Dyke-Davidoff-Masson Syndrome

NIGAM PRAKASH NARAIN Rakesh Kumar Bhupendra Narain

ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is characterized by seizures, facial asymmetry, contralateral hemiplegia and mental retardation. The characteristic radiologic features are cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. We report a case of DDMS in an 18-monthold girl who presented with right sided focal seizures, hemiparesis of the same side, and delayed milestones.

Key words: Hemiatrophy, seizures.

First description of Dyke-Davidoff-Masson syndrome (DDMS) dates back to 1933, when Dyke, Davidoff and Masson described the plain skull radiographic and pneumatoencephalographic changes in a series of nine patients(1). Since then only few pediatric cases of DDMS have been reported.

CASE REPORT

An 18-month-old female baby, born full term, presented with recurrent tonic seizures of right half of body since the age of 6 month. There was no history of significant antenatal or perinatal

From the Department of Pediatrics, Patna Medical College, Patna, Bihar, India.

Correspondence to: Dr. Nigam Prakash Narain, Associate Professor, Department of Pediatrics, Patna Medical College, Patna, Bihar, India. E-mail: nigampn@gmail.com

Manuscript received: April 30, 2007; Initial review completed: June 13, 2007; Revision accepted: February 27, 2008. complication. Neurological examination revealed right-sided spastic hemiparesis, brisk deep tendon reflexes, and extensor plantar response. The baby could not stand without support and spoke bisyllables only. There was no neurocutaneous marker or asymmetry of face or body and head circumference was normal. Vision and hearing were normal and cranial nerves were intact. Hematological profile and cerebrospinal fluid examination were normal. Computed Tomography (CT) scan of head revealed hemiatrophy of left cerebral hemisphere, dilatation of the left lateral ventricle, widening of ipsilateral sulci, a well defined cystic lesion and calvarial expansion on the left side. There was shift of the midline to the left. We made a diagnosis of Dyke-Davidoff-Masson syndrome.

DISCUSSION

DDMS is a rare condition characterized clinically by variable degrees of facial asymmetry, seizures, contralateral hemiparesis, mental retardation and learning disabilities in association with the classical radiological findings of asymmetry of cerebral hemispheric growth with atrophy on one side, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses(1-4).

Both sexes and any of the hemispheres may be affected but male gender and left hemisphere involvement are more frequent. Age of presentation depends on time of neurologic insult and characteristic changes may be seen only in adolescence. The clinical findings may be of variable degree depending on the extent of the brain injury. Varying degrees of atrophy of one half of body, sensory loss, speech and language disorder, mental retardation or learning disability and psychiatric manifestations like schizophrenia may also be present. In the present case, the findings of dilated cortical sulci and widening of ipsilateral diploi reflect a late onset of brain insult probably of vascular origin involving left middle cerebral artery.

A proper history, thorough clinical examination and radiologic findings provide the correct diagnosis.

INDIAN PEDIATRICS

CASE REPORTS

The condition needs to be differentiated from Basal ganglia germinoma, Sturge Weber syndrome, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis(5,6).

A possible etiological relation between cerebral atrophy and seizures has been reported in two different studies from India(7,8). Prognosis is better if hemiparesis occurs after the age of 2 yrs and in absence of prolonged or recurrent seizures. Children with intractable disabling seizures and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases.

Contributors: NPN diagnosed this case, supervised the management and drafted the manuscript; he will act as guarantor of the paper. BN and RK were involved in patient management and review of literature.

Funding: None.

Competing interests: None stated.

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.

Multiple Foreign Bodies in a Neonate

ANURAG MEDATWAL PPGupta RK Gulati

ABSTRACT

We report a rare instance of nine foreign bodies in a neonate that included a coin, safety pin, screw, cotton piece, polythene piece, and four glass pieces. Of these, six foreign bodies were removed by esophagoscopy and endoscopy, two glass pieces were passed in feces and one could not be removed. The child died 5 days after admission.

Keywords: Esophagus, Foreign body, Gastrointestinal tract, Newborn.

- Tasdemir HA, Incesu L, Yazicioglu AK, Belet U, Gungor L. Dyke Davidoff Masson syndrome. Clin Imaging 2002; 26: 13-17.
- Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff Masson syndrome. Indian J Radiol Imaging 2006;16:165-166.
- 4. Dix JE, Coil WS. Cerebral hemiatrophy: Classification on the basis of MR Imaging findings of mesial temporal sclerosis and childhood febrile seizures. Radiology 1997; 203: 269-274.
- 5. Sener RN, Jinkins JR. MR of craniocerebral atrophy. Clin Imaging 1992; 16; 93-97.
- 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. 212-214.
- Nair KP, Jayakumar PN, Taly AB, Arunodaya GR, Swamy HS, Shanmugam V. CT in simple partial seizures in children: a clinical and computed tomography study. Acta Neurol Scand 1997; 95: 197-200.
- 8. Garg RK, Karak B. Cerebral hemiatrophy: a possible etiological relation with febrile seizures. Indian Pediatr 1998; 35: 79-81.

A 12 day-old female child having normal parents, presented with a complaint of recurrent vomiting, fever and respiratory distress for 11 days. During this period of 11 days she was treated by many pediatricians, with antibiotics and other supporting medicines. On admission, the general condition was poor and she was put on antibiotics and CPAP. Chest *X*-ray revealed three metallic foreign bodies (a coin, safety pin, and screw) in the upper, mid-esophagus

From the Department of Pediatrics, Jay-Kay Mother and Child Hospital, Medical College, Kota, Rajasthan, India.

Correspondence to: Anurag Medatwal, Department of Pediatrics, Jay-Kay Mother and Child Hospital, Medical College, Kota, Rajasthan, India. E-mail: amedatwal@yahoo.co.in

Manuscript received: August 8, 2006; Initial review completed: December 28, 2006; Revision accepted: February 27, 2008.

INDIAN PEDIATRICS

VOLUME 45-NOVEMBER 17, 2008