

Dyke-Davidoff Masson Syndrome : Presentation as Hemiplegic Cerebral Palsy



Medical Science

KEYWORDS : Dyke-Davidoff –Masson Syndrome , hemiatrophy , sepsis.

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ABSTRACT

Dyke-Davidoff Masson Syndrome is characterized by atrophy of cerebral hemisphere and distinctly being unilateral (hemiatrophy) and varied presentation of the contralateral side of the body . It can be congenital or acquired due to insult on central nervous system leading to loss of volume of one cerebral hemisphere. We present case of a 7 years old female child with DDMS secondary to probable sepsis and hemorrhage during the early neonatal period.

INTRODUCTION :

Acquired Dyke-Davidoff -Masson Syndrome is also known as hemispheric atrophy . It was 1st described by Dyke Davidoff and Masson in 1933 (1) on plain skull skiagram and pneumatoencephalogram. Alpers and Deer defined two types of cerebral hemiatrophy – congenital and acquired (2).

Congenital is seen secondary to intrauterine vascular occlusion or malformation in utero & in neonatal period (3). Acquired variety includes Birth trauma , Perinatal intracranial hemorrhages , Rasmussen encephalitis , Postictal hemiatrophy, infections and vascular abnormalities (4) . Plain X – Ray skull , computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) are helpful in diagnosis of Dyke-Davidoff -Masson Syndrome. Here we present case of Dyke-Davidoff -Masson Syndrome with unilateral motor involvement.

CASE REPORT :

A 7 year old female child presented in the OPD with complaint of disability and weakness of leg arm and left leg . Parents also complained that the child is not able speak properly on detailed history , birth history had significant evidence of umbilical hemorrhage and umbilical sepsis leading to CNS infection .

As per stated by her parents , the development of the child was delayed globally . Later parents noticed that child is not able to perform activities from left arm and has weakness in the left leg . There is no other significant past history .Family history showed no related details or significance on examination ;vitals-HR:80/min,RR=24/min ; temp =Afebrile and B.P =100/68mmHg. No pallor, icterus, clubbing or cyanosis seen. Bilateral cervical lymph nodes were palpable but were found to be in significant. On general examination deformity in left hand and feet was found. The tone was found to be increased in both left hand and leg. The plantar reflex was found to be unequal- left showed extensor (Babinski sign positive) and right showed flexor (normal response). No other systemic finding was found to be significant.

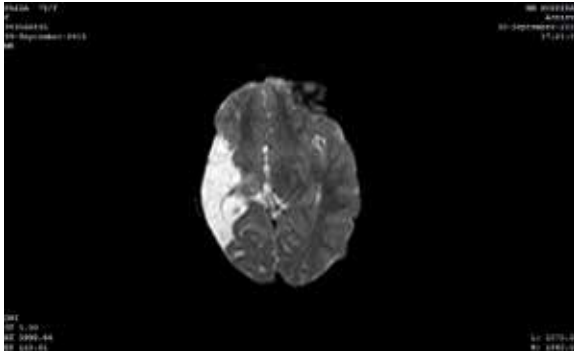
Speech assessment showed that the language perception was received properly but formation of words and delivery of same was affected. Hearing and intelligence Quotient was found to be normal. Provisional diagnosis of spastic Hemiplegic cerebral palsy was made and child was subjected to further radiological inves-

tigation in form of MRI-brain. Report was suggestive of :Chronic infarct right MCA territory with encephalomalacia and gliosis with Wallerian degeneration with changes of right cerebral hemiatrophy (Acquired Dyke – Davidoff – Masson syndrome).

Fig 1: Patient showing the involvement of left side of the body.



Fig 2: MRI finding showing involvement of the right side of the cerebral hemisphere.



DISCUSSION :

Similar to case presented , cerebral hemiatrophy is an entity with diverse etiologies and pronounced asymmetry of cerebral hemisphere(5). Its well known that the cause of Dyke-Davidoff -Masson Syndrome can be congenital (Primary) or acquired (secondary). In our case the supporting evidences towards a secondary cause is well d

Justified with a fact of positive history of insult during early infancy . Post natal causes include diverse etiologies such as traumatic CNS injuries , infection , tumor , ischemic and hemorrhagic insults and prolonged febrile convulsions(6).

The fact that any vascular insult before 3 years of age shows finding of compensatory thickening of calvarium and enlargement of sinuses to fill vacuum created by hypoplastic brain . As evidenced same in our case that with the wallerian degeneration there is ipsilateral calvarian thickening with loss of volume of right cerebral Hemisphere ; rightly explaining the involvement of left half of the body.

If the insult occurs in intrauterine life , there will be shifting of midline structures towards the affected side and prominence of sulcuses replacing absent gliotic tissues . These features can differentiate affection of the brain in utero from early life (7). Conditions that are associated with cerebral hemiatrophy such as Rasmussen encephalitis , Sturge weber syndrome , some brain tumors , Silver's syndrome , linear nervous sebaceous syndrome and progressive multifocal leucoencephalopathy should be differentiated . Cerebral hemiatrophy without seizure most likely cause due to cerebrovascular disease(8).

Treatment modalities are still to be optimized . It is basically symptomatic targeting convulsions , hemiparesis , learning difficulties , etc. Functional hemispherectomy with 85% success rate (9,10). Although in our case patient lost the follow up.

CONCLUSION :

Dyke-Davidoff -Masson Syndrome is a rare disease encountered. It can be diagnosed with detailed history and a good radiological support. Treatment is basically symptomatic and the prognosis is found better if hemiparesis is started after the age of two years.

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