INTERNATIONAL JOURNAL OF CURRENT MEDICAL AND PHARMACEUTICAL RESEARCH

IJCMPR

ISSN: 2395-6429

Available Online at http://www.journalcmpr.com

DOI: http://dx.doi.org/10.24327/23956429.ijcmpr20170078

RESEARCH ARTICLE

DYKE-DAVIDOFF-MASSON SYNDROME: CT &MR IMAGING

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ARTICLE INFO

Article History:

Received 11th February, 2017
Received in revised form 14th March, 2017
Accepted 20th April, 2017
Published online 28th May, 2017

Key words:

Hemiatrophy, refractory seizures, magnetic resonance imaging (MRI)

ABSTRACT

Dyke-Davidoff-Masson syndrome also known as cerebral hemiatrophy is a rare entity characterized by unilateral cerebral atrophy secondary to in utero or early childhood cerebral insult such as infarct, trauma or infection. There is ipsilateral compensatory calvarial, diploic hypertrophy and contralateral hemiparesis. We present a case of this uncommon entity and discuss its imaging features, differential diagnosis, treatment options and prognosis.

INTRODUCTION

Dyke-Davidoff-Masson syndrome is rare and is characterized by cerebral hemiatrophy secondary to brain insult in utero or early childhood cerebral insult. There is ipsilateral compensatory osseous hypertrophy and contralateral hemiparesis (1). We present a case of this uncommon condition and discuss its imaging features, differential diagnosis, treatment options and prognosis.

CASE REPORT

A 12-year-old male child presented with complaints of focal seizures, characterized by brief episodic movements of right side of body after which child regains consciousness. This started approximately at 3 years of age. The frequency of convulsions increased gradually and more so in the last year. There was history of speech and language delay as well. His birth history and family history was unremarkable. On examination, patient had spastic hemiparesis on the right side. Treatment history revealed patient was consulting local medical practitioner and was on phenytoin. However, patient had stopped responding recently.

Initial non-enhanced CT scan (1mm slice thickness with multiplanar reconstructions) followed by contrast enhanced Magnetic resonance imaging (Siemens, Avanto 1.5T MRI) of brain showed hemiatrophy with gliotic changes of left cerebral hemisphere, ipsilateral dilated lateral ventricle and thickening of diploic space. The falx was inserted midline. Left frontal sinus was prominent (Figures 1,2). Features were suggestive of Dyke-Davidoff-Masson syndrome. Other antiepileptic drugs were added in his medication and he almost became seizure free. He was advised speech therapy too and was kept on regular follow-ups.

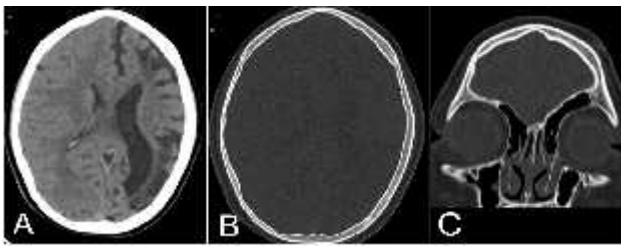

Figure 1 A. Left cerebral hemiatrophy with prominent sulci and ventricles, falx is midline.
B. Thickened Calvaria on left side.
C. Prominent left frontal sinus.

DISCUSSION

Dyke-Davidoff-Masson syndrome is defined by variable degrees of hypoplasia or atrophy of unilateral cerebral hemisphere with compensatory osseous changes. The cause could be either vascular insult during intrauterine life resulting in hypoplasia of a cerebral hemisphere or acquired causes like trauma, vascular abnormalities intracranial haemorrhage and

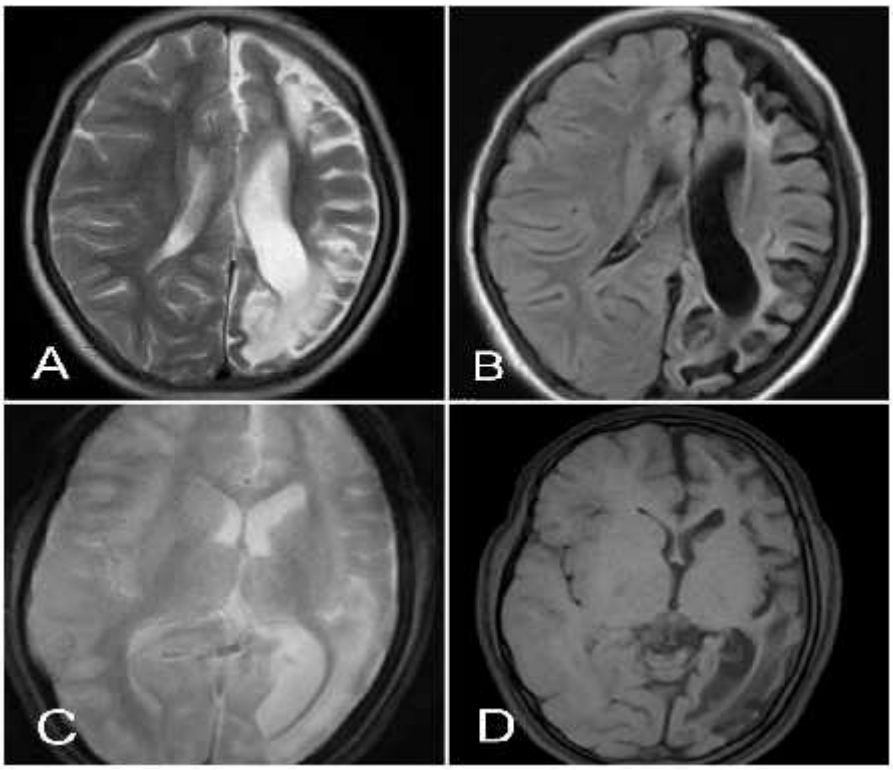

Figure 2 A.T2WI showing striking left cerebral hemiatrophy with enlarged left lateral ventricle, midline falx and interhemispheric fissure.
B. FLAIR sequence showing gliosis of left cerebral hemisphere. Thickening of calvaria noted on left side.
C. GRE sequence showing no calcification.
D. Post gadolinium T1FS showing no enhancement of pia.

uncommonly infection in the perinatal period or shortly thereafter, causing cerebral hemiatrophy (2, 3). The cause of cerebral atrophy is reduction in the formation of brain derived neurotropic factors (4,5).

The syndrome was first described by Dyke, Davidoff and Masson in 1933 on plain skull radiographs and pneumoencephalograms in a series of nine patients (2). The clinical manifestations depend upon extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation or learning disability, speech or language disorders and facial asymmetry. Uncommonly, it can also present with sensory symptoms and psychiatric disorders like schizophrenia. Seizures may present months to years after the onset of hemiparesis and mental retardation is not seen in all cases (5-7).

The diagnosis is based on the typical radiological features on computed tomography (CT) and MRI scans which include cerebral hemiatrophy with dilated ipsilateral lateral ventricle. Also there is thickening of calvarium with enlargement of frontal, ethmoid and sphenoid sinuses and elevation of greater wing of sphenoid and petrous ridge. The osseous abnormalities can also be depicted on plain skull films (5).

These compensatory cranial changes occur to take up the relative vacuum created by the atrophied cerebral hemisphere.[9,10] Shen *et al*.[11] depicted three MR imaging patterns of cerebral hemiatrophy: MR imaging pattern I corresponds to diffuse cortical and subcortical atrophy; pattern II corresponds to diffuse cortical atrophy coupled with porencephalic cysts; and pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory. In our case, pattern III was present.

The differential diagnoses are sturge weber syndrome, chronic Rasmussen encephalitis and large territorial MCA infarcts. However, sturge-weber syndrome additionally shows enhancing pial angiomas typical dystrophic cortical calcifications and enlarged choroid plexus. Rasmussen encephalitis doesn't show calvarial changes and demonstrates more focal encephalomalacia, typically around medial temporal lobe and around sylvian fissure (3,6). Large territorial MCA infarcts that occur after two or three years do not cause calvarial changes.

Management consists of control of seizures with appropriate anticonvulsants, as most patients with this disorder present with refractory seizures. Additionally, domiciliary physiotherapy, occupational and speech therapy have a crucial role. Hemispherectomy is indicated in patients with hemiplegia and intractable disabling seizures and is successful in 85% of the cases. Prognosis is poor in cases of prolonged or recurrent seizures and if hemiparesis occurs before two years of age. Hence, it is indeed very important for radiologists and clinicians to be familiar with this condition for its early diagnosis and treatment (5-8).

References

1. Park KI, Chung JM, Kim JY. Dyke-davidoff-masson syndrome: cases of two brothers and literature review. *J Epilepsy Res* 2014; 4:24-7.
2. Goyal J, Shah V, Rao S & N. Jindal: Dyke Davidoff Masson syndrome in Children. *The Internet Journal of Pediatrics and Neonatology.* 2009, 10:2.
3. Ozgur K. (2002). Dyke-Davidoff-Masson syndrome (Cerebral hemiatrophy). Available at: http://www.radiology.co.uk/srs-x/cases/092/d.htm. Accessed on: 12/04/2011.

4. Lee JH, Lee ZI, Kim HK, *et al*. A case of Dyke-Davidoff-Masson syndrome in Korea. *Korean J Pediatr* 2006; 49:208-11.
5. Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-davidoffmasson syndrome. *Indian J Radiol Imaging* 2006; 16:165-6.
6. Singh P, Saggar K, Ahluwalia A. Dyke-Davidoff-Masson syndrome: Classical imaging findings. *J Pediatr Neurosci* 2010; 5:124-5.
7. Behera MR, Patnaik S, Mohanty AK. Dyke-Davidoff-Masson syndrome. *J Neurosci Rural Pract* 2012; 3:411-3.
8. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson syndrome. *Indian Pediatr* 2008; 45:927-8.
9. Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: Clinicoradiological analysis of 19 cases. *Pediatr Int.* 2007; 49:70-5. [PubMed]
10. Kochar DK, Jain N, Sharma BV. Dyke-Davidoff-Masson syndrome-Neuroimage. *Neurol India*. 2001; 49:417-8. [PubMed]
11. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. *J Formos Med Assoc.* 1993; 92:995-1000. [PubMed]

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