



Dyke-Davidoff-Masson Syndrome: A case report

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Abstract:

Dyke-Davidoff-Masson Syndrome (DDMS) is a rare condition characterized by seizures, facial asymmetry, contralateral hemiplegia, learning disabilities and mental retardation which is usually due to an insult to the developing brain in fetal or post natal life. The characteristic radiologic features are cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. In this article, we report a case of 45 yr old man who presented with seizures, cognitive impairment and right sided hemiparesis since birth and with classic imaging features on cross sectional CT and MRI study which was subsequently diagnosed as DDMS.

Key words: Cerebral hemiatrophy, hemiparesis, hyperpneumatisation, seizures, sinuses

Introduction:

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to an insult to the developing brain in fetal or early childhood period [1-3]. DDMS is a rare condition characterized by seizures, facial asymmetry, contralateral hemiplegia, learning disabilities and mental retardation [1-3]. The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults [3-6]. However, it can also be seen in children [3,7]. Here, we report a case of 45 yr old man who presented with seizures, cognitive

impairment and right sided hemiparesis since birth and with classic imaging features on cross sectional CT and MRI studies.

Case Report

A 47 year old male with history of weakness of right half of body and seizures, on prophylactic antiepileptic drugs since birth presented with convulsions to the emergency department due to irregular medication in the recent past. Patient was managed with anticonvulsants and stabilised. Detailed clinical history was taken and general and systemic examination was done. General examination was normal with no signs of neurocutaneous markers seen. Neurologic

examination revealed right sided hemiparesis with features of upper motor neuron signs like spasticity, extensor plantar response and brisk tendon reflexes while rest of the other systemic examinations being normal. MR imaging was done which revealed atrophy and gliosis involving left cerebral hemisphere with encephalomalacic changes including dilatation of ipsilateral lateral ventricle, widening of sulci and ipsilateral sylvian fissure (figures 1 and 2). Also noted were hyperpneumatisation of frontal sinus and thickening of calvarium on the same side (figure 3). CT was subsequently done out of academic interest to better demonstrate the calvarial and sinus changes which correlated with findings seen on MRI. With such classic findings diagnosis of Dyke-Davidoff-Masson syndrome was made. Patient responded well with the treatment and was discharged following improvement and advised to be on regular medications for seizures with regular follow up.

Discussion

Dyke-Davidoff-Masson syndrome was initially described by C.G.Dyke, L.M.Davidoff and C.B.Masson in 1933 in plain skull x-ray and pneumatoencephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial-asymmetry and mental retardation [1-6]. But now in the modern era of advanced imaging techniques especially with CT and MRI, characteristic classical findings associated with the syndrome can very well be demonstrated.

DDMS is a rare clinical condition which refers to atrophy or hypoplasia of one cerebral hemisphere i.e. cerebral hemiatrophy. Both sexes can be affected while slight male predominance is seen. Age of presentation depends on time of neurologic insult. The clinical findings may be of variable degree and depends on the extent of the brain injury. The characteristic features include variable degrees of facial asymmetry, seizures, contralateral hemiparesis, mental retardation, speech and language disorders and learning disabilities [3].

Classic imaging findings seen on cross sectional imaging includes cerebral hemiatrophy with compensatory cranial changes which occur to take up the relative vacuum created by the atrophied cerebral hemisphere. Left cerebral hemisphere involvement is frequent [3,8]. Homolateral hypertrophy of the skull and sinuses i.e. thickening of calvarium and hyperpneumatisation of ipsilateral frontal sinus and mastoid air cells, elevation of the greater wing of

sphenoid and petrous ridge and upward tilting of planum- sphenoidale will be present. Other features are enlargement of ipsilateral sulci, dilatation of ipsilateral ventricle and cisternal spaces and decrease in size of ipsilateral cranial fossa suggesting cerebral volume loss.

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 [9,10].

Correct diagnosis could be achieved with proper clinical history, detailed systemic examination and imaging findings. The prime differential diagnoses to be considered includes Sturge weber syndrome, Epidermal nevus syndrome, Haberland syndrome, Rasmussen encephalitis and Basal ganglia germinoma [7,11].

Sturge weber syndrome (encephalotrigeminal angiomas) is a phakomatosis with leptomeningeal angiomas and ipsilateral facial nevus in the area of trigeminal nerve distribution. Imaging findings include cerebral atrophy with tram-track calcifications, choroid plexus enlargement, cranial diploe prominence and venous abnormalities [12].

Rasmussen encephalitis is a rare childhood syndrome characterized by medically refractory focal seizures and progressive multifocal neurologic symptoms such as hemiplegia and cognitive impairment. It is chronic, progressive encephalitis due to an autoimmune mechanism elicited secondary to viral infection resulting in progressive cortical atrophy. The most commonly encountered imaging feature is progressive unilateral cerebral cortical atrophy.

Haberland syndrome (encephalocraniocutaneous lipomatosis) is a rare neurocutaneous disease characterised by unilateral lipomatous hamartomata of the scalp, ocular and facial lesions with ipsilateral cerebral malformations such as ipsilateral cortical atrophy, porencephalic cysts, cranial asymmetry etc [13].

Epidermal nevus syndrome is a neurocutaneous syndrome associated with epidermal nevus and variety of congenital CNS disorders. Unilateral hemimegalencephaly is the most commonly found CNS abnormality. Other neuronal migration abnormalities like microgyria, heterotopia are also seen [14].

Basal ganglia germinoma is a rare tumor of the brain, which may present with progressive hemiparesis and cerebral hemiatrophy [15].

Refractory seizures is the usual presentation in patients with DDMS and thus treatment should be directed towards control of seizures with suitable antiepileptic drugs. Along with drugs, physiotherapy, occupational therapy, and speech therapy play a significant role in long-term management. Hemispherectomy is the treatment of choice in intractable disabling seizures and hemiplegia with a success rate of 85% in selected cases [3,7].

Thus, appropriate diagnosis of this condition by combined clinical examination and radiological investigation with prompt supportive treatment with antiepileptic drugs and physiotherapy can help improve the patient.

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References

1. Dyke C G, Davidoff L M and Masson C B. Cerebral Hemiatrophy and homolateral hypertrophy of the skull and sinuses. *Surg Gynecol Obstet* 1933; 57: 588-600
2. Sharma S, Goyal I, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff-Masson Syndrome. *Ind J Radiol Imag*. 2006;16:165-6.
3. Manas R Behera, Sibabrata Patnaik, Aswini K Mohanty. Dyke-Davidoff-Masson Syndrome. *J Neurosci Rural Pract*. 2012 Sep-Dec; 3(3): 411-3.
4. Singh P, Sagar K, Ahluwalia A. Dyke-Davidoff-Masson Syndrome: Classical imaging findings. *J Pediatr Neurosci*. 2010;5:124-5.
5. Koshy B, Surendrababu NR. Dyke-Davidoff-Masson Syndrome. *Ann Acad Med Singapore*.2010;39:501-2.
6. Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson Syndrome. *Neurol india*. 2003;51:136.
7. Narain NP, Kumar R, Narain B. Dyke-Davidoff-Masson Syndrome. *Indian Pediatr*. 2008;45:927-8.
8. Unal O, Tombul T, Cirak B, Anlar O, Incesu L, Kayan M. Left hemisphere and male sex dominance of cerebral hemiatrophy(DDMS). *Clin Imaging*. 2004;28:163-5.
9. Shen WC, Chen CC, Lee SK, Ho YJ, Lee KR. Magnetic resonance imaging of cerebral hemiatrophy. *J Formos Med Assoc*. 1993;92:995-1000.
10. Sener RN, Jinkins JR. MR of craniocerebral atrophy. *Clin Imaging* 1992; 16; 93-7.
11. Rao KCVG. Degenerative diseases and hydrocephalus. In: Lee SH, Rao KCVG, Zimmerman RA, editors. *Cranial MRI and CT*. Fourth edition. New York: McGraw-Hill; 1999. p.212-4.
12. Bonmati LM, Menor F, Poyatos C, Cortina H. Diagnosis of Sturge Weber Syndrome: Comparison of efficacy of CT and MR imaging in 14 cases. *AJR* 1992;158:867-71.
13. Rubegni P, Risulo M, Sbrano P, Buonocore G, Perrone S. Encephalocraniocutaneous lipomatosis (Haberland syndrome) with bilateral cutaneous and visceral involvement. *Clin Exp Dermatol* 2003;28:387-90.
14. Zhang W, Simos PG, Ishibashi H, Wheless JW, Castillo EM et al. Neuroimaging features of epidermal nevus syndrome. *AJNR* 2003;24:1468-70.
15. Liu E, Robertson RL, Plessis A, Pomeroy SL. Basal ganglia germinoma with cerebral hemiatrophy. *Pediatric Neurology*. 1999;20:312-4.

Figures:

Figure 1: T2 weighted Coronal MR image showing hemiatrophy of left cerebral hemisphere with osseous thickening of cranial bones on ipsilateral side (solid arrow). Dilatation of left lateral ventricle is also noted (open arrow).

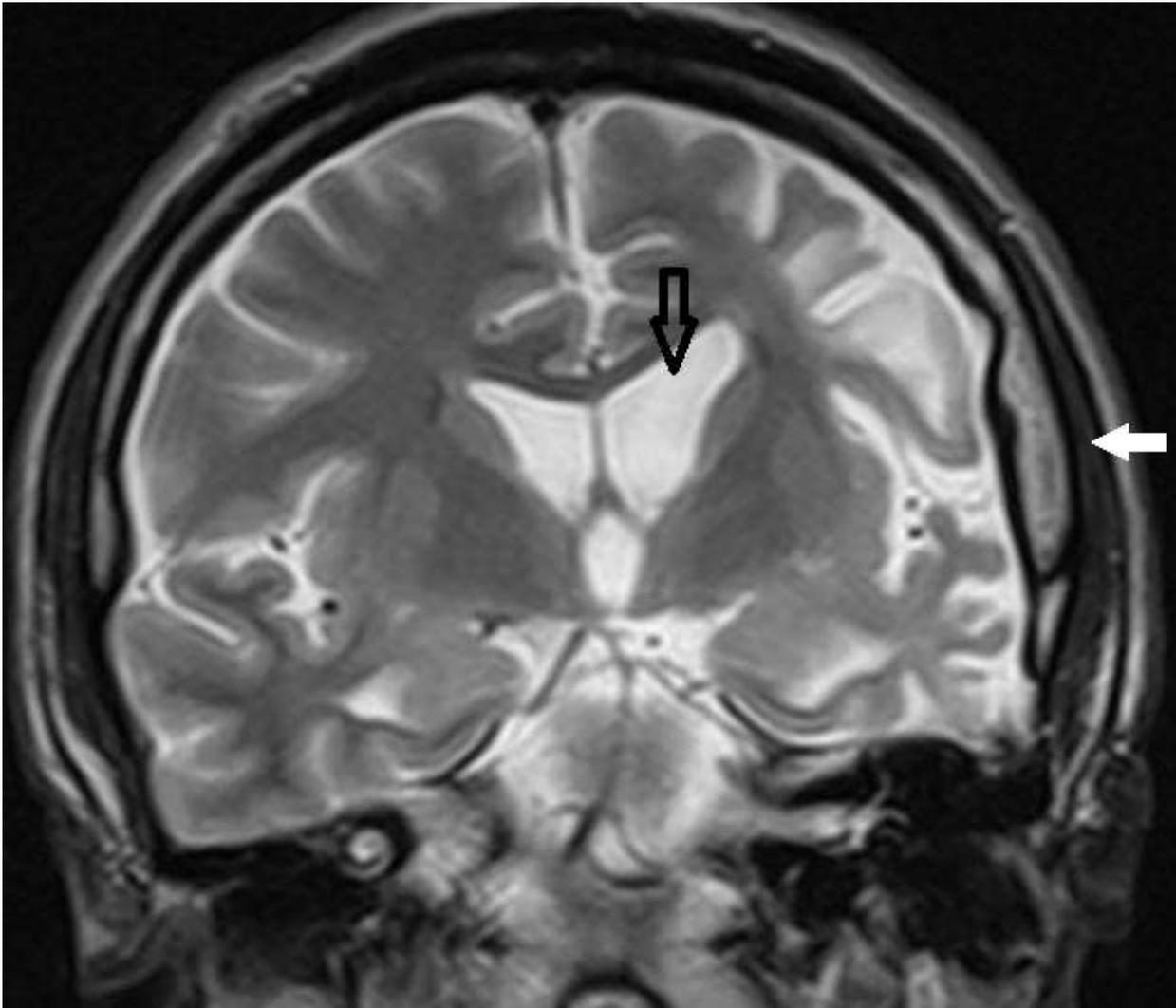


Figure 2: FLAIR axial MR image showing hemiatrophy of left cerebral hemisphere, gliotic changes (short arrow), prominent left sylvian fissure and dilated temporal horn of left lateral ventricle (star).

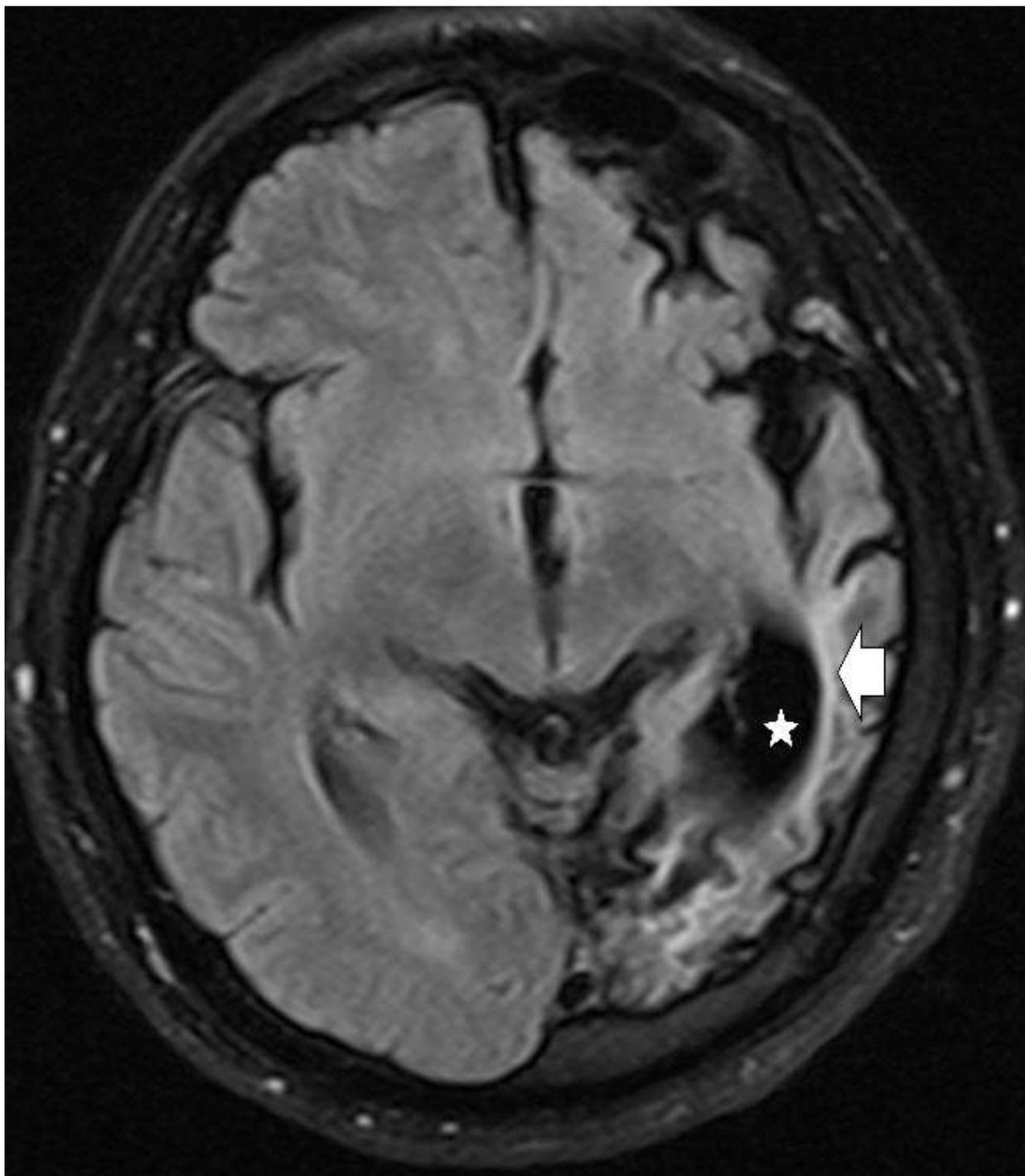


Figure 3: NCCT axial image showing hyperpneumatized left frontal sinus (asterisk) with ipsilateral calvarial thickening.

