Dyke-Davidoff-Masson Syndrome: Cerebral Hemiatrophy - Five cases with Review of literature

Dr.Chetana Ramesh Ratnaparkhi[1], Dr. Kajal Ramendranath Mitra[2], Dr. Prashant Madhukarrao Onkar[3]

ABSTRACT

Introduction-Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS) is uncommon entity in pediatric age group. It is characterized clinically by hemiparesis, seizures, mental retardation and has hallmark radiological picture which includes hemiatrophy of the cerebral hemisphere with hypertrophy of ipsilateral skull vault, hyerpneumatization of paranasal sinuses, mastoid air cells and elevation of petrous ridge. Most of the published data about this syndrome is of case reports. We report series of five pediatric cases from central India of cerebral hemiatrophy with classical clinical and radiological findings with short review of literature.

Case report- We have reported five cases of DDMS over a period of two years between July 2010 to August 2012 in pediatric age group who reported to us for imaging. Age of the patient varied between 5 months to 8 years which included one female child and four male children. Presenting feature were hemiplegia, mental retardation, seizures. On imaging cerebral atrophy contralateral to the side of hemiplegia is seen with calvarial thickening.

Conclusion- DDMS is characterized by triad of hemiparesis, contralateral cerebral hemiatrophy and seizures along with classical radiological features. Imaging has distinguished role in diagnosing this rare entity and differentiating it from other causes of cerebral hemiatrophy.

Key Words: cerebral hemiatrophy, Computed Tomography (CT), **Dyke**-Davidoff-Masson syndrome, Magnetic Resonance Imaging (MRI)

[1]Associate Professor, [2]Professor & Head, [3]Associate Professor

Department of Radio diagnosis, NKP Salve Institute of Medical sciences and Lata Mangeshkar Hospital, Digdoh Hills, Higna Road, Nagpur, Maharashtra, India.

Corresponding author mail: chetanaratnaparkhi@gmail.com

Conflict of interest—Nil

SEAJCRR

INTRODUCTION

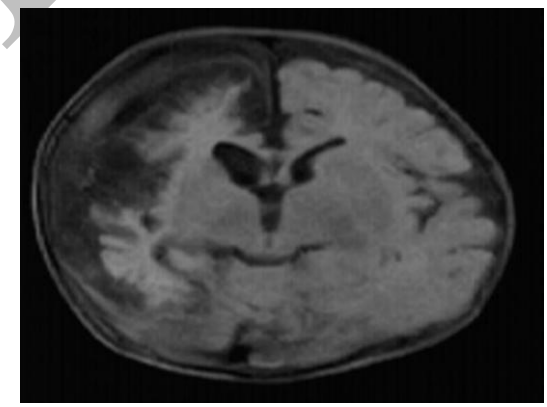
The radiological changes of Cerebral hemiatrophy were first described by Dyke, Davidoff and Masson in a series of nine patients on skull radiographs and pneumoencephalogram in 1933[1] This condition is suspected on clinical presentation of spastic hemiparesis, seizures, cerebral palsy & variable mental retardation. Radiological findings of contralateral hemiatrophy, calvarial and paranasal sinuses hypertrophy confirm the diagnosis [1-5].

Very few cases are reported in pediatric patients so far. Appropriate history, clinical examination along with radiologic findings offers a correct diagnosis. We encountered five cases of DDMS over a period of two years i.e. between July 2010 to August 2012 in pediatric age group who reported to us for imaging.

CASE REPORT SERIES

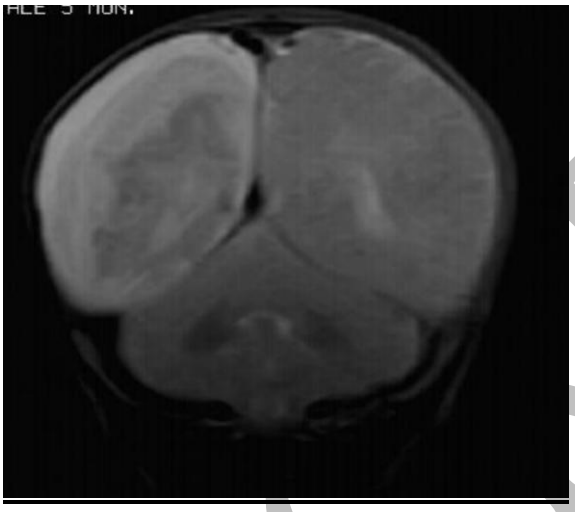
Case 1: A five months old male child presented with tonic clonic convulsions left sided hemiparesis and developmental delay. Birth history was unremarkable. Magnetic resonance imaging showed right cerebral hemiatrophy in the form of dilatation of the sulcal spaces, lateral ventricles and deepening of sylvian fissure in the right hemisphere (Figure1A & 1 B). The frontal sinuses were yet to be pneumatized. Widening of diploic space with chronic subdural collection was seen in right occipital and posterior parietal region.

Figure 1 A - MRI, Fluid attenuated inversion recovery sequence, axial image shows loss of volume on right side marked by dilatation of the sulcal spaces and dilatation of the ipsilateral lateral ventricles. Note hypointense subdural collection at parietal and frontal region along the convexity.



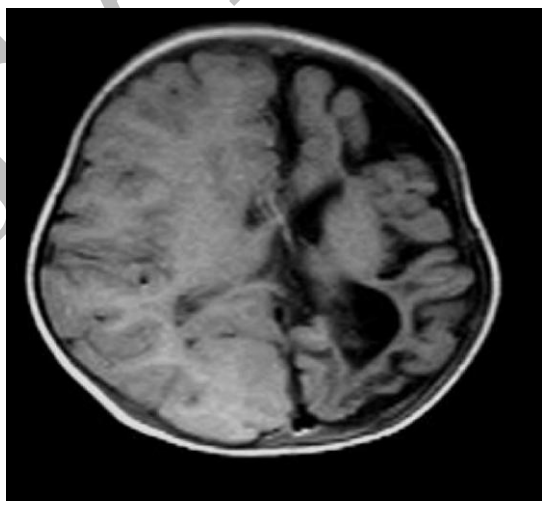
SEAJCRR

Figure 1B- Coronal T2 weighted image shows cerebral atrophy on right side with hyperintense subdual collection suggestive of chronic nature.



Case 2: One year old male child presented with right sided hemiparesis, difficulty in walking and cerebral palsy. On MRI brain, hemiatrophy of left cerebral hemisphere was seen (Figure 2). Calvarial thickening was not pronounced.

Figure 2- Axial T1 weighted image shows cerebral atrophy on left side.



SEAJCRR

Case 3: Eight year old male child presented with left sided hemiparesis and tonic clonic convulsions with mental retardation. On CT scan brain atrophy of right cerebral hemisphere was seen with thickening of the ipsilateral calvarium (Figure 3 A and 3B). Hyerpneumatization of ipsilateral mastoid air cells was also noted.

Figure 3A- CT brain axial image shows cerebral atrophy on right side with exvuo-dilatation of occipital horn of ipsilateral lateral ventricle.

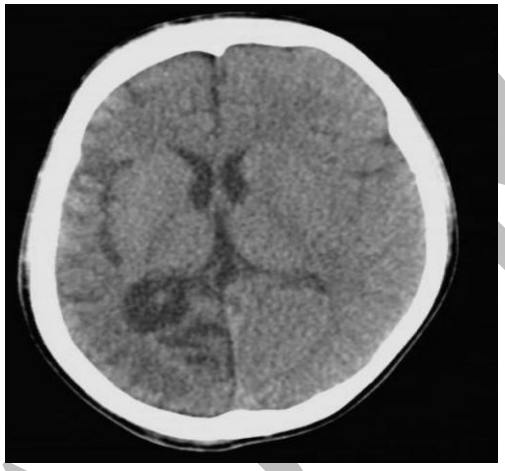
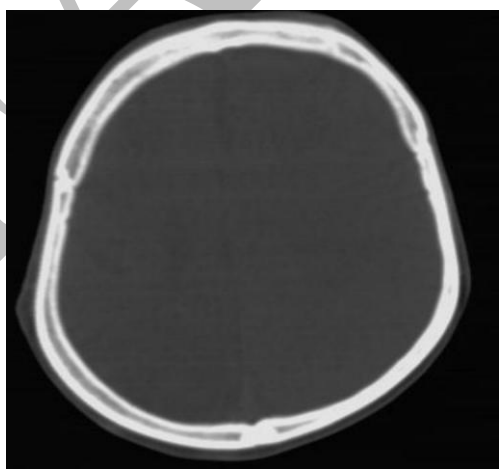


Figure 3B- CT brain bone window axial image shows calvarial thickening on right side on parietal region.



Case 4: Seven year old male child presented with left sided hemiparesis, seizure disorder of one year duration and developmental delay. MRI of the brain showed right cerebral hemiatrophy,

ipsilateral calvarial thickening and hyerpneumatization of ipsilateral mastoid air cells (Fig 4 A and Fig 4 B).

Figure 4 A & Figure 4 B - MRI Fluid attenuated inversion recovery sequence axial and coronal image respectively shows loss of volume on right side marked by dilatation of the sulcal spaces and dilatation of the ipsilateral lateral ventricles associated with calvarial thickening.

Figure 4 A

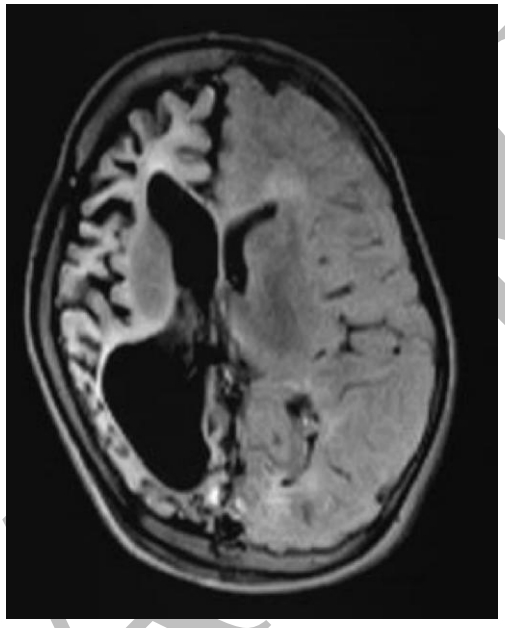
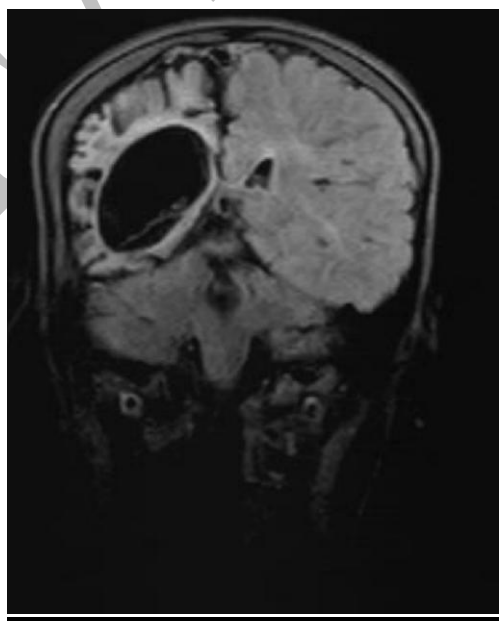


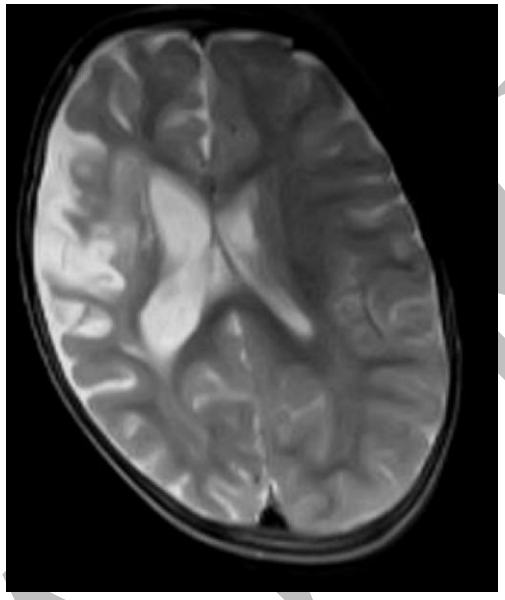
Figure 4 B



SEAJCRR

Case 5: Two year old female child presented with left sided hemiparesis and mental retardation. MRI of brain showed loss of cerebral volume on right side with chronic infarct in corona radiata, posterior limb of internal capsule and part of the insular cortex (Figure 5).

Figure 5 - Axial T2 weighted image shows cerebral atrophy on right side with dilatation of right lateral ventricle.



Shift of the midline structures and cerebellar atrophy was not seen in any of the patients. Intracranial calcification was not seen any of the patients.

On clinical and radiological findings, diagnosis of Dyke Davidoff Masson syndrome was made. All the cases were of secondary or acquired variety.

DISCUSSION

The presenting features of DDMS are hemiparesis (typically spastic hemiplegia), seizures, variable degree of mental retardation and facial asymmetry. Clinical features vary depending upon time and severity of brain damage. Most of the reported cases show male predilection and left cerebral hemisphere involvement [2]. On the contrary, in our case series, right cerebral hemisphere involvement (4:1) is predominant, however male predilection (4:1) is same as described in literature. Other clinical features mentioned are sensory involvement, speech impairment and crossed cerebellar atrophy. Psychiatric illnesses like schizophrenia have also been reported in a patient with DDMS [3]. There

was no history of psychiatric illness in our patients however in our case series all the patients were mentally challenged. No structural abnormality was seen in the cerebellum in our patients.

C.G Dyke, L.M Davidoff and C.B Masson have reported radiographic findings of this rare syndrome in series of nine patient ways back in 1933 [1]. They had done skull radiography and pneumoencephalography and reported a radiographical finding which includes contralateral cerebral atrophy in the form of widening of the sulci and dilatation of the ipsilateral lateral ventricle, thickening of adjacent calvarium, hypertrophy of frontal and ethmoid sinuses. Most of these findings were seen on imaging in our cases. Other findings are elevation of the greater wing of sphenoid and petrous ridge [1-5]. All these findings are better appreciated on computed tomography or magnetic resonance imaging than radiographs. Currently these are the modality of choice to confirm the diagnosis of DDMS.

Etiology of cerebral atrophy is divided into congenital or primary and acquired or secondary. In congenital or primary variety, intrauterine vascular occlusion is an underlying pathology. In acquired or secondary variety, ischemic brain insult occurs in perinatal life or later secondary to trauma, infections, congenital vascular malformations, hypoxia, intraparemchymal / intraventricular hemorrhages, coarctation of mid aortic arch and amniotic band [1, 5, 6, and 7]. The involvement of the middle cerebral artery territory is common. All the five cases in this series demonstrated the involvement of middle cerebral artery territory. Hageman et al, proposed the term cerebral hemi-hypoplasia or unilateral cerebral hypoplasia for primary or congenital cerebral atrophy as it is lack of cerebral development than true atrophy [8].

Hemiatrophy of the cerebral hemisphere is not very common in pediatric patient. The brain attains half of its adult size during the first year of life and three-fourths by third year. Growing brain is a major contributing factor for development of the skull vault. When brain growth is hampered, the adjacent structures viz. skull vault and sinuses grow inward, resulting in enlargement of the frontal sinus, diploic space and the elevation of the greater wing of sphenoid and the petrous ridge on the affected side [9]. These changes are evident only when the brain damage has occurred early in life i.e. before three years of age.

SEAJCRR

The characteristic radiologic findings in congenital or primary variety of hemiatrophy are shift of midline structures towards the side of the disease, sulcal prominence and gliotic changes [10]. These features differentiate it from acquired variety where midline shift and gliotic changes like porencephalic cyst (cystic encephalomalacia) are absent as seen in our cases.

Differential diagnoses of cerebral hemiatrophy are Sturge-Weber Syndrome, Linear-nevus syndrome, Basal ganglia Germinoma, Fishman syndrome, Silver-Russell syndrome, Rasmussen encephalitis and Parry –Romberg syndrome [4, 10].

Hemiparesis in Sturge –Weber syndrome is secondary to the unilateral cerebral involvement which is seen in 80% of the patients [11].The classical port wine stain, vascular angiomas and cortical calcification differentiates hemiatrophy in Sturge –Weber syndrome from DDMS.

Linear-nevus syndrome is marked by developmental delay, seizures, flat nevus on face associated with hemimegalencephaly, in few cases cortical atrophy with dilated ventricles has also been seen [12].

Cerebral hemiatrophy is seen in one third of the patients with basal ganglia Germinoma which shows minimally enhancing mass in the basal ganglia region which causes tumor infiltration of ganglion cells and nerve fibers resulting in degeneration of efferent and afferent fibers from the basal ganglia and thalamus [13].

Fishman syndrome is a neurocutaneous syndrome characterized by unilateral cutaneous lipomatous hamartomata involving scalp, eyelid, and outer globe of the eye, cerebral or cerebellar lipomas, and ipsilateral cortical atrophy with or without calcification with developmental delay [14].

Silver-Russell syndrome is genetic disorder characterized by disparity in the size of two halves of the body and growth retardation and underlying cause for asymmetry is a disturbance in the control of symmetry [15].

In Rasmussen encephalitis, progressive unilateral cerebral atrophy is seen mainly limited to frontal and temporal region, i.e., insular and periinsular cortex and atrophy of the basal ganglia associated with presence of oligoclonal or monoclonal banding on CSF examination [16].

Parry –Romberg syndrome (PRS) is marked by progressive hemifacial atrophy and on imaging shows increased signal intensity in the white matter, leptomeningeal

SEAJCRR

enhancement and variable degree of cerebral atrophy and cerebral calcifications [17].

Some reported cases mention postictal cerebral hemiatrophy in patients due to extensive neuronal loss involving entire hemisphere [18].

Conclusion`: - Diagnosis of DDMS is made by triad of hemiparesis, contralateral hemiatrophy and seizures along with classical radiological features. Imaging plays important role in diagnosing this rare entity and differentiating it from other causes of cerebral hemiatrophy.

REFERENCES:

[1] Dyke CG, Davidoff LM, Masson LB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinus. Surg Gynecol Obstet 1933; 57: 588-600.

[2] Unal O, Tombul T, Cirak B, Anlar O, and Incesu L, Kayan M: Left hemisphere and male sex dominance of cerebral hemiatrophy (Dyke-Davidoff-Masson Syndrome). Clin. Imaging. 2004; 28:163-65.

[3] Puri BK, Hall AD, Lewis SW. Cerebral hemiatrophy and schizophrenia, Br J Psychiatry 1994; 165:403-5.

[4] Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff Masson syndrome. Indian J Radiol Imaging 2006; 16:165-166.

[5] Tasdemir HA, Incesu L, Yazicioglu AK, Belet U, Gungor L. Dyke Davidoff Masson syndrome. Clin Imaging 2002; 26: 13-17.

[6] Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE,Jacoby CG: Cerebral hemiatrophy, hypoplasia of internal carotid artery and intracranial aneurysm. A rare association occurring in an infant. Arch Neurol 44:232-235, 1987.

[7] Stred SE, Byrum CJ, and Bove EL, Oliphant M: Coarctation of the midaortic arch presenting with monoparesis. Ann. Thorac Surg 42:210-212, 1986.

[8]Hageman G, Gooskens R H J M, Willemse J, A cerebral cause of arthrogryposis: Unilateral cerebral hypoplasia. Clin Neurol Neurosurg 1985; 87: 119-119-122.

[9]Parker C E, Harris N and Mavalwala J, Dyke-Davidoff-Masson Syndrome: Five case studies and deduction from dermatoglyphics. Clinical Pediatrics 1972; 11(5):288-292.

[10] Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging l992; 16:93-7.

[11]Gilbert Vezina and A. James Barkovich. The Phakomatoses.

SEAJCRR

In: A. James Barkovich, Charles Raybaud, editors. Pediatric Neuroimaging 5th edition, Lippincott Williams & Wilkins, a Wolters Kluwer busniness 2012, p. 605-609.

[12] Cheng-Chun Chen, Shao-Hung Lien, Mu-Ling Hsu, Chiung-Hsi Tien, Chun-Jung Chen,Pei-Yuan Chang et al. Linear Nevus Sebaceous Syndrome. J Med Sci 2006; 26(2):077-082.

[13] Tamaki N, Lin T, Shirataki K, Hosoda K, Kurata H, Matsumoto S, et al. Germ cell tumors of the thalamus and the basal ganglia. Childs Nerve Syst 1990; 6:3–7.

[14] Sergiusz Jozwiak (2012), MD, PhD; Chief Editor: Dirk M Elston. Haberland Syndrome. Retrieved from http://emedicine.medscape.com/article/1117060-overview.

[15] Tanner JM, Lejarraga H, Cameron N: The natural history of the Silver-Russell syndrome: a longitudinal study of thirty-nine cases. Pediatric Res 1975, 9:611-23.

[16] Gary Hedlund, James F.Bale, JR, A. James Barkovich. Infections of the Developing and Mature Nervous System. In: A. James Barkovich, Charles Raybaud, editors. Pediatric Neuroimaging 5th edition, Lippincott Williams & Wilkins, a Wolters Kluwer busniness 2012, p. 1022-1024.

[17] Fry JA, Alvarellos A, Fink CW, Blaw ME, Roach ES. Intracranial findings in progressive facial hemiatrophy. J Rheum 1992; 19:
956 –958

[18] Vosskämper, M., & Schachenmayr, W. (1990). Cerebral hemiatrophy: a clinicopathological report of two cases with a contribution to pathogenesis and differential diagnosis. Clin Neuropathol, 9(5), 244-50.

SEAJCRR