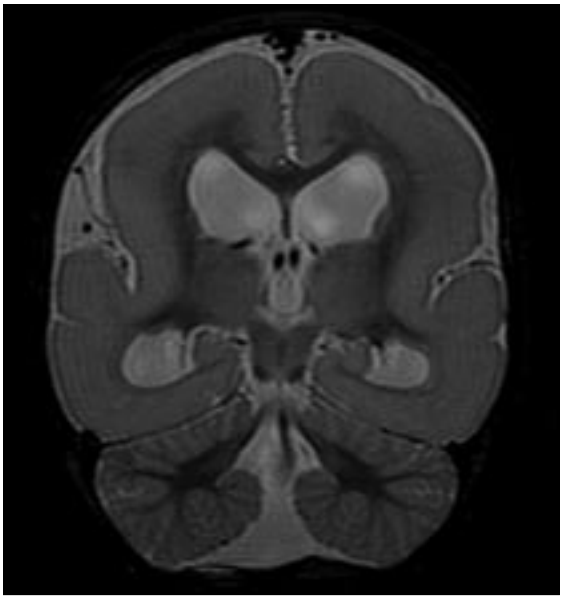

Figure 4. Coronal T2WI of the brain of 1-year-old girl with seizures, demonstrating the classical findings in complete lissencephaly. The cerebral cortex is remarkably thick and lacks gyri (smooth surface), the gray/white matter interface is smooth, and the sylvian fissures are underdeveloped. This appearance is very similar to the immature fetal brain.

pears after the age of 12 years.[15] The predilection of these lesions to cerebellum, brainstem and basal ganglia is in keeping with the distribution described in the literature.[16] Characteristically, the areas of myelin vacuolization do not enhance; however, it is not unusual for these lesions to increase in size and number between the age of 2 to 12 years and they should not be mistaken for a neoplasm.

During the eighth fetal week the neurons start migrating from the germinal zone to the cortex. Several etiological factors have a role in interruption of neuronal migration, resulting in variable degrees of migrational abnormalities due to arrest of neurons at a distance short of the normal location of the cortex. In type 1 (classical) lissencephaly (Figure 4), the brain surface is completely smooth (agyria) or has broad, flat gyri separated by a few, shallow sulci (pachygyria).[1] In addition, imaging studies show a thick cortex and shallow, vertically oriented sylvian fissures (lack of opercularization) giving the cerebrum the shape of a figure of eight.[17] More than one gene defect has been proved responsible for classical lissencephaly. In some types of classical lissencephaly, the mothers and sisters of the affected patients have another type of migrational defect (band heterotopia or double cortex).[18] On the other hand, the previously termed type 2 lissencephaly is no longer considered lissencephaly, but a brain malformation that forms part of the cobblestone complex that is often seen in association with muscular dystrophy.[1] Only one patient was identified in this series with this type of cerebral malformation. Gray matter heterotopia refers to normal neurons in abnormal location. Patients with heterotopia almost always present with epilepsy, the severity and prognosis of which is a function of the severity of heterotopia.[17] The heterotopic gray matter may be located subependymally, subcortically, or between the ventricles and cortex. The heterotopic islands may have a nodular or band configuration and their signal intensity on MRI follows that of the cerebral cortex on all pulse sequences.[19] These lesions are not surrounded by edema and do not enhance.

Among the neural tube closure disorders identified in this study are the meningoencephaloceles, Chiari malformations, and dermoid. Cephaloceles are extracranial extensions of intracranial structures through a skull defect. Classically, the herniated intracranial structures remain connected to the intracranial ones through the skull defect, but in rare cases this connection might be lost and the cephalocele is then called a sequestrated cephalocele or meningocele.[20] Cephaloceles may be isolated anomalies, may be associated with other anomalies, or may be part of a syndrome.[17] The association between holoprosencephaly and cephalocele that has been identified in one patient in this series is an extremely rare association.[21] The hallmark of Chiari I malformation is the cerebellar tonsillar herniation below foramen magnum. This malformation results in hydrocephalus and sometimes syringomyelia, but is usually not associated with other cerebral malformations. Chiari II malformation on the other hand is virtually always associated with lumbar myelomeningocele, and frequently cerebral malformations. In this study the most commonly associated cerebral malformations with Chiari II were polymicrogyria and corpus callosum dysgenesis. The hallmark of Chiari II is the small posterior fossa due to a low tentorial insertion leading to herniation of cerebellum superiorly above the tentorium and inferiorly below the foramen magnum. Several other changes occur in the posterior fossa secondary to this cerebellar herniation, such as cerebellar creeping around the brainstem, downward displacement of the medulla oblongata, elongation of the fourth ventricle, and concavity of the clivus. Chiari III malformation is an extremely rare condition[2] and only one case was identified in this study. In this malformation there is, in addition to the usual Chiari II changes, posterior herniation of the cerebellum and sometimes of the brainstem by spina bifida at the C1 or C2 level. Dermoid results from improper disjunction of neuroectoderm from cutaneous ectoderm during the third or fourth week of gestation (ectodermal heterotopia). Dermoid may be associated with dermal sinus and skull defects, which were noticed in all four patients in this study. In these case, the patient may present with meningitis.

When neurons reach the cortex area but fail to develop into normal gyri the condition is referred to as a disorder of cortical organization, which includes polymicrogyria, schizencephaly, and focal cortical dysplasia. Disorders of cortical organization may be focal or diffuse. There is a lack of normal gyral formation with a thick cortex. Congenital bilateral perisylvian syndrome is a familial condition characterized by polymicrogyria involving the cortex for a variable extent around the sylvian fissures.[22] Imaging of polymicrogyria requires careful selection of MR pulse sequences to adequately identify the gyral abnormality, which is frequently missed on standard pulse sequences. The 3-dimentional SPGR sequence has proved its role in imaging such conditions.

In conclusion, a predominance of neural tube closer defects, cortical migrational abnormalities, and corpus callosum anomalies has been demonstrated in this study, and the results are similar to what has been observed in other parts of the world. Although most congenital cerebral malformations followed the usual and commonly described pattern and appearance, some did not. Unusual patterns and associations were found, which required further follow-up imaging, neurological evaluation, and genetic workup and counseling.

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