

Case Report

DYKE DAVIDOFF MASSON SYNDROME: A CASE REPORT OF A RARE CLINICAL ENTITY

Harpreet Singh, Sameer Arora, *Ankit Kalra, Nikhil Govil, Saroj Dhankar and Vikram S Tanwar

Department of Medicine, Pandit B. D. Sharma PGIMS, Rohtak, Haryana, India

*Author for Correspondence

ABSTRACT

A 38 year old woman presented with recurrent seizures, spastic left sided hemiparesis since childhood with normal mental and speech capabilities. CT Imaging revealed right sided cerebral hemiatrophy with dilated ipsilateral ventricle, prominent sulcal spaces, osseous hypertrophy of same side with hyperpneumatization of frontal sinus diagnostic of acquired variety of Dyke Davidoff Masson Syndrome (DDMS).

Keywords: Hemiatrophy, Dyke Davidoff Masson Syndrome

INTRODUCTION

Dyke Davidoff Masson Syndrome (DDMS) is a syndrome characterised by constellation of multiple clinical and radiological features. Clinical features include seizures, mental retardation, facial asymmetry, contralateral hemiplegia and learning disabilities. Radiological features include cerebral hemiatrophy, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses (Sharma *et al.*, 2006). We report a case of DDMS in a 38 year old woman who presented with seizures and left sided weakness since childhood.

CASES

A 38 year old woman presented with history of multiple seizure episodes limited to left half of the body. Comprehensive clinical history revealed complains of weakness of left half of the body along with history of seizure disorder since childhood. Past history revealed episode of prolonged febrile seizures at the age of 1 year. There was no history of any medication use despite symptoms. Results of neurological examination showed normal mental status, vision, hearing and head circumference. There was evidence of left sided paresis, spastic in nature with brisk deep tendon reflexes and extensor plantar response on the left side. Additionally, musculoskeletal examination showed asymmetry in the two arms with left being less bulky.



Figure 1: CT Scan of the patient showing dilatation of right sided lateral ventricle with prominent sulci on same side

Case Report



Figure 2: CT scan of the patient showing osseous hypertrophy on the right side and hyperpneumatization of frontal sinus

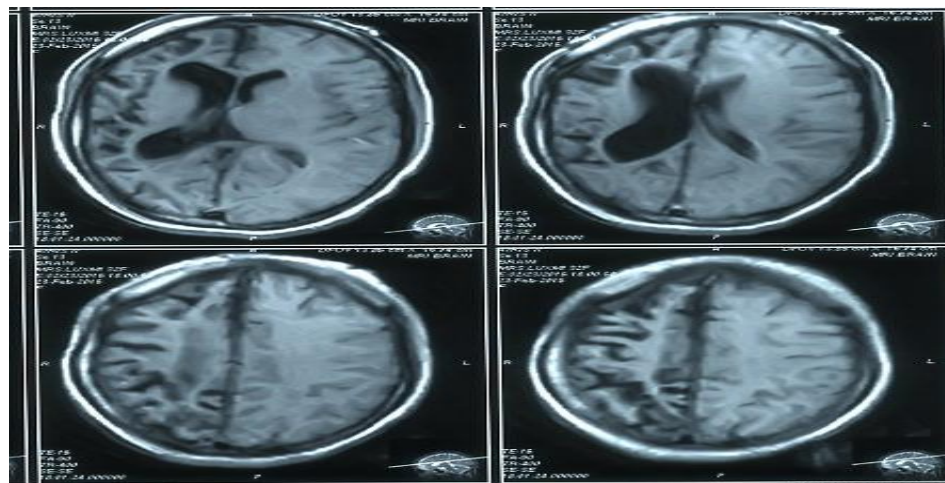


Figure 3: Axial T2 Weighted Image revealing sulcal prominence andencephalomalacia on right side

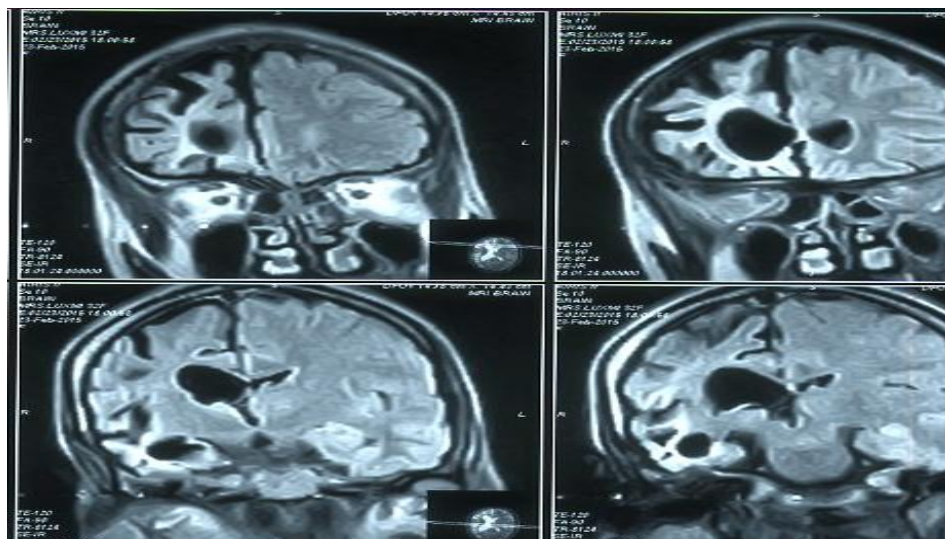


Figure 4: Coronal Flair MRI Image revealing the right sided atrophy of cerebrum with dilated right ventricle

Case Report

Plain CT scan of the head (Figure-1) revealed atrophy of the right cerebral hemisphere with exvacuo dilatation of the ipsilateral ventricle with right sided bony hyperostosis and hyperpneumatization of the frontal sinus (Figure-2). MRI Brain revealed areas of cystic encephalomalacia in right frontotemporo parietal cortical and subcortical regions involving right basal ganglia, perisylvian region and extending in periventricular regions with dilated adjacent sulci and right lateral ventricle (Figure-3 and 4). Based on clinical and radiological picture, a diagnosis of Dyke Davidoff Masson Syndrome (DDMS) was made.

DISCUSSION

In 1933, first description of DDMS was made by Dyke, Davidoff, and Masson on plain skull radiographic and pneumatoencephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial-asymmetry, and mental retardation (Dyke and Masson, 1933). DDMS is a rare entity encountered in clinical practice and is defined as hypoplasia of one cerebral hemisphere secondary to brain insult in fetal or childhood period (Sharma *et al.*, 2006). Male gender and left hemisphere involvement are more frequent though either sex or hemisphere may be affected (Unal *et al.*, 2004). The age of presentation with charactersitic changes are mainly seen in adolescence. Trauma, inflammation or vascular malformations and occlusions stand out to be the etiology behind DDMS (Parker *et al.*, 1972).

Compensatory cranial changes with thickening of calvaria and diploic space, enlargement and hyperaeration of paranasal sinuses and mastoid fill up the vacuum created by the atrophied hemisphere if insult occurs in first 2 years of life (Parker *et al.*, 1972; Solomon *et al.*, 1970)

There are two types of DDMS: congenital and acquired. Congenital variant is attributed to in-utero insult to the brain with shift of midline structures to diseased side with no sulcal prominence. Acquired variant resulting from cranial trauma, cerebrovascular accident, prolonged febrile seizures or inflammatory process shows prominent sulcal spaces on imaging as in present case. Triad of mental retardation, seizures and contralateral hemiplegia may be encountered in some though mental retardation is not always present and seizures may postdate the hemiparesis by months or years (Sener and Jinkins, 1992).

The differentials of DDMS include Sturge Weber syndrome, Basal cell germinoma, Linear-Nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis. Sturge Weber Syndrome (encephalotrigeminalangiomatosis) comprises of cerebral atrophy with leptomeningealangioma with characteristic features of port-wine facial nevus, intracranial tramtrack calcification, and the absence of midline shift (Jacoby *et al.*, 1977). Basal ganglia germinoma is a rare tumor of the brain, which may present with progressive hemiparesis and cerebral hemiatrophy (Liu *et al.*, 1999). Linear nevus syndrome is characterized by typical facial nevus, mental retardation, recurrent seizures, and unilateral ventricular dilatation resembling cerebral hemiatrophy (Jacoby *et al.*, 1977). Fishman syndrome or encephalocraniocutaneuslipomatosis is a rare neurocutaneous syndrome comprising unilateral cranial lipoma, lipodermoid of eye. The patient may present with seizure and cerebral imaging may show calcified cortex and hemiatrophy (Amar *et al.*, 2000). Characteristic features of Silver-Russel Syndrome are poor growth, delayed bone age, clinodactyly, normal head circumference, normal intelligence, classical facial phenotype (triangular face, broad forehead, small pointed chin, and thin wide mouth), and hemihypertrophy (Qiu and Shi, 2007). Rasmussen encephalitis lacks calvarial changes typical of DDMS and has more focal encephalomalacia mainly in the medial temporal lobe and around sylvianfissure (Sheybanl *et al.*, 2011).

Key to right diagnosis include detailed clinical history, examination and imaging modality like CT or MRI. MRI has the ability to differentiate between congenital and acquired types of DDMS by enlightening changes in the cerebral hemispheres as well as highlighting bony structures. MRI demonstrates encephalomalacia with shrunken hyperintensegyri on T2W images. DDMS neither enhances on T1W nor shows restricted diffusion but prominent sulci and cisterns with hemispheric volume loss are seen on T1W images.

Symptomatic treatment is the mainstay of therapy targeting convulsion, hemiplegia, hemiparesis and learning difficulties. Patients with refractory seizures may require multiple anticonvulsants.

Case Report

Hemispherectomy yields good success rate of about 85% in selected cases with candidates being children with intractable disability (Narain *et al.*, 2008). Prognosis is favourable if the onset of hemiparesis is after 2 years of age and in absence of prolonged or recurrent seizure (Pendse *et al.*, 2004).

REFERENCES

- Amar DJ, Kornberg AJ and Smith LJ (2000).** Encephalocraniocutaneouslipomatosis (Fishman Syndrome): A rare neurocutaneous syndrome. *Journal of Paediatrics and Child Health* **36** 603–5.
- Dyke CG DL and Masson CB (1933).** Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. *Surgery, Gynecology, and Obstetrics* **57**:88-600
- Jacoby CG, Go RT and Hahn FJ (1977).** Computed tomography in cerebral hemiatrophy. *American Journal of Roentgenology* **129** 5–7.
- Liu E, Robertson RL, Plessis A and Pomeroy SL (1999).** Basal ganglia germinoma with cerebral hemiatrophy. *Pediatric Neurology* **20** 312–4.
- Narain NP, Kumar R and Narain B (2008).** Dyke-Davidoff-Masson Syndrome. *Indian Pediatrics* **45** 927–8.
- Parker CE, Harris N and Mavalwala J (1972).** Dyke-Davidoff-Masson Syndrome: Five case studies and deductions from dermatoglyphics. *Clinical Pediatric Endocrinology* **11** 288–92.
- Pendse NA, Bapna P, Menghani V and Diwan A (2004).** Dyke-Davidoff-Masson Syndrome. *Indian Journal of Pediatrics* **71** 943.
- Qiu BP and Shi CH (2007).** Silver-Russel Syndrome: A case report. *World Journal of Pediatrics* **3** 68–70.
- Sener RN and Jinkins JR (1992).** MR of craniocerebralhemiatrophy. *Clinical Imaging* **16** 93–7.
- Sharma S, Goyal L, Negi A, Sood RG, Jhobta A and Surya M (2006).** Dyke-Davidoff-Masson Syndrome. *Indian Journal of Radiology and Imaging* **16** 165–6.
- Sheybanl L, Schaller K and Seeek M (2011).** Rasmussen encephalitis: An update. *Schweizer Archiv für Neurologie und Psychiatrie* **162** 225–31.
- Solomon GE, Hilal SK, Gold AP and Carter S (1970).** Natural history of acute hemiplegia of childhood. *Brain* **93** 107–20.
- Unal O, Tombul T, Cirak B, Anlar O, Incesu L and Kayan M (2004).** Left hemisphere and male sex dominance of cerebral hemiatrophy(DDMS). *Clinical Imaging* **28** 163–5.