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Dyke Davidoff Masson Syndrome

Authors
Sree Siva Kumar Raja Addagalla[1], Jagadeesh Metta[2], Udaya Bhaskar Pamarthi[3]
RMC, KAKINADA

Abstract
Dyke Davidoff Masson syndrome is a rare condition with variable degree of facial asymmetry, contralateral hemiparesis / hemiplegia, seizures with a radiological finding of asymmetry of cerebral hemisphere growth with atrophy on one side usually associated with ipsilateral osseous hypertrophy and hyperpneumatisation of sinus.
Keywords: *Dykes Davidoff Masson Syndrome, ExVacuo dilatation, Cerebral hemiatrophy.*

Introduction

Dyke Davidoff Masson syndrome is a rare condition with variable degree of facial asymmetry, contralateral hemiparesis / hemiplegia, seizures with a radiological finding of asymmetry of cerebral hemisphere growth with atrophy[1] on one side usually associated with ipsilateral osseous hypertrophy and hyperpneumatisation of sinus.

Case Report

A 30-year-old female patient was brought to emergency department with chief complaint of generalized tonic clonic seizures for 2 days, with about 4-5 episodes per day, associated with tongue bite, involuntary micturition & postictal confusion for about 15-20 minutes.

Past history revealed that she had similar complaints since the age of 7 years and has been using anti epileptics since then but stopped using since last 1 month. There is no history suggestive of any CNS infections in childhood, head injury or any delayed developmental milestones.

On examination patient has been found to have facial asymmetry with left half being smaller relative to right half of the face. On further examination she was found to have mild weakness of left upper limb and lower limb and also has mirror movements.

Investigations

MRI Brain was done which revealed diffuse atrophic changes in right cerebral hemisphere with ex-vacuo dilatation of Lateral Ventricle along with right sided hyper pneumatization of Frontal sinus.

In the present case scenario, the finding of right cerebral hemiatrophy with enlarged cortical sulci with ex-vacuo dilatation of Lateral Ventricle and history of seizures and evidence facial asymmetry and hemiparesis suggest a diagnosis of DDMS.

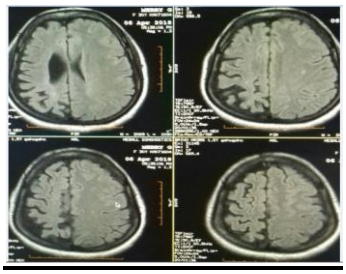

Fig: 01

Discussion

Patient with DDMS usually present with refractory seizures and should focus on control of seizures and treatment should focus on control of seizures with suitable anticonvulsants may be needed. Along with drugs, physiotherapy, occupational therapy and speech therapy play a significant role in long-term management. Prognosis is better if the onset of hemiparesis is after 2 years age and in absence of prolonged or recurrent seizure[1][2].

Hemispherectomy is the treatment of choice for children with intractable disabling seizures hemiplegia with a success rate of 85% in selected cases

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