DYKE-DAVIDOFF-MASSON SYNDROME

Naba Raj Koirala, Roshna Khadka, Manoj Bhattarai, Dibya Tulachan, Ajay Kumar Das, and Jwalanta Poudel

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. Here we report a case of a 16 years young girl who presented with seizures severe mental retardation and weakness of left upper-limb and on CT brain was diagnosed to have DDMS.

Key words: DDMS, Seizure, mental retardation, Hemiatrophy, hypoplasia, cerebral hemisphere

Introduction:

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.

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. Hypoplasia of one cerebral hemisphere (hemiatrophy) is mostly secondary to brain insult in fetal or early childhood period. Hemiatrophy is not frequently encountered in clinical practice. We present here a case of a 16 years young girl who presented with seizures and weakness of left upper-limb and on CT was diagnosed to have DDMS.

Case report:

A 16 years young girl born full term, presented to the psychiatry OPD of Nobel Medical College Teaching Hospital with complaints of seizures for near about past 15 years and weakness in left upper limb since birth. Her seizures were controlled with tablet Phenytion sodium 300 mg/day but now for last 4 months she had seizures even when on regular medication. Neurological examination revealed right-sided spastic hemiparesis, brisk deep tendon reflexes, and extensor plantar response. The baby could not stand without support and speak only monosyllabic words. There was no neurocutaneous marker or asymmetry of face or body. Her head circumference, and visual and hearing capacities were normal, so her cranial nerves. Her hematological profile and cerebrospinal fluid examination were also normal. Her 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:

In 1933, Dyke, Davidoff, and Masson described the plain skull radiographic and

pneumatoencephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial-asymmetry, and mental retardation[1]. The plain skull radiographic changes included thickening of calvarium and dilatation of ipsilateral frontal and ethmoid sinuses. Also, there was elevation of the greater wing of sphenoid and petrous ridge elevation of the greater wing of sphenoid and petrous ridge and upward tilting of planum- sphenoidale.

In 1939, Alpers and Dear defined two types of cerebral hemiatrophy[2]. In the primary (congenital) type, the entire cerebral hemisphere is characteristically hypoplastic. The secondary type results from a cerebrovascular lesion, inflammatory process, or cranial trauma. Twenty-two cases of primary variety were collected from the literature until 1939. A clinical triad of hemiplegia, seizures and mental retardation was defined. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis[3].

According to Hageman et al, since primary cerebral atrophy is actually a lack of cerebral development rather than atrophy the term cerebral hemi-hypoplasia or unilateral cerebral hypoplasia would be more appropriate[4]. The brain reaches half of its adult size during the first year of life and reaches three-fourths of that size by the end of third year. As it enlarges, the brain presses outward on the bony tables and is partly responsible for the gradual enlargement and general shape of the adult head. When the brain fails to grow properly, the other structures tend to direct their growth inward, thus accounting for the enlargement of the frontal sinus, the increased width of the diploic space and the elevations of the greater wing of sphenoid and the petrous ridge on the affected side[5]. These changes can occur only when brain damage is sustained before three years of age however, such changes may become evident as soon as nine-months after brain damage was sustained[6].

A vascular cause of cerebral hemiatrophy (hypoplasia), first proposed in 1860 was confirmed in later studies[7,8,9]. It was proposed that a vascular anomaly occurring in very early gestation (five or six weeks) may result in a major defect in brain development whereas those occurring later may produce more localized lesions[8]. It was reported that decrease in carotid artery flow due to coarctation of aorta can also cause cerebral hemiatrophy. Crossed cerebellar atrophy is usually associated with long standing, extensive and unilateral cerebral lesions with onset during infancy or early childhood. The manifestations of DDMS may be so subtle as to be overlooked on plain radiographs; however, CT is the diagnostic modality of choice[8,9].

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.

A proper history, thorough clinical examination and radiologic findings provide the correct diagnosis. The condition needs to be differentiated from Basal ganglia

germinoma, Sturge Weber syndrome, Linear nervous 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[10,11,12,13.] 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.

Conclusion:

Although Dyke- Davidoff-Masson syndrome is a rare condition, its typical radiological findings can derive its diagnosis in the background of appropriate clinical features.

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. **Alpers B J and Dear R B**. Hemiatrophy of the brain. J Nerv Ment Dis 1939; 89: 653-651.

3. **Zilkha A**. CT of cerebral hemiatrophy, AJR 1980; 135: 259-262.

4. **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.

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

6. **Solomon G E, Hilal S K, Gold A P, et al.** Natural history of acute hemiplegia of childhood. Brain 1970; 93: 107-120.

7. **Teal J S, Rumbaugh C L, Bergeron R T, et al.** Congenital absence of internal carotid artery associated with cerebral hemiatrophy, absence of the external carotid artery. AJR 1973; 18: 534-545.

8. **Parker J C, Gaede J T**. Occurrence of vascular anomalies in unilateral cerebral hypoplasia "cerebral hemiatrophy". Arch Pathol Lab Med 1970; 90: 265-270.

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

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

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. 212-214.

12. **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.

13. **Garg RK, Karak B**. Cerebral hemiatrophy: a possible etiological relation with febrile seizures. Indian Pediatr 1998; 35: 79

Correspondence Address: Dr.*Naba Raj Koirala,* Professor and Head, Department of Psychiatry and Mental Health, Nobel Medical College and Teaching Hospital, Biratnagar. E-mail: drnabaraj@yahoo.com.