Case Report

Dyke-Davidoff-Masson syndrome as sequelae of typhoid encephalitis?

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Abstract

Dyke-Davidoff-Masson syndrome (DDMS) is characterized by cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses in association with contralateral hemiplegia, seizures, mental retardation, difficulty, and impairment of speech development. Among the various complications of typhoid fever, neuropsychiatric manifestations constitute a major portion. However, DDMS post typhoid encephalitis has not been reported in the literature. We report a case of DDMS in an 8-year-old boy who presented with multiple seizures, impaired speech, behavioral changes, and mental retardation following typhoid encephalitis.

Key words: *Dyke-Davidoff-Masson syndrome, Hemiatrophy, Seizures, Typhoid encephalitis*

Dyke-Davidoff-Masson syndrome (DDMS) is characterized by unilateral atrophy of cerebral hemisphere, enlargement of the ipsilateral sulci, ventricles and cisternal spaces, decrease in the size of ipsilateral cranial fossae, overdevelopment of paranasal sinuses and mastoid air cells, and unilateral thickening of the skull. It can be congenital or acquired and is usually seen during adolescence. However, it can also be seen in childhood which is secondary to brain insult in fetal or early childhood period. We report a case of an 8-year-old boy who presented with multiple seizures, behavioral changes, progressive impairment of speech, and mental retardation after an episode of typhoid fever with encephalitis and on magnetic resonance imaging (MRI) was diagnosed to have DDMS. A high incidence of neuropsychiatric manifestations of typhoid has been reported during different stages of typhoid fever. However, on extensive literature search, we could not find cerebral hemiatrophy as a long-term complication of typhoid encephalitis.

CASE REPORT

An 8-year-old boy presented to the emergency room with history of recurrent episodes of tonic, clonic and myoclonic seizures, several episodes in a day since 5 months. There was progressive impairment of speech in the form of slurring of speech, difficulty in articulation, and decreased speech output. This was associated with abnormally aggressive and wandering behavior and progressive learning disability. Motor, sensory, cerebellar, and cranial nerve examination was normal. There was no facial asymmetry. Child was a product of normal vaginal delivery with an uneventful postnatal period. Developmental milestones were normal.

Six months prior to this presentation, he was admitted in our hospital with a history of high grade fever for 10 days, altered sensorium for 2 days and one episode of tonic clonic convulsion. On examination, there was relative bradycardia with hepatosplenomegaly (liver-2 cm, spleen-1 cm soft). Investigations revealed leukopenia, positive IgM Typhidot with widal titres of To-1/320, TH-1/320. Blood culture was, however, sterile. Contrast-enhanced computed tomography was normal. Malaria antigen test and peripheral smear for malarial parasite were negative. He was diagnosed and treated as enteric encephalitis. On discharge, his neurological status was normal.

Blood and cerebrospinal fluid studies were unremarkable. During this presentation MRI of the skull revealed a generalized dilatation of the cortical sulci and lateral ventricles in the left cerebral hemisphere, consistent with left cerebral hemiatrophy (Figures 1 and 2).

DISCUSSION

DDMS is a rare condition which was first described by Dyke et al. in 1933 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. Age of presentation depends on time of neurologic insult, and characteristic changes may be seen only in adolescence. Male gender is more commonly affected. Furthermore, left hemisphere involvement is more frequent. The clinical findings may be of variable degree depending on the extent of the brain injury. Seizures may not be present initially and may appear many months or years after onset of hemiparesis. Similarly, hemiparesis may not be present initially and may appear sometime after the onset of the seizure [2]. Varying degrees of atrophy of one half of the body, sensory loss, speech and language disorder, mental retardation or learning disability, and psychiatric manifestations like schizophrenia may also be present [3].

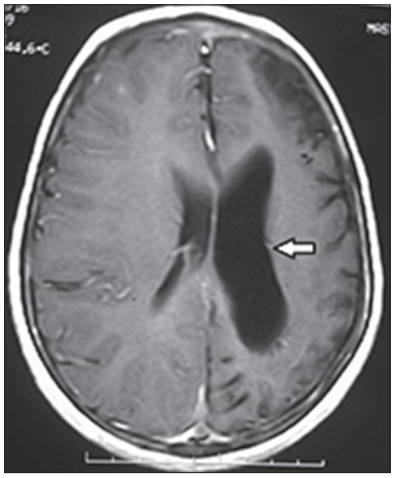

Figure 1: Flair magnetic resonance imaging sequence showing left cerebral hemiatrophy with dilatation of ipsilateral ventricle and midline shift

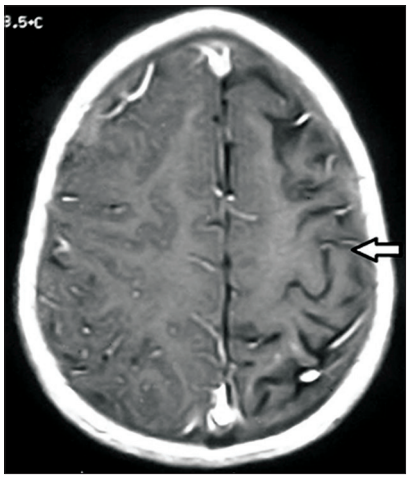

Figure 2: Flair magnetic resonance imaging sequence T2W showing dilatation of cortical sulci

In our patient, however, some of the typical clinical manifestations such as hemiparesis and facial asymmetry were not present. This may be due to early diagnosis, and the typical clinical features may evolve eventually.

Imaging studies help in clinching the diagnosis. There is unilateral atrophy of the cerebral hemisphere with ipsilateral shift of the ventricles. The gyri on the involved side are wide and often replaced by gliotic brain tissue. There is ipsilateral osseous hypertrophy and hyperpneumatization of sinuses [1,4]. All these changes were present in our patient.

DDMS has been reported in association with cerebral malaria [5] and encephalitis [6,7] after few years of its occurrence. The exact mechanism of cerebral atrophy is still unclear in either type. It is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which in turn lead to cerebral atrophy [8].

Vascular occlusions related to cerebral malaria were postulated as the cause of acquired cerebral changes [5].

Infantile (congenital) type of DDMS, in contrast to adult (acquired) DDMS, shows enlargement of calvarium, diploic space, and paranasal sinuses [6]. Prominent sulcal spaces will be absent as ischemia occurs during embryogenesis when the formation of gyri and sulci is deficient. In contrast, the atrophied cerebral hemisphere will have prominent sulcal spaces if the vascular insult occurs after birth [2].

In the present case, the findings of dilated cerebral sulci and lateral ventricles with ipsilateral midline shift and a relatively acute course reflects a late onset of brain insult probably due to inflammation and or vascular occlusion secondary to typhoid encephalitis.

Neuropsychiatric complications in typhoid fever range from 5% to 35% in various studies. Most of the neurological complications are seen during the acute course of illness, but some like motor neuron disease, cognitive deterioration occur well after recovery [9]. Hemiatrophy of cerebral hemispheres as a sequelae of typhoid encephalitis presenting late after recovery has not been documented in the literature.

The possible mechanisms responsible for the neurological manifestations of typhoid fever are hyperpyrexia (>43°C), fluid and electrolyte disturbances, typhoid neurotoxin, vasculitis with peri-vascular cuffing, autoimmune mechanism, pressure effect on blood vessels resulting in cerebral infarction and acute disseminated encephalomyelitis [9,10] post mortem histology in fatal cases has revealed congestion, diffuse edema, and perivenous lymphocytic infiltrations.

We contemplate that the cause of cerebral hemiatrophy in our case could be vasculitis or other vessel-related changes occurring in typhoid fever as mentioned above. MR angiography if performed at that time could have revealed vasculopathy. Hence, it is prudent to look for DDMS as sequelae, particularly in tropical countries where enteric fever is endemic, if the child presents with neurobehavioral changes and or seizures post enteric encephalitis.

Other differential diagnoses to be considered in a patient of cerebral hemiatrophy are Sturge-Weber Syndrome, Basal ganglia germinoma, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome, and Rasmussen encephalitis.

REFERENCES

1. Dyke CG, Davidoff LM, Masson LB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinus. Surg Gynecol Obstet. 1933;57:58.
2. Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE, Jacoby CG. Cerebral hemiatrophy, hypoplasia of internal carotid artery, and intracranial aneurysm. A rare association occurring in an infant. Arch Neurol. 1987;44(2):232-5.
3. Sener RN, Jinkins JR. MR of craniocerebral hemiatrophy. Clin Imaging. 1992;16(2):93-7.
4. Zilkha A. CT of cerebral hemiatrophy. AJR Am J Roentgenol. 1980;135(2):259-62.
5. Karuppiah S, Rodgman C, Lombard J. Dyke-Davidoff-Masson syndrome in postcerebral malaria. J Child Neurol. 2009;24(4):487-90.
6. Atalar MH, Icagasioglu D, Tas F. Cerebral hemiatrophy (Dyke-Davidoff-Masson syndrome) in childhood: Clinicoradiological analysis of 19 cases. Pediatr Int. 2007;49(1):70-5.
7. Shetty DS, Lakhkar BN, John JR. Dyke-Davidoff-Masson syndrome: Neuroimage. 2003;51(1):136.
8. Lee JH, Lee ZI, Kim HK, Kwon SH. A case of Dyke – Davidoff – Masson syndrome in Korea. Korean J Pediatr. 2006;49(2):208-11.
9. Osuntokun BO, Bademosi O, Ogunremi K, Wright SG. Neuropsychiatric manifestations of typhoid fever in 959 patients. Arch Neurol. 1972;27(1):7-13.
10. Haque A. Neurological manifestations of enteric fever. In: Chopra JS, Sawhney IM, editors. Neurology in Tropics. India: BI Churchill Livingstone; 1999. p. 506-12.

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