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Case Report

Gradenigo syndrome and acute disseminated encephalomyelitis: An infective and inflammatory combo

Subhashini K[1,*], **P Paranthaman**[1]

[1]*Dept. of Internal Medicine, Govt. Kilpauk Medical College and Hospital, Chennai, Tamil Nadu, India*

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ABSTRACT

A 14 year old male presented with complaints of giddiness for two days.He had a history of left sided ear pain and a purulent discharge from the left ear one month ago. He had signs of cerebellar ataxia.On examining his left ear three point tenderness and tragal tenderness was present. On Day 4 he developed diplopia, facial pain and a GCS of 15/15. On examination he revealed a left lateral rectus palsy. CT Brain showed no evidence of raised intracranial tension. MRI Brain showed evidence of encephalitis. A cerbrospinal fluid analysis was within normal limits. The patient was suspected with Acute Disseminated Encephalomyelitis (ADEM) following otitis media of the left ear with an active episode of mastoiditis causing Gradenigo syndrome. Patient was treated with intravenous antibiotics, steroids and his condition improved.

Key Messages: Acute meningoencephalitis and Acute Disseminated Encephalomyelitis need to differentiated at the earliest to facilitate a faster recovery and aslo to minimize the neurological deficits. Combination of an infectious and inflammatory pathology can pose a serious problem.

1. Introduction

This case mimic an acute meningoencephalitis but is actually a post infectious encephalitis with a Gradenigo syndrome producing cranial nerve palsies adding to the confusion. This case has overlapping components of central and peripheral causes of vertigo. Also this is a case of Acute Disseminated Encephalomyelitis with no episode of seizures.

2. Case History

Year old male presented with complaints of giddiness for two days.

He had a history of left sided ear pain and a purulent discharge from the left ear one month ago. He had no history of seizures. Patient had no comorbid illnesses. On admission patient vital signs were BP-11/70 mmHg Pulse rate of 88bpm, SPO_2 98% room air, respiratory rate of 15 per minute, capillary blood glucose level of 133mg/dL and Jugular venous pressure was not elevated. His cardiovascular, respiratory and abdominal examinations were unremarkable. On examining his nervous system, he had a Glasgow Coma Scale of 15/15. His speech and memory were intact. His cranial nerve examination showed no evidence of abnormalities on Day 1 of admission. His motor system examination was unremarkable and his deep tendon reflexes were in the grade of 2+. Bilateral Babinski's sign was seen. His cerebellar function examination revealed pastpointing of the upper limbs, intention tremors, a bidirectional nystagmus and an ataxic gait. He has no meningeal signs and his sensory system was intact. Cerbellar abscess was considered a possibility but was later ruled out by imaging studies. On examining his left ear three point tenderness and tragal tenderness was present.

On Day 4 he developed diplopia, facial pain and a GCS of 15/15. On examination he revealed a left lateral rectus palsy. CT Brain showed no evidence of raised intracranial

* *Corresponding author*.
E-mail address: subhashini95@gmail.com (Subhashini K).

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tension and a false localising sign was ruled out [Figure4]. There was no evidence of papilledema or optic neuritis.

MRI Brain showed a left mastoiditis with non diffusion restricting T2/FLAIR hyperintenities in left thalamus, left cerebral peduncle, tegmentum of midbrain, tegmentum of pons, superior, middle and inferior cerebellar peduncles.(Figures 1, 2 and 3)

Fig. 1: T2/FLAIR hyperintensity in left thalamus

Fig. 2: T2/FLAIR hyperintensity in left mid brain

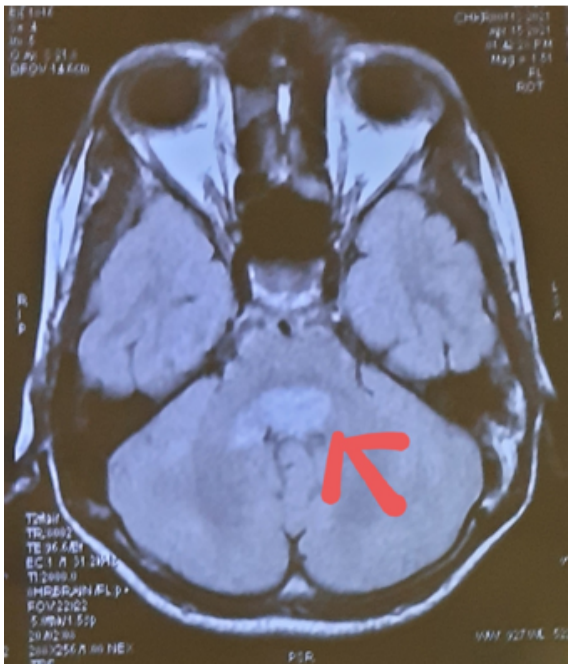

Fig. 3: T2/FLAIR hyperintensity in pons

Cerbrospinal fluid analysis showed sugar, protein and cellular counts within normal limits.

The patient was suspected of having an Acute Disseminated Encephalomyelitis following otitis media of the left ear with mastoiditis causing Gradenigo syndrome

Patient was treated with intravenous antibiotics, steroids and other supportive measures and his condition started to improve.

3. Discussion

Acute disseminated encephalomyelitis (ADEM) is an acute demyelinating disorder of the central nervous system, and is characterised by multifocal white matter involvement. Diffuse neurological signs along with multifocal lesions in brain and spinal cord characterise the disease.[1] ADEM is theorized to be an immunologically mediated demyelinating disease triggered by a febrile illness or recent vaccination, eliciting an inflammatory response affecting the central nervous system (CNS). Possible mechanisms may include either molecular mimicry or direct inflammatory damage to myelinated neurons.[2] ADEM can occur following any infection. The prevalence of ADEM is higher in children and young adults and is thought to be related to the increased frequency of viral infections and vaccination in this patient population.[3] The distinction between it and acute viral encephalitis is difficult but patients with ADEM respond better to steroids. Hence identifying it is very essential.

Gradenigo's syndrome is a rare disease, which is characterized by the triad of the following conditions: suppurative otitis media, pain in the distribution of the first

and the second division of trigeminal nerve, and abducence nerve palsy. The full triad may often not be present.[4] It is thought to be caused by infection spreading from the middle ear via air-cell tracts, as well as lymphatic and vascular channels, to the petrous apex of the temporal bone, causing the osteomyelitis termed 'petrous apicitis,' and may result in concurrent abducence and trigeminal nerve inflammation if the suppurative process proceeds to affect Dorello's canal and Meckel's cave. The triad of otorrhoea, facial pain in the distribution of the trigeminal nerve and ipsilateral abducence nerve palsy, doesn't often present in its classical form. In one retrospective study, only seven of forty-four patients (16%) had an abducence nerve palsy. Facial pains and otitis were a more common constellation, seen in thirty-seven patients (84%).[5,6]

The combination of ADEM and Gradinego syndrome pose a diagnostic dilemma as cranial nerve involvements can occur in both and each has a different management. The combination of an infective and inflammatory pathology also poses problems in administering drugs like steroids.

Hence the early differentiation between an infective and an inflammatory pathology, though difficult, is quite essential.

4. List of abbreviations

ADEM – Acute Disseminated Encephalomyelitis

5. Source of Funding

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6. Conflicts of Interest

There are no conflicts of interest.

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Author biography

Subhashini K, Junior Resident https://orcid.org/0000-0002-5895-1293

P Paranthaman, HOD

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