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Case Report

Acquired dyke-davidoff-masson syndrome: a clinicoradiographic correlation

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Abstract. Dyke-Davidoff-Masson syndrome (DDMS) is characterized by unilateral cerebral atrophy or hypoplasia (hemiatrophy), ipsilateral osseous hypertrophy with hyperpneumatization of paranasal sinuses accompanied with varying degrees of contralateral hemiparesis, hemifacial atrophy and mental retardation. DDMS may be infantile or acquired later in life as a result of trauma, infection, ischemic or hemorrhagic insult to the central nervous system (CNS). We present here an 8-year-old girl with DDMS secondary to a febrile CNS infection at the age of 2 years.

Key words: Dyke-Davidoff-Masson syndrome; unilateral cerebral hemiatrophy; hemiparesis; seizure

1. Introduction

DDMS takes place in the differential diagnosis of unilateral cerebral atrophy with ipsilateral hypertrophy. Age of clinical presentation and appearance of the entire spectrum of characteristic clinical and radiological features depends upon the timing of the CNS insult.

2. Case report

An-8-year-old ambidextrous girl presented to us with a history of a generalized tonic clonic seizure (GTCS). She was apparently well till the age of 2 years when she had had an acute febrile illness following which she had developed partial seizures confined to the right side of the face and the right upper limb. Her partial seizures had been well controlled with sodium valproate till about 10 days ago when she had her first GTCS prompting a visit to our emergency room (ER). She was the product of a non-consanguineous marriage with no complications reported in the antenatal or post-natal period. She was born full term and had a birth weight of about 6.6 pounds. Childhood vaccination record was up to date and she attained all milestones till around 2 years of age when she had had an acute febrile illness necessitating intensive care. Details about this febrile illness were not available but since then she had a history of weakness at the right side of her body. Over time, the parents also noticed that the right side of her face and her right arm and leg were smaller as compared to the left. At the time of presentation to our ER, she was a fourth grade student of average scholastic performance. Examination revealed a thin built girl with right hemiatrophy involving the face and the upper and lower limbs (Fig 1, 2, 3). Deep tendon reflexes were brisker on the right as compared to the left. She scored 20/30 in the mini-mental state examination (MMSE). No neurocutaneous stigmata were identified. Non contrast head computed tomography scan (NCCT) revealed evidence of irregular large gliotic area in the left frontal temporal region extending up to the

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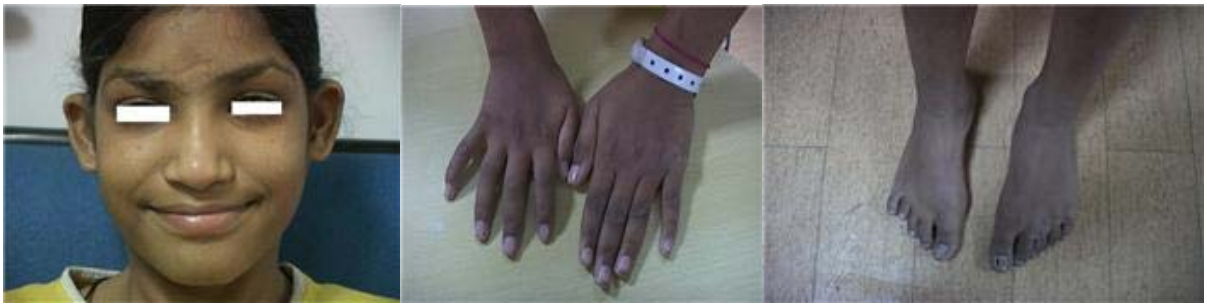

Fig. 1, 2, 3. Photos of the patient show hemiatrophy involving the right face and upper and lower limb.

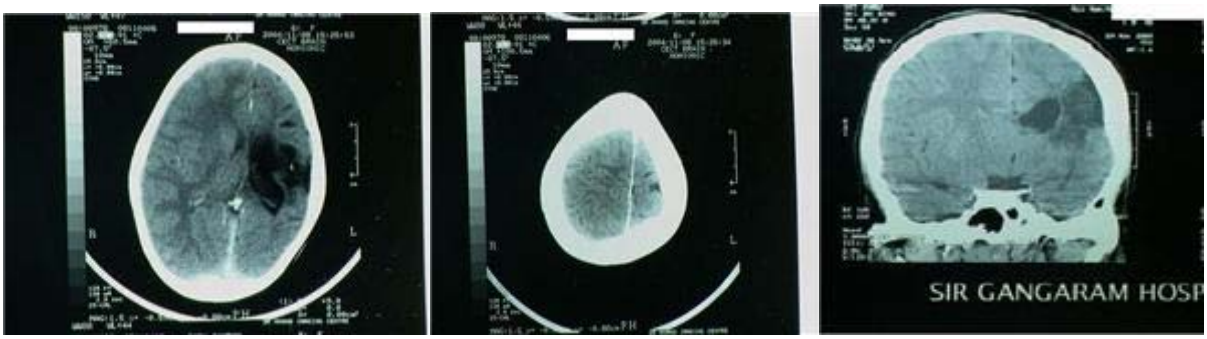

Fig. 4, 5, 6. NCCT shows irregular large gliotic area in the left frontal temporal region extending up to the cortical surface with thickening of the ipsilateral left overlying calvarium with smaller size of the left cerebral hemisphere.

cortical surface with thickening of the ipsilateral left overlying calvarium with smaller size of the left cerebral hemisphere (Fig. 4,5,6). NCCT scan of the paranasal sinuses revealed mucosal thickening in the right sphenoid and right posterior ethmoid air cells with prominence of the left sphenoid and left frontal sinuses (Fig. 7). The possibility of acquired DDMS was considered. The electroencephalogram showed left frontoparietal localization related spike wave discharges.

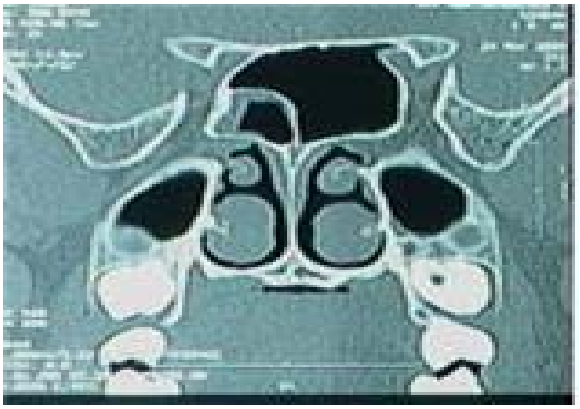

Fig 7. NCCT scan of the paranasal sinuses shows mucosal thickening in the right sphenoid and right posterior ethmoid air cells with prominence of the left sphenoid and left frontal sinuses.

3. Discussion

Dyke-Davidoff-Masson syndrome first described by Dyke, Davidoff and Masson in 1933 is characterized by asymmetry of the cerebral hemispheric growth with hemiatrophy, ipsilateral osseous hypertrophy with hyper pneumatization of the paranasal sinuses accompanied with varying degrees of contralateral paresis (1,2). The etiology of DDMS can be classified as congenital or primary and acquired or secondary. Congenital hemiatrophy is usually a sequelae of intrauterine carotid artery occlusion or hypoplasia. Other prenatal and perinatal causes include hypoxic ischemic encephalopathy (HIE), birth trauma, coarctation of midaortic arch, amniotic bands and intracranial hemorrhage. Included among the postnatal causes of DDMS are diverse etiologies such as traumatic CNS injuries, infection, tumor, ischemic and hemorrhagic insults and prolonged febrile seizures (3,4). In their clincoradiological review of 19 cases of DDMS, Atalar et al. documented 12 cases as congenital and 7 cases as acquired type. Among the acquired cases, CNS infections were described in 2 patients, cranial trauma in another two and postnatal hypoxic ischemia in the remained three. A history of seizures was documented in all 19 cases (5). Chung et al. reported a 23-year-old man with

DDMS associated with angiographically documented occlusion of the internal carotid artery while Karuppiah et al. reported the syndrome in an 18-year-old emigrant woman from Ghana in association with cerebral malaria at 13 years of age (6,7). Age of clinical presentation and appearance of characteristic clinical and radiological features depends upon the timing of the insult. Hence it is the infantile (congenital) type of DDMS which shows enlargement of the calvarium and of the paranasal sinuses and diploic space. It is hypothesized that the compensatory skull changes reflect an adaptation to the unilateral decrease in brain parenchymal volume. Various changes have been documented in the literature namely ipsilateral calvarial thickening (of both the inner table and diploic space), loss of convolutional markings of the inner table of skull, overgrowth of paranasal sinuses, elevation of the petrous ridge and sphenoidal wing and hypoplasia of the floor of the anterior and middle cranial fossa. Brain development proceeds at a rapid pace in the newborn child reaching three fourths of the adult full size by the third year of life. Thus another useful marker to indicate the timing of the CNS insult is the gyral and sulcal formation of the hemiatrophic brain. If vascular insult occurs during embryogenesis no prominent sulci are visualized while prominent sulcation is seen if vascular insult occurs after birth. Differential diagnosis of cerebral hemiatrophy apart from DDMS includes neurocutaneous syndromes such as Sturge-Weber, leucodystrophies in the atropic stage and Rasmussen's encephalitis (hemiconvulsion-hemiplegia-epilepsy syndrome) (8). Children with DDMS and with intractable disabling seizures are candidates for hemispherectomy which is successful in eliminating or substantially reducing seizures in as many as 85% of carefully selected cases. Prognosis is better for children who develop hemiparesis after the age of 2 years in the absence of prolonged or recurrent seizures.

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