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IMAGING SPECTRUM IN PEDIATRIC CASES WITH CEREBRAL HEMIATROPHY: A BRIEF REVIEW OF LITERATURE

Radio-Diagnosis

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ABSTRACT

Childhood pediatric hemi atrophy is a rare imaging finding with various etiologies (congenital and acquired) which can further be clinically classified to progressive and non-progressive causes. Here, we describe MRI and CT imaging findings in 6 pediatric cases of cerebral hemiatrophy presenting with contralateral hemiplegia, refractory seizures, developmental delay and cognitive impairment, all of which underwent MRI brain study and CT brain study in few cases. A clinico-radiological approach plays a major role in early diagnosis of cerebral atrophy and its causes, which in turn helps in early treatment and a better prognosis.

KEYWORDS

Cerebral Hemiatrophy, Rasmussen Encephalitis, Dyke-Davidoff-Masson syndrome, Sturge-Weber syndrome, treatment

INTRODUCTION

Childhood cerebral hemiatrophy is an uncommon neuroimaging finding with many different etiologies which can be congenital (genetic or in-utero insults) or acquired (as a result of postnatal cerebrovascular causes, inflammatory process, or traumatic). Clinical features are often a result of contralateral hemiplegia, refractory seizures, cognitive impairment, emotional and behavioral issues.

A clinical approach to etiologies of cerebral hemiatrophy classifies it into a non-progressive (presumed single insult) or a progressive (ongoing insult) etiology. Non progressive causes are ischemic or hemorrhagic stroke, cranial trauma, cranial irradiation. Progressive causes include Sturge Weber syndrome, Parry Romberg syndrome, Rasmussen encephalitis, germinoma-associated cerebral hemiatrophy, vascular malformations, status epilepticus including hemiconvulsion-hemiplegia-epilepsy syndrome.

Table 1: Approach to imaging features in cases of cerebral hemiatrophy (Referred from Tan AP et al. Figure 1& 2)

Imaging features	Etiology
1) Distribution of atrophy	
• *Parieto-occipital*	SWS
• *Perisylvian fronto-temporal*	DDMS
• *MCA territory*	Arterial ischemic stroke
2) Intracranial calcification	
• *Gyriform/subcortical (tram-track predominance)*	SWS
• *Discrete focal*	PRS
3) Leptomeningeal enhancement +	SWS (almost always +) PRS
4) Ipsilateral white matter signal abnormalities	RE
5) Atrophy of ipsilateral caudate nucleus	RE
6) Mass like lesion with enhancing solid component	Off midline germinoma
7) vascular anomalies	Congenital ICA hypoplasia Moya Moya disease
8) Ipsilateral calvaria thickening, enlargement of paranasal sinuses & elevated greater wing of the sphenoid	DDMS

SWS: Sturge-Weber syndrome, DDMS: Dyke-Davidoff- Masson syndrome, RE: Rasmussen encephalitis, PRS: Parry-Romberg syndrome

Case Study

Case 1: A 3-years-old female child came with complaints of right sided focal seizure and weakness since 1 year. There was history of fever and vomiting 1 year back following which the seizure started along with regression of milestones. Birth history was normal. MRI brain (refer figure 1) showed marked left cerebral hemiatrophy involving cortex, white and deep grey matter and insular cortex with encephalomalacia and marked gliotic changes with ex vacuo dilatation of ipsilateral lateral ventricle. Left caudate nucleus was also mildly atrophied. However, no leptomeningeal enhancement was noted in post contrast study. On history and MRI correlation, a diagnosis of Rasmussen encephalitis was given.

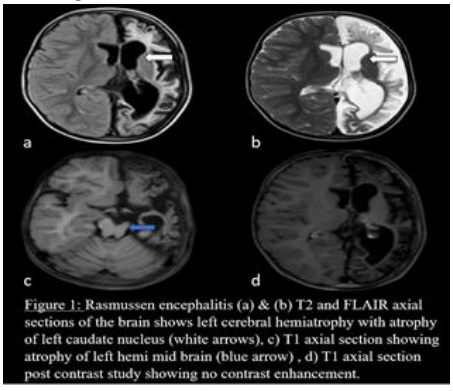

Figure 1: Rasmussen encephalitis (a) & (b) T2 and FLAIR axial sections of the brain shows left cerebral hemiatrophy with atrophy of left caudate nucleus (white arrows), c) T1 axial section showing atrophy of left hemi mid brain (blue arrow) , d) T1 axial section post contrast study showing no contrast enhancement.

Case 2: 5 years old female child came with complaints of right sided focal seizure with right hemiparesis since 1 year. Past history reveals fever with altered sensorium 1 year back following which the symptoms started. Birth history was normal. No developmental delay. MRI brain (refer figure 2) revealed left cerebral hemiatrophy involving the left fronto-parietal lobes with focal gliotic chnages and encephalomalacia in the region and ipsilateral lateral ventricle dilatation. In addition, mild left caudate nucleus atrophy was also noted. However, there was no compensatory hypertrophy of the bony calvaria and hyperpneumatization of paranasal sinuses. Based on history and MRI correlation, a diagnosis of Rasmussen encephalitis was given.

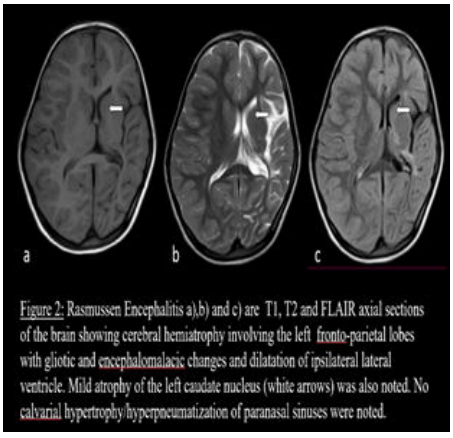

Figure 2: Rasmussen Encephalitis a),b) and c) are T1, T2 and FLAIR axial sections of the brain showing cerebral hemiatrophy involving the left fronto-parietal lobes with gliotic and encephalomalacic changes and dilatation of ipsilateral lateral ventricle. Mild atrophy of the left caudate nucleus (white arrows) was also noted. No calvarial hypertrophy/hyperpneumatization of paranasal sinuses were noted.

Case 3: A 15 years old female child with seizure disorder presented

with GTCS type of seizure lasting for 5-10 mins. 1st episode was at 3 months of age. The child is on antiepileptic treatment since then. No history of weakness of limbs was given. Birth history was normal with no history of NICU admission. There is history of developmental delay. No records of previous imaging. MRI brain was done (refer figure 3) and showed right cerebral hemiatrophy predominantly involving the right frontal and anterior temporal lobes with T2/FLAIR hyperintensity involving cortical and subcortical white matter causing prominence of sulcal spaces, right sylvian fissure and CSF spaces with exvacuo dilatation of right lateral ventricular system. Diffuse thinning of body of corpus callosum was also noted. In addition, ipsilateral thickening of calvaria on right side & hyperpneumatization of right frontal sinus were also noted.

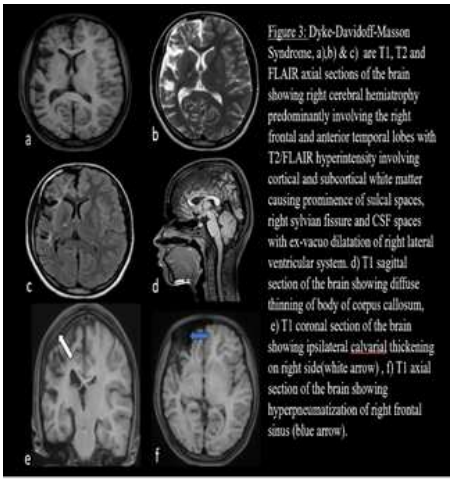

Figure 3: Dyke-Davidoff-Masson Syndrome, a)b) & c) are T1, T2 and FLAIR axial sections of the brain showing right cerebral hemiatrophy predominantly involving the right frontal and anterior temporal lobes with T2/FLAIR hyperintensity involving cortical and subcortical white matter causing prominence of sulcal spaces, right sylvian fissure and CSF spaces with ex-vacuo dilatation of right lateral ventricular system. d) T1 sagittal section of the brain showing diffuse thinning of body of corpus callosum, e) T1 coronal section of the brain showing ipsilateral calvarial thickening on right side(white arrow) , f) T1 axial section of the brain showing hyperpneumatization of right frontal sinus (blue arrow).

Case 4: 10-years-old female child was referred for MRI brain with left sided focal seizure. 1st episode was noted at the 1 month of age, following which there was hospital admission for 3 days. Birth history and milestones were normal. However, parents give history of poor academic performance. MRI brain (refer figure 4) showed diffuse right sided cerebral hemiatrophy with gliotic changes and dilatation of right lateral ventricle with prominence of cortical sulci. Thickening of adjacent calvaria with hyperpneumatization of right maxillary sinus and left mastoid were noted. Leptomeningeal enhancement was not seen on post contrast study. A diagnosis of Dyke-Davidoff-Masson syndrome was given on clinico-radiological correlation.

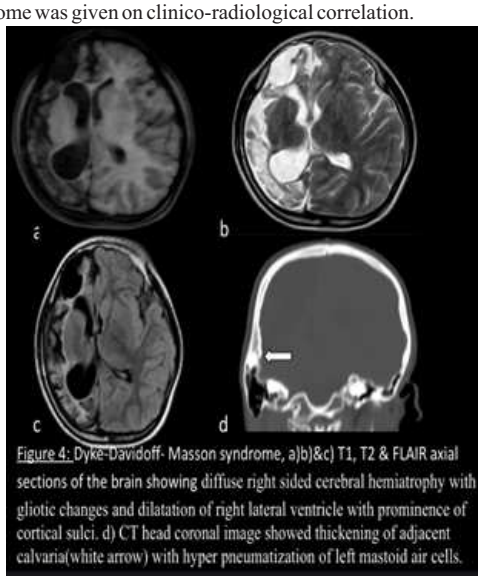

Figure 4: Dyke-Davidoff- Masson syndrome, a)b)&c) T1, T2 & FLAIR axial sections of the brain showing diffuse right sided cerebral hemiatrophy with gliotic changes and dilatation of right lateral ventricle with prominence of cortical sulci. d) CT head coronal image showed thickening of adjacent calvaria(white arrow) with hyper pneumatization of left mastoid air cells.

Case 5: A 4 years old female child presented with left sided tonic-clonic seizure. Birth history and developmental history were normal. On examination, port wine stain noted in right forehead. On MRI brain (refer figure 5), there is mild focal atrophy of right occipital lobe along with T2/FLAIR hyperintensities in temporooccipital lobes showing significant post contrast enhancement.Sulcal enhancement noted in bilateral frontal and left parieto-occipital lobes. On CT, foci of hyperdensity noted in cortical region of right occipital lobe- s/o calcification. On correlating, clinical & MRI findings, a diagnosis of Sturge Weber syndrome was given.

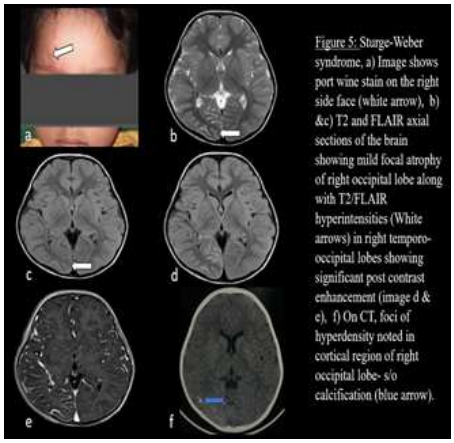

Figure 5: Sturge-Weber syndrome, a) Image shows port wine stain on the right side face (white arrow), b) &c) T2 and FLAIR axial sections of the brain showing mild focal atrophy of right occipital lobe along with T2/FLAIR hyperintensities (White arrows) in right temporo-occipital lobes showing significant post contrast enhancement (image d & e), f) On CT, foci of hyperdensity noted in cortical region of right occipital lobe- s/o calcification (blue arrow).

Case 6: A 16 days old female baby was referred with history of 2 episodes of seizure, focal type involving the right upper limb and head Birth history was normal. On examination, port wine stain noted on the left side of face since birth. MRI brain (refer figure 6) was requested and showed left sided cerebral hemiatrophy with widening of sulcal and CSF spaces. On post contrast study, lepto-meningeal enhancement was noted on the left side. Based on clinical history, examination and MRI study, it was diagnosed as a case of Sturge weber syndrome.

Figure 6: Sturge Weber syndrome, a) Image shows port wine stain on left side of face, b) &c) T2 and FLAIR axial sections of brain showing left sided cerebral hemiatrophy and d) T1 post contrast axial section showing leptomeningeal enhancement on left side.

DISCUSSION:

Cerebral hemiatrophy can be of congenital and acquired etiologies. Furthermore, to link clinical and neuroimaging findings and aid in management and avoiding further investigations, the etiologies are further classified to progressive and non-progressive causes. Progressive cerebral atrophy requires urgent management to delay progression of the disease and a have better prognosis.

Rasmussen encephalitis is a chronic inflammatory neurodegenerative disease usually affecting one cerebral hemisphere and leading to progressive neurological deficits and refractory seizures. Etiology is unclear, however, cytotoxic T cell reaction against the neurons was considered in the pathogenesis. Clinical examination, EEG and imaging helps in diagnosis and assessing progression of the disease. Cortical swelling is usually the first finding with with T2/FLAIR hyperintensity with progression across the hemisphere. Later on, cerebral hemiatrophy with widened cortical sulci and ipsilateral ventricle dilatation is noted and finally, there is disappearance of the increased signal, leaving a marked atrophy of one cerebral hemisphere and mild ipsilateral caudate nucleus atrophy. Some of these findings were noted in our patients (Figure 1 & 2).

Dyke-Davidoff-Masson syndrome is characterized by cerebral hemiatrophy/hypoplasia due to brain insult in-utero or early childhood associated with compensatory osseous hypertrophy. Clinical features

can vary in severity and includes contralateral hemiparesis, refractory seizures, behavioral/cognitive impairment. The diagnosis depends on imaging findings on CT and MRI which includes atrophy of one cerebral hemisphere & dilated ipsilateral lateral ventricle, ipsilateral thickening of the calvaria , hyperpneumatization of paranasal sinuses, mastoid air cells and elevation of greater wing of sphenoid and petrous ridge. Our patients presented with many of these findings (Fig 3 & 4).

Sturge-Weber syndrome is a phakomatosis associated with neurological abnormalities including seizures, hemiplegia, headache, developmental delays. Imaging findings includes leptomeningeal enhancement on CE-MRI of the brain, cerebral hemiatrophy, cortical/subcortical calcification, ipsilateral enlarged choroid plexus. Many of these abnormalities were demonstrated in the imaging of our patients (Figure 5 & 6).

CONCLUSIONS

Cerebral hemiatrophy is a rare but dangerous neuroimaging findings of different etiologies. Correct diagnosis and differentiation based on clinical examination, EEG and neuroimaging between the progressive and non-progressive disorders of cerebral hemi atrophy can aid in timely early intervention leading to better prognosis of the patients and also avoiding further investigations.

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