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MOLECULAR MIMICRY AT PLAY: POST-MUMPS ADEM IN A FULLY VACCINATED CHILD

DR.SUENERA .P.V[1], DR.RADHA KUMAR[2], DR.PRADEEP S[3], DR.JOSHUA RAJAN X[4], DR. SAVITHA LAKSHMI[5]

[1,2,3,4]PAEDIATRICS, SAVEETHA MEDICAL COLLEGE ANDHOSPITAL, SAVEETHA INSTITUTE OF MEDICAL AND TECHNICALSCIENCES(SIMATS), SAVEETHA UNIVERSITY, CHENNAI ,INDIA

[5]SENIOR LECTURER, DEPARTMENT OF ORAL& MAXILLOFACIAL SURGERY, SREE BALAJI DENTAL COLLEGE & HOSPITAL, CHENNAI, INDIA

CORRESPONDING AUTHOR -DR.JOSHUARAJAN,
e-mail: pvsk.med@gmail.com

SUMMARY

Acute disseminated encephalomyelitis (ADEM), being an inflammatory, demyelinating disorder following an infection or immunization is due to molecular mimicry induced by the exposure triggering CNS autoantigen production. We report a case of a developmentally normal ,fully immunized child who developed weakness of both the upper and lower limb with features of encephalopathy with MRI brain showing hyperintensities of the basal ganglia ,thalamus ,pons and mid brain .The weakness followed an infection with mumps a week earlier ,which was proved serologically by raised IgM titres in blood .The Child was symptomatically manged and responded favourably with pulse methylprednisolone therapy and intravenous immunoglobulin therapy .This case is reported due to the rarity of presentation of a ADEM following mumps infection in a fully vaccinated child .

BACKGROUND

The incidence of mumps virus infection, known for its high neuroinvasiveness, has significantly decreased following the inclusion of the MMR vaccine immunization programs worldwide. Despite its rarity post-vaccination, mumps infections can still occur, often presenting with benign meningitis during the acute phase. ADEM an important complication of mumps, is an autoimmune inflammatory condition of the CNS, believed to be immune-mediated as it often follows an infection or immunization in up to three-fourths of cases. The estimated annual incidence ADEM is 0.2 to 0.5 per 100,000 children. The average age of onset in children is between 3 to 7 years (1)

CASE PRESENTATION

A 9 yearold developmentally normal, fully immunized child presented with complaints of inability to sit and stand without support ,withweakness which initially involved the lower limb and then progressed to involve the upper limb since 1 week . Child had history of fever 1 week earlier which was associated with swelling of the both parotid glands. Child did not have any other gastrointestinal, respiratory or genitourinary symptoms .On examination, the vitals were stable and pupils were equally reactive to light with a GCS of 14 (E4V4M6).Neurological examination revealed reduced power in both upper and lower limbs (upper limb - 3/5 and lower limb power of 2/5 by MRC grading). Deep tendon reflexes were absent in both lower limbs with plantar reflex being extensor in both limbs.

INVESTIGATIONS *If relevant*

Blood investigations revealed normal complete blood count,negative C Reactive protein with normal liver and renal function tests and no growth in blood culture .

In view of the noncontributory initial lab results with no growth detected in blood, CSF studies were ordered which revealed lymphocytosis(100% lymphocytes;n=13), no growth in culture, negative results in CSF AFB, HSV, Mumps and enterovirus studies. However, IgM positive was noted for mumps virus.

His MRI Brain showed hyper intensities of midbrain, cerebellum, basal ganglia and pons.

T1-Few symmetrical hypointensities and T2-Hyperintensities in the bilateral basal ganglia and bilateral mammillary bodies,bilateral postero superior and posteroinferior cerebellum

T1W- Symmetrical hypointensities,T2W-hyperintensities in the mid brain tegmentum

T1W-Symmetrical subtle hypointensities,T2W-hypointensities in the pons of the medial longitudinal fasciculus and central tegmental tract ,hyperintensity in the aqueduct opening into the fourth ventricle

NEUROMYELITIS OPTICA MYELIN OLIGODENDROCYTE ANTIBODY -negative
NEUROMYELITIS OPTIC AQUAPORIN ANTIBODY-negative

DIFFERENTIAL DIAGNOSIS *If relevant*
The differential diagnosis which were considered were mumps encephalitis which is due to direct invasion of the mumps virus into the CNS but mumps virus not detected in CSF studies .The second differential diagnosis which was considered was Wilson's disease because of the presence of hyperintensities of the basal ganglia ,urinary copper levels and serum ceruloplasmin levels sent were in normal range ,there was no KF ring on Ophthalmic examination hence Wilson's disease was ruled out .Third differential diagnosis which was considered was Thiamine basal ganglia disease in view of waxing and waning course hence thiamine and biotin supplements were added

TREATMENT
Considering the initialdiagnosis of Viral Encephalomyelitis, child was started on Inj ceftriaxone and Inj acyclovir. There was no improvement following antibiotics and Antivirals.Pulse dose of Inj methylprednisolone @30 mg/kg (600 mg IV OD) was given for 5 days .Child was intubated on day 6 of hospital stay in view of poor respiratory efforts.Since there wasdeteriorationin power of the upper limb to 2/5 and lower limbspower to 1/5 Intravenous Immunoglobulin 2g/kg was given in 4 divided doses over the course of 2 days .Oral steroids (prednisolone) was given at a dose of 1 mg/kg/day and later tapered to 0.5 mg/kg /day.Child was extubated on day 25 of hospital stay after respiratory efforts improved . Child had abnormal facial movements and neurology opinion was obtained and further started on sodium valproate. In view of increasing extra pyramidal symptoms , child was started on bromocriptine and trihexyphenidyl .Regular physiotherapy and swallowing interventions were added in addition to high protein diet.
OUTCOME AND FOLLOW-UP
Child was given regularly physiotherapy and was continued on maintenance dose of steroids and is now able to walk with support .Power of the upper and lower limbs improved to 4/5atdischarge . Child was initially on Ryle's tube feeds now is able to swallow well and is able to take food on his own .regular physiotherapy and speech therapy was continued
On follow up child's power was improved to 5/5,his speech improved and he was able to talk in 3-4 wordsentences

DISCUSSION

Mumps, caused by a single-stranded, pleomorphic RNA virus from the Paramyxoviridae family is typically marked by fever and swelling of the parotid glands, either on one or both sides of the face. The central nervous system (CNS) is the next most frequently affected area after the salivary glands in children.(2) Meningitis occurs in 1%–10% of mumps cases, usually with a favorable prognosis, whereas mumps encephalitis, occurring in less than 1% of cases, can be fatal. The underlying pathology of mumps encephalitis may involve either direct viral invasion of the CNS, leading to primary meningoencephalitis, or immune-mediated demyelination, which typically arises days to weeks after the initial onset of parotitis.ADEM, or acute disseminated encephalomyelitis, is an immune-related neurological disorder marked by inflammation and the loss of myelin in the brain's white matter; often follows a viral infection or vaccination(3)
Diagnosing ADEM relies on clinical presentation and imaging findings, with MRI being the most crucial diagnostic tool. The MRI results can vary greatly, typically showing lesions in deep and subcortical white matter, and/or gray matter lesions in areas like the thalami and basal ganglia, with sizes ranging from 5 mm to over 5 cm.(2)
Symptoms of ADEM generally emerge 1-3 weeks after the onset of a mumps virus infection. Despite the likely post-infectious origin suggested by the recent history of illness, initial treatment with antimicrobials is recommended until an active infection is definitively excluded. The primary treatment for ADEM is high-dose steroid therapy. If the patient does not respond to steroids, plasma exchange and intravenous immunoglobulin may be considered.
In the case report by Verma et al., a 3-year-old girl presented with fever, weakness in all four limbs, urinary retention, respiratory distress, and altered sensorium. MRI scans revealed multiple cerebral T2-hyperintense signals and extensive spinal cord hyperintensities. The patient was diagnosed with ADEM and showed complete clinical and radiological improvement following treatment with intravenous methylprednisolone within one month.(2)
Another case reported by Jha et al. involved an 8-year-old girl who developed ADEM after a confirmed mumps infection. She presented with loss of consciousness, right-sided weakness, and myoclonic seizures. MRI findings and cerebrospinal fluid analysis supported the diagnosis of ADEM. She was treated with intravenous methylprednisolone, oral sodium valproate, baclofen, and oral prednisolone, showing significant improvement within four weeks.(3)
A third case described by Gonçalves et al. 21-year-old male who developed ADEM after mumps infection despite being vaccinated. He presented with disorientation, lower limb weakness, and urinary retention. MRI scans showed lesions in the splenium of the corpus callosum and the anterior arm of the right internal capsule. Despite initial antimicrobial treatment, his condition worsened, leading to the diagnosis of ADEM and subsequent treatment with high-dose steroids, resulting in partial motor recovery.(4)

Analysis of the previously reported cases show that all had problems in sensorium along with motor weakness. These case reports illustrate the variability in clinical presentations and outcomes of ADEM following mumps infection. In the present study, we report a similar weakness affecting both limbs bilaterally witha power of 2/5 in upper limb and 2/5 in lower limb. The child subsequently developed loss of neck control, swallowing dysfunction and deterioration of power in lower limbs ,loss of speech and choreoathetoid movements. Though pulse high dose steroid and Immnunoglobulin therapy were given, there was no immediate improvement with clinical course showing waxing and waning of improvement.Child was offered regular physiotherapy and training for swallowing dysfunction and speech.However, at discharge power in upper and lower limb power improved to3/5.on follow up of child after 1 month his power improved to 5/5 and child was able to carry on his daily activities .

LEARNING POINTS/TAKE HOME MESSAGES *3-5 bullet points*

- ADEM is a clinical diagnosis supported by MRI ,CSF and serum findings
- Vaccination does not fully rule out the possibility of mumps and its complications
- Major mechanism of ADEM is molecular mimicry

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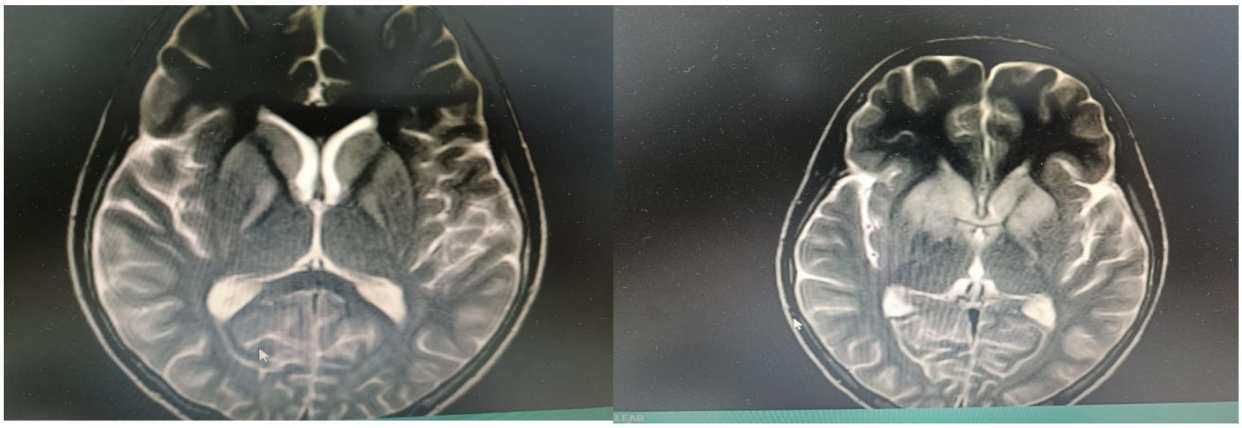
FIGURE/VIDEO CAPTIONS

FIGURE-1 -SHOWS THE MRI FINDINGS OF THE PATIENT

FIGURE 2-SHOWS THE REPEAT MRI FINDINGS OF THE PATIENT

FIGURE 1

INITIAL MRI FINDINGS OF THE CHILD



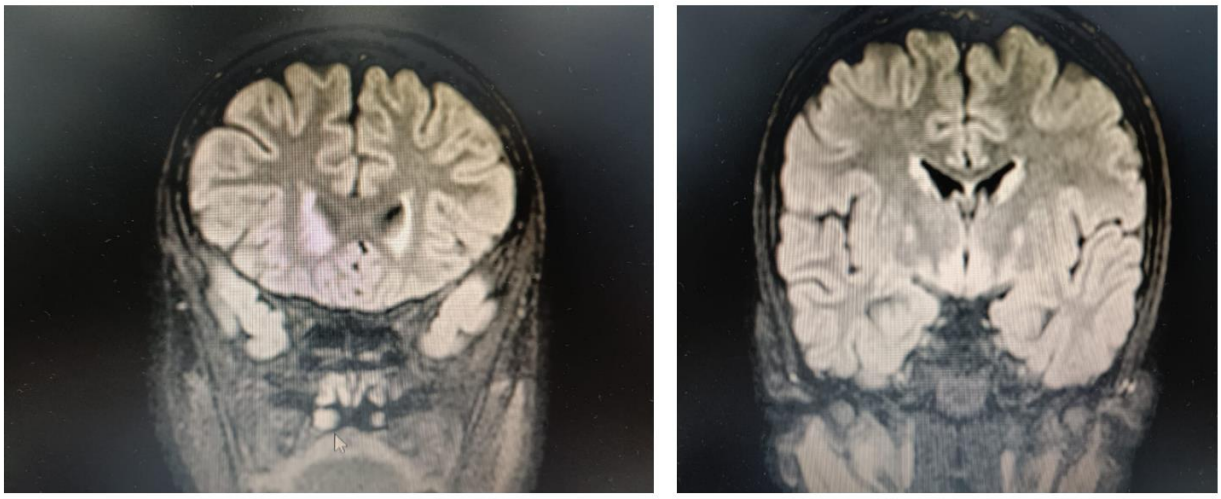
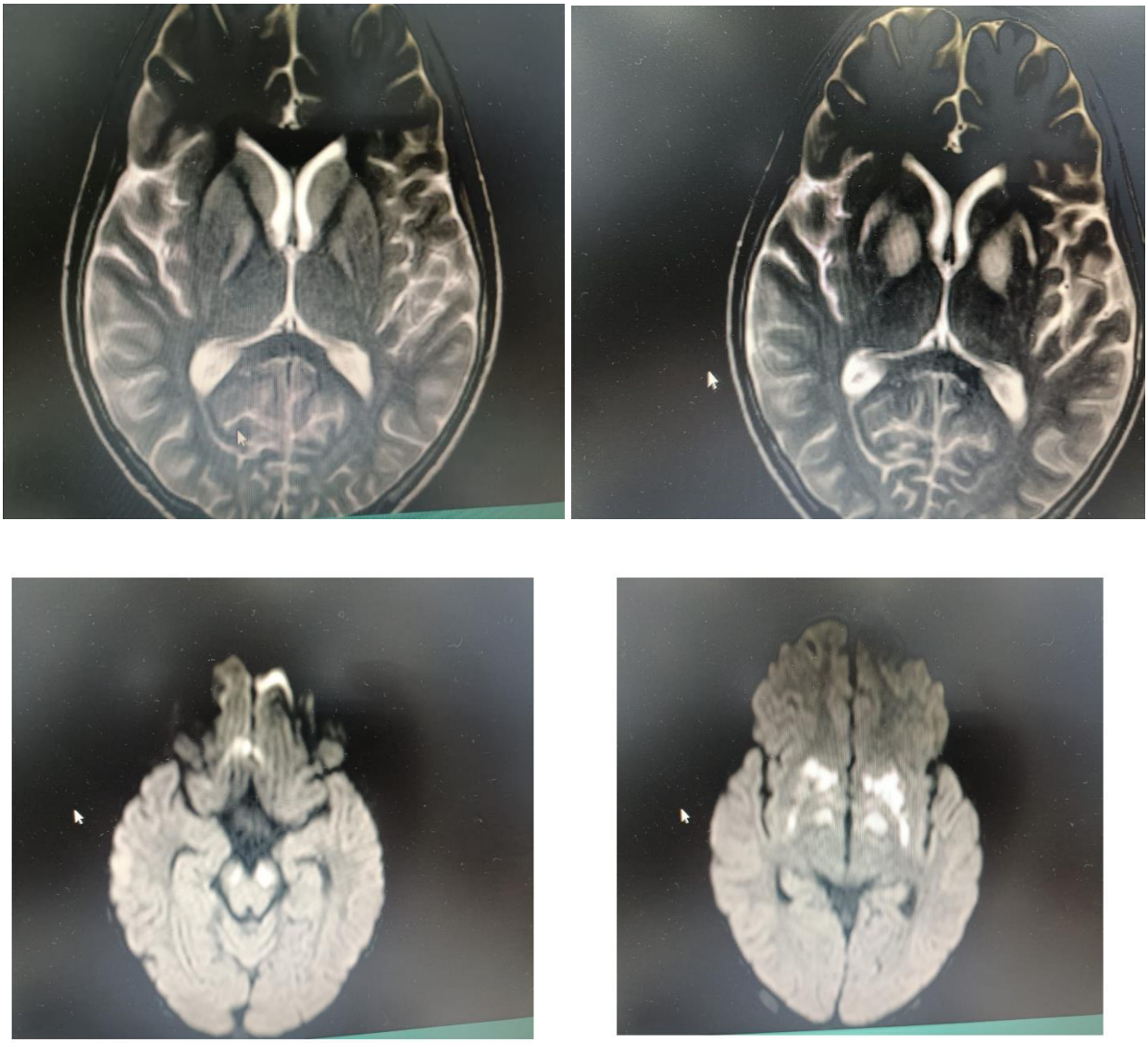


FIGURE 2

REPEAT MRI BRAIN OF THE CHILD



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