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KEY WORDS: Cerebral hemiatrophy; Magnetic resonance imaging; Rasmussen encephalitis; Dyke-Davidoff-Masson syndrome.

NEUROIMAGING SPECTRUM OF CEREBRAL HEMIATROPHY ON MRI: A CLINICORADIOLOGICAL STUDY

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ABSTRACT

Background Cerebral hemiatrophy is an uncommon clinicoradiological entity characterized by unilateral cerebral volume loss resulting from diverse congenital and acquired etiologies. Patients commonly present with seizures, hemiparesis, developmental delay, and cognitive impairment. Magnetic resonance imaging (MRI) plays a pivotal role in identifying the underlying etiology and associated structural abnormalities. **Materials and Methods** This retrospective clinicoradiological study included ten patients with cerebral hemiatrophy who underwent MRI evaluation at a tertiary care centre. Clinical presentation and neuroimaging findings were analyzed to determine the underlying etiology and imaging spectrum. MRI was performed using a 1.5-T scanner and included T1-weighted, T2-weighted, FLAIR, DWI/ADC, and SWI sequences. Post-contrast imaging was performed when clinically indicated. **Results** A total of ten patients with cerebral hemiatrophy were included in the study. The age of patients ranged from 4 to 36 years. Seizures were the most common presenting symptom and were observed in all patients. Hemiparesis and developmental delay were also frequent clinical manifestations. The etiological distribution included Rasmussen encephalitis (n = 4), Sturge-Weber syndrome (n = 3), and Dyke-Davidoff-Masson syndrome (n = 3). MRI demonstrated unilateral cerebral volume loss in all patients, with associated ex-vacuo ventricular dilatation being the most consistent imaging finding. Additional findings included cerebral peduncular atrophy, calvarial thickening, hyperpneumatization of paranasal sinuses, leptomeningeal vascular malformations, and cortical calcifications depending on the underlying etiology. **Conclusion** MRI is indispensable in the evaluation of cerebral hemiatrophy, enabling differentiation among various etiologies based on characteristic imaging patterns and facilitating appropriate clinical management.

Table 1: Etiological distribution of cerebral hemiatrophy

Etiology	Number of Cases
Rasmussen encephalitis	4
Sturge-Weber syndrome	3
Dyke-Davidoff-Masson syndrome	3
Total	10

Table 2: Major MRI findings

MRI Finding	Number of Cases
Cerebral hemiatrophy	10
Ex-vacuo ventricular dilatation	10
Cerebral peduncular atrophy	5
Calvarial hickening/hyperpneumatization	3
Leptomeningeal vascular malformations	3
Cortical calcifications	3

INTRODUCTION

Cerebral hemiatrophy refers to atrophy of one cerebral hemisphere in the presence of a relatively normal contralateral hemisphere. It may occur secondary to congenital or acquired insults affecting the developing brain. Congenital causes include intrauterine vascular injury and developmental anomalies, whereas acquired causes include perinatal hypoxia, infection, trauma, ischemia, inflammatory disorders, and prolonged seizures. The clinical manifestations vary depending on the timing and extent of cerebral injury, with seizures, hemiparesis, developmental delay, cognitive impairment, and speech disturbances being the most common presentations.

Magnetic resonance imaging (MRI) serves as the imaging modality of choice for evaluating cerebral hemiatrophy due to its excellent soft-tissue resolution and ability to characterize associated abnormalities. Characteristic MRI findings such as unilateral cerebral volume loss, ex-vacuo ventricular dilatation, calvarial thickening, hyperpneumatization of paranasal sinuses, cerebral peduncular atrophy, leptomeningeal angiomatosis, and cortical calcifications assist in narrowing the differential diagnosis and determining the underlying etiology. MRI not only characterizes the extent of hemispheric atrophy but also identifies disease-specific imaging patterns that aid in etiological diagnosis and prognostication.

Dyke-Davidoff-Masson syndrome (DDMS) is characterized by cerebral hemiatrophy associated with compensatory osseous changes resulting from early-life cerebral insult. Rasmussen encephalitis is a chronic inflammatory disorder characterized by progressive hemispheric atrophy and medically refractory epilepsy. Sturge-Weber syndrome (SWS) is a neurocutaneous disorder characterized by leptomeningeal angiomatosis and facial capillary malformations.

The present clinicoradiological study describes the MRI spectrum of cerebral hemiatrophy in ten patients with varied etiologies, including DDMS, Rasmussen encephalitis, and SWS, highlighting the role of MRI in accurate diagnosis and management.

CASE 1: A 12-year-old girl presented with recurrent focal seizures involving the right side of the body since 5 years of age. There was a history suggestive of perinatal hypoxic insult requiring neonatal intensive care admission after birth. She gradually developed weakness of the right upper and lower limbs with difficulty in performing daily activities. Neurological examination revealed spastic right hemiparesis with exaggerated deep tendon reflexes and extensor plantar response. MRI demonstrated left cerebral hemiatrophy with encephalomalacic changes and ex-vacuo dilatation of the left lateral ventricle. Associated ipsilateral calvarial thickening and hyperpneumatization of the left frontal sinus and bilateral mastoid air cells were noted. Based on the clinical and imaging findings, a diagnosis of Dyke-Davidoff-Masson syndrome was established.

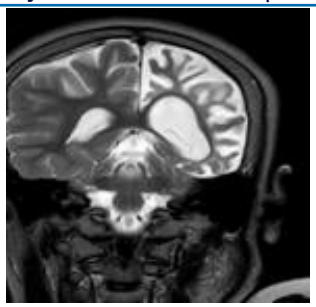


Fig 1:T2 coronal

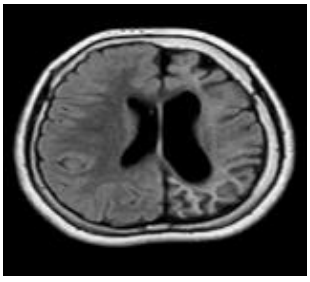


Fig 2:FLAIR axial

T2 coronal and FLAIR axial images show left cerebral atrophy with hyperintense leukomalacia and ex-vacuo dilatation of left lateral ventricle.

CASE 2: A 25-year-old male with long-standing seizure disorder and intellectual disability presented with generalized tonic-clonic seizures. MRI revealed marked left cerebral hemiatrophy with surrounding gliosis and widening of cortical sulci. Ex-vacuo dilatation of the left lateral ventricle was present along with atrophy of the left cerebral peduncle and bilateral caudate nuclei. Compensatory changes included ipsilateral calvarial thickening and hyperpneumatization of the frontal, sphenoid, and mastoid air cells. The characteristic imaging findings were consistent with Dyke-Davidoff-Masson syndrome.

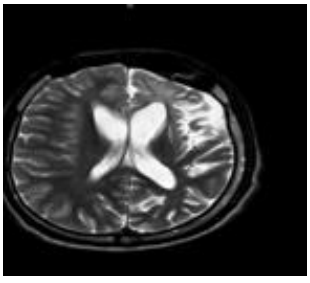


Fig 3:T2 axial

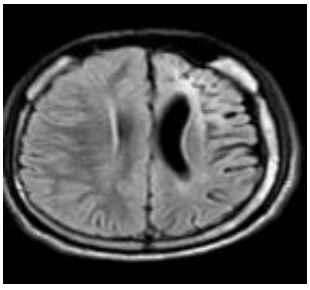


Fig 4:FLAIR axial

T2 axial and FLAIR axial images show unilateral atrophy with surrounding gliosis & widening of cortical sulci of the left cerebral hemisphere and ex-vacuo dilatation of the left lateral ventricle.

CASE 3: An 11-year-old female presented with recurrent focal seizures and weakness of the right upper and lower limbs. Neurological examination revealed right-sided hypertonia, hyperreflexia, and reduced motor power. MRI demonstrated diffuse gyral thickening involving the left cerebral hemisphere with T2/FLAIR hyperintensity and diffusion restriction, associated with ipsilateral ventricular prominence. EEG revealed epileptiform activity. Based on the clinical presentation and imaging findings, a diagnosis of Rasmussen encephalitis was established.

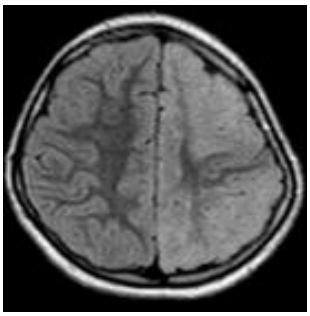


Fig 5: FLAIR axial

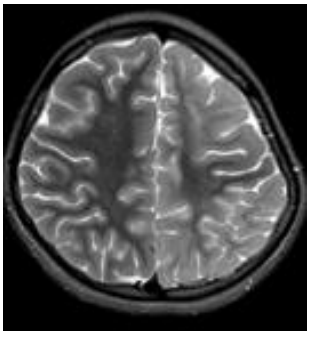


Fig 6:T2 axial

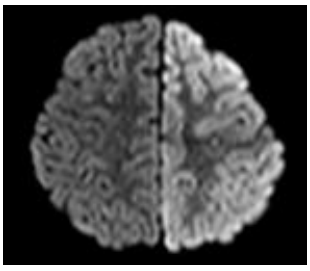
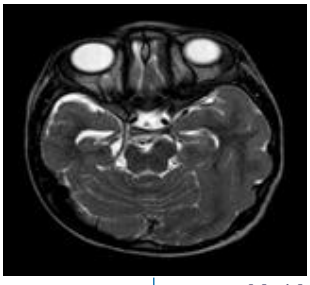


Fig 7:DWI axial

Diffuse gyral thickening involving left cerebral hemisphere showing T2/FLAIR hyperintense signal with associated diffusion restriction.

CASE 4: An 8-year-old boy presented with recurrent focal seizures involving the left side of the body, developmental delay, and progressive gait abnormality. MRI demonstrated right cerebral hemispheric atrophy with mild ex-vacuo dilatation of the temporal and occipital horns of the right lateral ventricle, along with atrophy of the right cerebral peduncle. The progressive clinical course and characteristic imaging findings favoured a diagnosis of Rasmussen encephalitis.



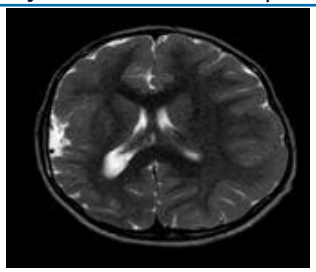


Fig 8 and 9:T2 axial images

T2 axial images showing atrophy of right cerebral hemisphere with mild dilatation of temporal and occipital horn of right lateral ventricle with associated atrophy of right cerebral peduncle noted.

CASE 5: A 6-year-old girl presented with recurrent seizures since infancy and delayed developmental milestones. Clinical examination revealed a right-sided facial port-wine stain involving the ophthalmic division of the trigeminal nerve. MRI demonstrated atrophy of the right cerebral hemisphere with enlargement of the ipsilateral choroid plexus. Susceptibility-weighted imaging revealed prominent vascular channels traversing the right cerebral white matter with focal blooming in the right parietal region suggestive of cortical calcifications. These findings were consistent with Sturge-Weber syndrome.

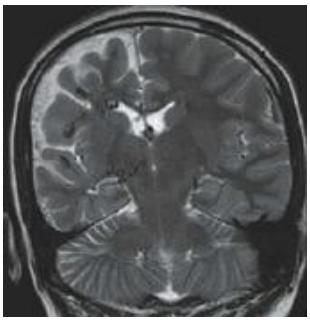


Fig 10:T2 coronal

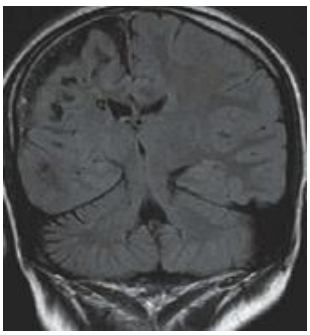


Fig 11: FLAIR coronal

Coronal T2 and FLAIR images show volume loss involving the right parietal lobe with prominence of the overlying cortical sulci and mild ex-vacuo dilatation of the adjacent right lateral ventricle. Multiple serpiginous cortical and extra-axial vascular structures are noted over the right frontoparietal convexity.

CASE 6: A 14-year-old male presented with recurrent focal seizures involving the right side of the body with progressive right-sided weakness. MRI demonstrated mild atrophy of the left cerebral hemisphere with ex-vacuo dilatation of the left lateral ventricle and associated atrophy of the left cerebral peduncle. Absence of compensatory calvarial changes favored an acquired etiology. Based on the clinical and imaging findings, a diagnosis of Rasmussen encephalitis was established.

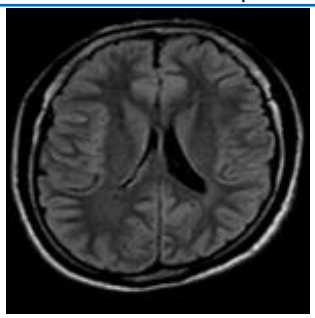


Fig 12: FLAIR axial

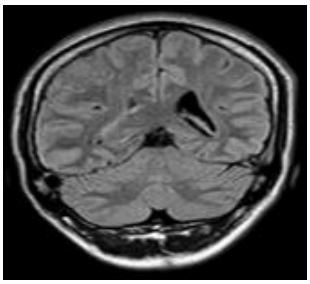


Fig 13: FLAIR coronal

FLAIR axial and coronal images showing mild left cerebral hemiatrophy with ex-vacuo dilatation of the ipsilateral lateral ventricle.

CASE 7: A 36-year-old male with a history of epilepsy since childhood presented with generalized tonic-clonic seizures. MRI demonstrated mild left cerebral and cerebellar hemiatrophy with ex-vacuo dilatation of the left lateral ventricle. EEG revealed bilateral hemispheric dysfunction with epileptiform discharges. The constellation of imaging and clinical findings favoured a diagnosis of Rasmussen encephalitis.

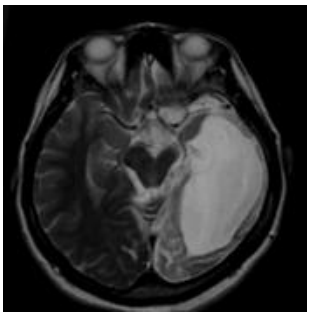


Fig 14:T2 axial

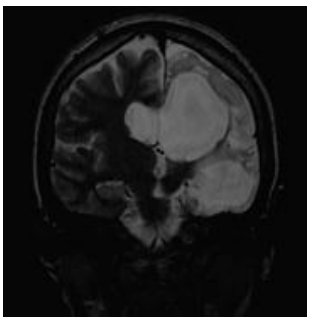


Fig 15:T2 coronal

T2 axial image shows severe atrophy of left cerebral hemisphere with dilatation of temporal horn of left lateral ventricle; atrophy of left cerebral peduncle is also noted. T2 coronal image demonstrates left cerebral hemiatrophy with ex-vacuo dilatation of the body and temporal horn of the left lateral ventricle.

CASE 8: A 4-year-old male child presented with recurrent seizures since infancy and developmental delay. Clinical examination revealed a right-sided facial port-wine stain involving the ophthalmic division of the trigeminal nerve. MRI demonstrated right parieto-occipital volume loss with prominence of cortical sulci and mild ex-vacuo dilatation of the adjacent right lateral ventricle. Multiple serpiginous cortical and extra-axial vascular structures were noted over the right frontoparietal convexity suggestive of leptomeningeal angiomatosis. These findings were consistent with Sturge-Weber syndrome.

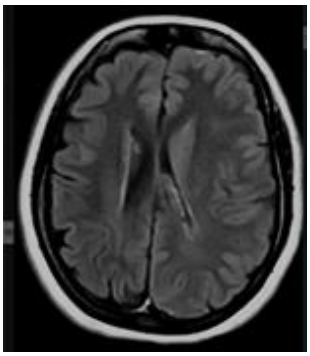


Fig 16: FLAIR axial

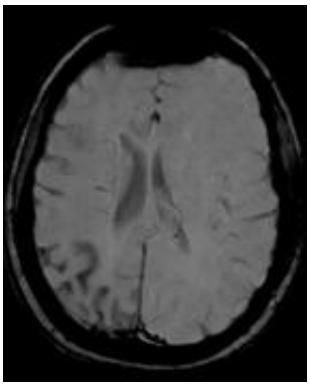


Fig 17: SWI axial

FLAIR axial image shows diffuse atrophy of right cerebral hemisphere. Susceptibility weighted axial image demonstrates foci of blooming along right occipital region suggesting calcification

CASE 9: An 8-year-old boy presented with recurrent focal seizures since infancy and mild developmental delay. Clinical examination revealed a right-sided facial port-wine stain. MRI demonstrated diffuse right cerebral hemiatrophy with prominence of cortical sulci and mild ex-vacuo dilatation of the ipsilateral lateral ventricle. Susceptibility-weighted imaging revealed focal blooming in the right occipital region representing cortical calcifications. A diagnosis of Sturge-Weber syndrome was established.

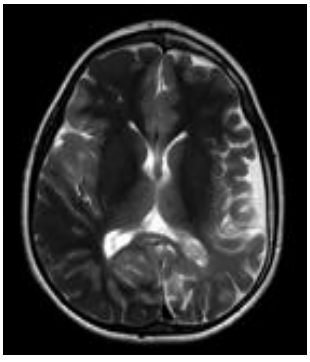


Fig 18: T2 axial

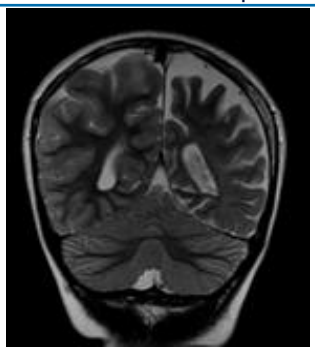


Fig 19: T2 coronal

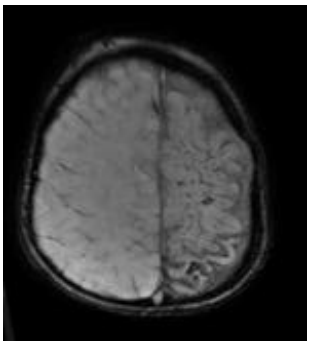


Fig 20: SWI axial

T2 axial and T2 coronal images show atrophy of left cerebellar hemisphere and also demonstrate enlargement of left choroid plexus. Prominent abnormal vascular channels are noted traversing the white matter of left cerebral hemisphere in axial susceptibility weighted image. It also shows subtle signal drop along left high parietal gyrus representing calcifications.

CASE 10: A 19-year-old male presented with recurrent seizures and mild left-sided weakness since childhood with a history suggestive of perinatal cerebral insult. MRI demonstrated marked right cerebral hemiatrophy with ex-vacuo dilatation of the right lateral ventricle and associated atrophy of the right cerebral peduncle. Compensatory osseous changes in the form of ipsilateral calvarial thickening and enlargement of the right frontal sinus were also noted. The imaging findings were characteristic of Dyke-Davidoff-Masson syndrome.

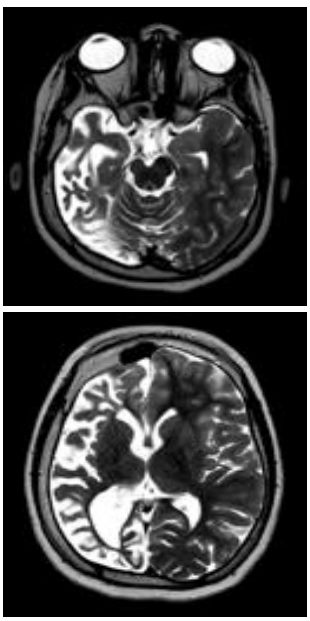


Fig 21 and 22: T2 axial images

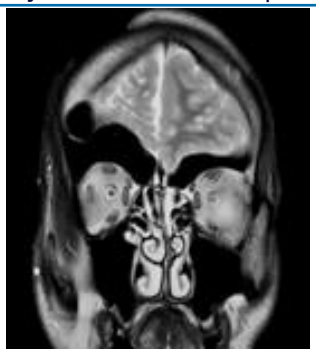


Fig 23:T2 coronal

T2 axial images demonstrate dilatation of right lateral ventricle secondary to atrophy of right cerebral hemisphere. Associated atrophy of right cerebral peduncle, calvarial thickening are also appreciated. T2 coronal image shows enlarged right frontal sinus.

DISCUSSION:

Dyke-Davidoff-Masson syndrome is a childhood hemiatrophy with compensatory ipsilateral calvarial hypertrophy.[10,11] DDMS occurs secondary to vascular, congenital or acquired ischaemic disease, traumatic or inflammatory insult occurring in utero or during the childhood period.[10] Calvarial changes occur only if brain damage occurs before 3 years of age.[11] Failure of brain growth results in compensatory changes like hyperpneumatized sinuses due to inward redirection of nearby calvarial growth. CT is preferred over conventional radiography in DDMS.[12]

Rasmussen encephalitis was discovered in 1958 by Theodore Rasmussen.[13,14] Rasmussen encephalitis is usually unicerebral and generally occurs in children under the age of 15 years with median age of 6 years.[13,14] It is a diagnosis of exclusion and typically insidious in onset.[13] It is characterized by unilateral hemispheric atrophy, abrupt appearance of focal, persistent motor seizures, followed by hemiplegia and progressive cognitive deterioration.[15] Atrophy of the head of the caudate, volume loss most accentuated in the insular and peri-insular regions, atrophy of hippocampus along with hemiatrophy as in our case have been seen in Rasmussen encephalitis.[13,14]

The prevalence of Sturge-Weber syndrome is 1 in 50,000 live births.[16] It is a sporadic neurocutaneous syndrome with predominant vascular manifestations. Contrast-enhanced MRI is the investigation of choice in SWS for depicting the presence and extent of leptomeningeal angiomatosis which is the hallmark of SWS. [17] Hemiatrophy in SWS is usually unilateral and confined to the parieto-occipital area, but can involve entire hemisphere or there can be bilateral involvement.[18,19] Gyriform hypointensities on T1- and T2-weighted MRI images seen as the areas of blooming on SWI occur due to venous collateralization and cortical calcification. These present as tramline calcifications on CT with underlying leptomeningeal angiomatosis.[19]

The present study highlights the broad spectrum of MRI manifestations of cerebral hemiatrophy and underscores the pivotal role of MRI in differentiating among various etiologies, thereby facilitating early diagnosis and appropriate management.

LIMITATIONS: The present study is limited by its retrospective design and relatively small sample size. Nevertheless, it highlights the diverse MRI manifestations of cerebral hemiatrophy and reinforces the diagnostic utility of MRI in etiological diagnosis.

CONCLUSION

Cerebral hemiatrophy encompasses a heterogeneous group of disorders with varied etiologies and clinical manifestations. MRI plays a pivotal role in identifying characteristic imaging features such as cerebral volume loss, ex-vacuo ventricular dilatation, calvarial changes, cerebral peduncular atrophy, and leptomeningeal vascular abnormalities. Familiarity with these imaging patterns enables accurate etiological diagnosis and facilitates appropriate patient management.

Ethical Approval: The study was conducted in accordance with institutional ethical standards. Patient confidentiality was maintained throughout the study.

Patient Consent: Informed consent was obtained from patients or their guardians for publication of clinical and imaging data.

Conflict of Interest: None declared.
Source of Funding: Nil.

REFERENCES:

1. McMonagle P, Deering F, Berliner Y, Kertesz A. The cognitive profile of posterior cortical atrophy Neurology. 2006;66:331–8
2. Parker CE, Harris N, Mavalwala J. Dyke-Davidoff-Masson syndrome: five case studies and deductions from dermatoglyphics. Clin Pediatr 1972;11:288-292.
3. Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging 1992;16:93-97.
4. Kandel D, Dhakal P, Bhandari S, Mahat S, Baniya J (2022) Rasmussen Encephalitis : A Case Report. J Med Case Rep Case Series 3(10) : https://doi.org/10.38207/JMCRCS/2022/OCT03100172 (01).
5. Rumboldt Z, Castillo M, Huang B, Rossi Andrea Brain Imaging with MRI and CT: An Image Pattern Approach. 2013 Cambridge University Press Cambridge Medicine:428
6. Zilkha A . CT of cerebral hemiatrophy. AJR 1980;135:259-262.
7. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy and homolateral hypertrophy of the skull and sinuses Surg Gynecol Obstet. 1933;57:588–600
8. Yerdelen D, Zafer F. Dyke–Davidoff–Masson syndrome Neurosurg Q. 2009;19:59–61
9. Rasmussen T, Olszewski J, Lloydsmith D. Focal seizures due to chronic localized encephalitis Neurology. 1958;8:435–45
10. Goyal J, Shah V, Rao S, Jindal N. Dyke Davidoff Masson syndrome in Children. The Internet Journal of Pediatrics and Neonatology 2008;10.
11. Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff-Mason-Syndrome. Neuroradiol 2006;16:165-6.
12. Radswiki R, Gaillard F. Cerebral hemiatrophy; 2011. Available from: http://radiopaedia.org/articles/cerebral-hemiatrophy. [Last accessed on 2020 04].
13. Akinkunmi M, Salisu M, Awosanya G. Rasmussen Encephalitis In A Nigerian Child: A Case Report. The Internet Journal of Radiology. 2009;12.
14. Bien CG, Granata T, Antozzi C, Cross JH, Dulac O, Kurthen M, et al. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: A European consensus statement. Brain 2005;128:454-71.
15. Zhang YH, Pu LH, Liu XY, Xiong H, Li YL, Liu XZ, et al. Clinical characteristics and treatment of Rasmussen syndrome in 16 children. Zhonghua Er Ke Za Zhi 2007;45:697-702.
16. Boyer RS. Disorders of histogenesis: Neurocutaneous syndromes. In: Osborn AG, editor. Diagnostic Neuroradiology. 1st ed. St Louis, Mo: Mosby; 1994. p. 72-113.
17. Zhou J, Li N, Zhou X, Wang J, Ma H, Zhang R. Sturge Weber Syndrome, A case report and review of literature. Chin Med J 2010;123:117-21.
18. Kelley TM, Hatfield LA, Lin DD, Comi AM. Quantitative analysis of cerebral cortical atrophy and correlation with clinical severity in unilateral Sturge-Weber syndrome. J Child Neurol 2005;20:867-70.
19. Marisa KB, Glenn AT. Pictorial essay: MRI of cerebral microhaemorrhages. AJR 2007;189:720-5.