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Case Report

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Osmotic demyelination syndrome presenting with parkinsonism

Weerasinghe NT, Safeek LM, Peiris PJP & Azher S

National Hospital Kandy

Correspondence:
Weerasinghe NT
Registrar in Medicine, National Hospital Kandy
E-mail: thivankaw@gmail.com

: https://orcid.org/0000-0001-8979-0763

Abstract

Osmotic demyelination syndrome (ODS) is a condition which occurs due to rapid osmotic changes which result in myelinolysis of the nervous system. Uncommonly the myelinolysis could involve areas beyond the pons resulting in extrapontine myelinolysis (EPM). We report a 70-year-old gentleman who presented with a 2-week history of reduced interaction and altered behavior with parkinsonism following oral correction of severe hyponatraemia. Magnetic resonance imaging (MRI) of the brain revealed hyperintensities in the T2-weighted FLAIR images involving the pontine region with no involvement of basal ganglia. This case illustrates the significance of adhering to recommended protocols when correcting hyponatraemia and the importance of timing brain imaging in ODS.

Keywords: Osmotic demyelination syndrome, parkinsonism, extrapontine myelinolysis, atypical parkinsonism, hyponatraemia

INTRODUCTION

Osmotic demyelination syndrome (ODS) was first described as central pontine myelinolysis (CPM) in 1959 by Adams RD et al. among malnourished, alcoholic patients who developed quadriplegia with retained tendon reflexes and Babinski sign[1]. This condition was later attributed to rapid correction of hyponatraemia as well as many other conditions causing sudden changes in tonicity[2][3]. Extrapontine myelinolysis (EPM) is well described in literature and Tomita I et al. presented a case of parkinsonism following rapid correction of hyponatraemia[4]. We present a case of a gentleman presenting with parkinsonism following oral correction of hyponatraemia with MRI evidence of CPM.

CASE PRESENTATION

A 70-year-old gentleman with diabetes mellitus presented to us with a two-week history of progressive altered behaviour. They noticed reduced speech and marked slowing of movements. On examination he had bradykinesia with postural instability. However, he did not have a resting tremor. Cogwheel rigidity could be elicited bilaterally and the gait was shuffling in character. He had mild cognitive impairment with short term memory loss. He did not have gaze palsies, nystagmus, pyramidal signs or urinary incontinence. He was able to communicate, with a limited vocabulary.

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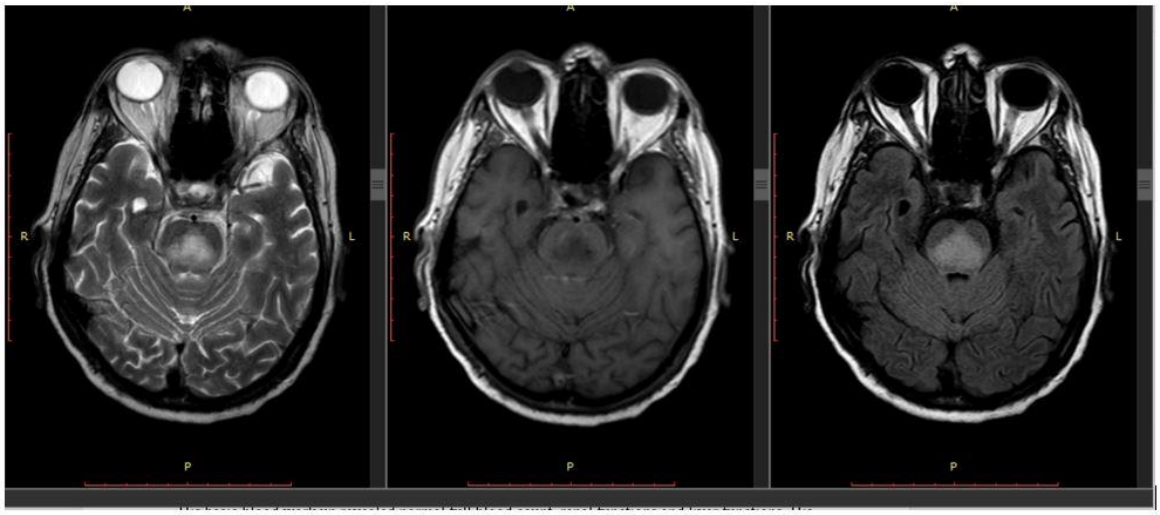
3 weeks back he had been admitted with vomiting and confusion and his serum sodium level was found to be low i.e. 110 mOsm/l. Medical records indicated that he was on hydrochlorothiazide and furosemide during that admission. Fluid restriction and oral correction with salt had been carried out. On discharge from that admission he had had a serum sodium level of 119 mOsm/l.

When he presented to us 3 weeks after being discharged from the previous admission, his serum sodium level was 138 mOsm/l. His basic blood work up revealed normal full blood count, renal functions and liver functions with a normal serum thyroid-stimulating hormone (TSH) level. Diuretics which were withheld during the last admission were not re-introduced. He was found to have poor glycaemic control as a subsequent fasting blood sugar value came as 278 mg/dl.

EEG did not reveal an encephalopathy pattern. T2-weighted FLAIR images of MRI brain revealed hyperintensities involving the pontine region. The classic "trident sign" was evident on the MRI brain as shown. T1-weighted images revealed hypointensities with minimal gadolinium enhancement involving the same region. These changes did not show diffusion restriction on diffusion weighted imaging (DWI). However interestingly, MRI did not reveal significant changes in the basal ganglia even though patient clinically showed extrapyramidal signs and symptoms. (Figure 1)



T2 T1 T2 FLAIR

Figure 1: MRI brain with T2-weighted FLAIR image showing pontine hyperintensities

He was commenced on levodopa/carbidopa combination to relieve his parkinsonism and physiotherapy was arranged as well. Over a period of 4 to 5 days he was able to move around independently and showed clinical improvement. Even though steroids and intravenous immunoglobulin has a place in the management of ODS, they were not commenced in this patient as he presented to us late in the course of the disease[5][6]. He is awaiting follow up MRI brain to assess progression and clinical assessment of motor symptoms and signs.

DISCUSSION

CPM and EPM are old terms which have been replaced by the term ODS. In 1959 Adams RD et al. described demyelination of the pontine region among malnourished, alcoholic patients who developed quadriplegia with preservation of eye movements[1]. Later EPM was described, where the demyelination was not confined to the pons. Patients presented with a variety of clinical manifestations ranging from movement disorders, cranial neuropathies, behavioural disorders to

psychiatric disorders, seizures and so on. The demyelination was observed in the external capsule, basal ganglia, thalamus, cerebral cortex and hippocampi[7].

The osmotic disequilibrium resulting in demyelination of extrapontine structures such as basal ganglia result in parkinsonism as a manifestation of ODS. Tomita I et al. described this observation following rapid correction of hyponatraemia[4]. Rizvi I et al. also presented a case where rapid rise in serum sodium levels had resulted in parkinsonism and MRI revealing evidence of EPM[8]. In literature there are cases where even correction of hyponatraemia done acoording to the recommended protocols had resulted in osmotic demyelination.

Brain imaging using MRI is the most reliable way of diagnosing ODS. However, MRI changes can get delayed compared to the clinical presentation. Myelinolytic lesions cannot be demonstrated within the first 2 weeks by using conventional MRI sequences[9]. Therefore, it is important not to exclude the diagnosis based on normal brain imaging, particularly if done early in the course of the disease.

The classic MRI finding of osmotic demyelination is the "trident" shaped T2 and FLAIR hyperintensity in the pontine region, which symmetrical[10]. There is associated T1 hypointensity with no contrast enhancement or mass effect. There can be diffusion restriction in DWI, even if imaged within first 24 hours[11].

In our patient, MRI brain showed T2 high signal intensities involving the pontine region bilaterally with the basal ganglia being spared, quite similar to the classic "trident" sign (Figure 1). However, the clinical presentation was more in favour of an EPM involving the basal ganglia. It could be concluded that the changes of the basal ganglia maybe too early to be seen on MRI. Therefore, it is crucial to time the brain imaging to get a more accurate result.

It is important to have an understanding regarding the categories of patients who are more prone to develop ODS. Over the years researchers have observed various groups of people who are at a higher risk of developing osmotic demyelination. This was described in the very first publication by Adams et al. in 1959 where he observed this phenomenon among "malnourished" and "alcoholic" patients[1]. Apart from rate of correction of hyponatraemia, other identified risk factors include hepatic dysfunction[12], post liver transplantation[13], Wilson disease[14], hyperosmolar states, hypokalaemia, hypophosphataemia, renal failure, hyperemesis and diabetes mellitus[7]. This patient being diagnosed with diabetes mellitus complicated with subnephrotic range proteinuria, might have played a role in pushing him towards osmotic demyelination with the correction of hyponatraemia.

This case reiterates the importance of the proper rate of correction of hyponatraemia to prevent osmotic demyelination, the importance of timing brain imaging for an accurate diagnosis and recognizing the high risk categories for the development of ODS.

Author declaration

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Author contributions

NTW under the supervision of PJPP and SA managed the patient referred to, in the case report. NTW drafted the manuscript and SA revised the final version.

Conflicts of interest

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Consent for participation

Informed written was obtained from the patient for the publication of case details.

Availability of data

Not applicable.

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