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# Extrapontine myelinolysis manifested selectively by acute severe parkinsonian syndrome

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**Abstract**

**OBJECTIVES:** Osmotic demyelination syndrome (ODMS) is a rare and serious neurologic disorder with acute myelin disintegration, usually in the pontine area (central pontine myelinolysis) and to a lesser extent, even in other areas of the central nervous system (extrapontine myelinolysis). The main underlying mechanism is the change of serum osmolality with quick correction of low mineral levels, mainly hyponatraemia. Clinical manifestation is various and depends on the localization.

**DESIGN:** We describe an acute isolated extrapontine myelinolysis causing acute onset of parkinsonism in a 61-year-old man who developed quickly progressing parkinsonian syndrome after the rapid correction of hyponatraemia.

**RESULTS:** Brain MRI revealed lesions only in the striatum, sparing the globus pallidus. Substitution therapy with high doses of levodopa significantly improved his clinical condition.

**CONCLUSION:** Extrapontine myelinolysis with isolated affection of basal ganglia is extremely rare. In such case, clinical manifestation of acute severe parkinsonism could be successfully treated by high dose of levodopa.

CASE REPORT

## INTRODUCTION

Although the most common cause of parkinsonism is Parkinson´s disease, a wide range of medical conditions may be associated with parkinsonian signs. In general, presentations of parkinsonism with rapid onset of gait disorder with postural instability suggest secondary causes of parkinsonism. Rarely, extrapontine myelinolysis (EPM) may be associated with acute parkinsonism (Post *et al.* 2009; Kwon *et al.* 2011).

Osmotic demyelination syndrome (ODMS) is a rare neurologic disorder with unknown pathogenesis. It is characterized by osmotic destruction of myelin and damage of oligodendrocytes in pons or extrapontine localisations while neurons and axons are spared (Martin, 2004). This syndrome is often related to quick correction of hyponatraemia (Adams *et al.* 1959). The clinical manifestation depends on the lesion localization.

In this case report, we present a 61-year-old patient with acute development of parkinsonian

symptoms, following a rapid correction of symptomatic hyponatraemia that had developed after profuse vomiting. It is a case of rare isolated extrapontine myelinolysis, where early levodopa substitution therapy significantly improved the serious clinical condition.

## CASE REPORT

A 61-year-old man with a past medical history of hypertension and hypercholesterolemia was transported to the hospital because a collapse with fall downstairs and with vertigo. His condition was preceded by 4 days of nausea with repeated profuse vomiting (up to 20 times daily). He presented slightly imprecise dystaxia in the left upper extremity. Slight instability of stance was apparent, but the patient was able to walk without help. He had a history of chronic alcohol abuse 20 years ago.

Acute brain MRI showed only lesions of gliosis in the white matter, probably of vascular-ischemic aetiology (Figure 1). On the day of acute admission his serum sodium concentrate was 104 mmol/l, potassium 2.29 mmol/l, chlorides 66 mmol/l, serum osmolality 229 mmol/kg. After the first 12 hours of intravenous substitution, the sodium level increased to 117 mmol/l (by 13 mmol/l). In the next 12 hours, it increased to 124 mmol/l. Total natraemia correction was 20 mmol/l in 24 hours. In the following days, the serum mineral levels were stable and the patient did not require further supplementation.

Five days after the discharge from the hospital, the patient gradually developed clinical manifestations of hypersalivation, speech worsening, body stiffness and serious deteriorating mobility. In the objective neurologic finding, severe bilateral parkinsonian syndrome was dominant, with intermittent resting tremor of extremities and significant rigidity, serious gait disorder, general restlessness and anxiety. Levodopa/carbidopa treatment was initiated with gradual titration. The control MRI of the brain (17 days after the initial MRI) showed bilateral diffuse increase of signal intensity in the level of basal ganglia (the striatum, sparing the globus pallidus), especially in the DWI (Figure 2) supporting the diagnosis of extrapontine myelinolysis.

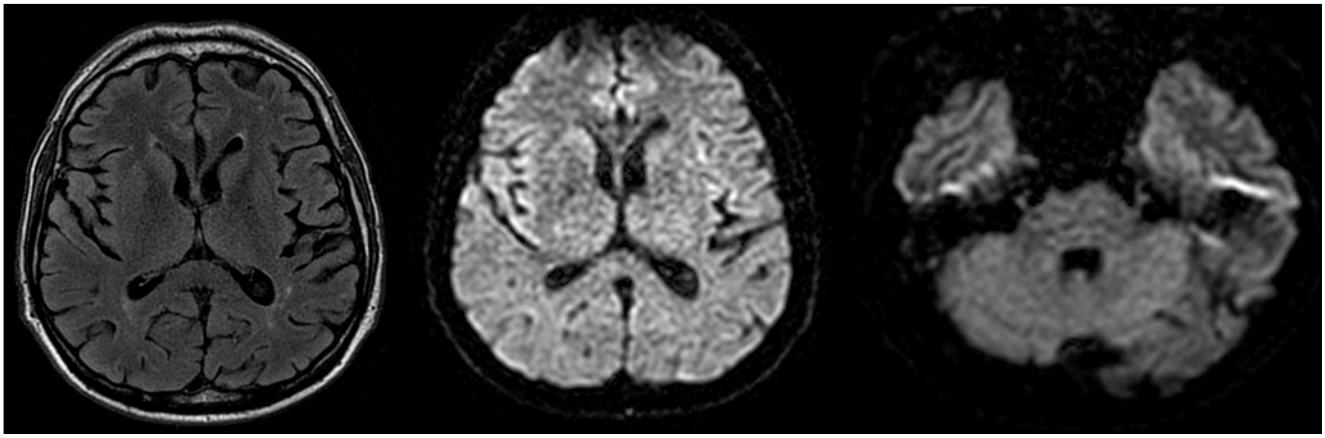

**Fig. 1.** Initial brain MRI. T2 weighted scans with lesions of gliosis in the white matter, probably of vascular-ischemic aetiology (Right), DWI scans without pathology in the basal ganglia (Middle) and pons (Left).

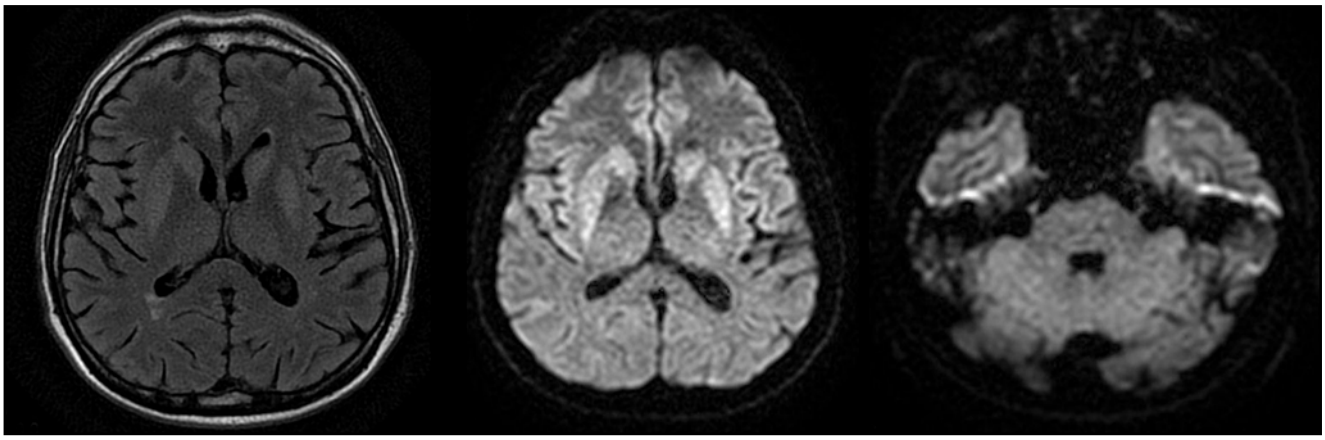

**Fig. 2.** Brain MRI after 17 days. T2 weighted scans with symmetrical hyperintensities involving basal ganglia (Right), DWI scans with bilateral diffuse increase of signal intensity in the same region with relative sparing of globus pallidus (Middle), DWI scans with no increase of signal intensity in pons (Left).

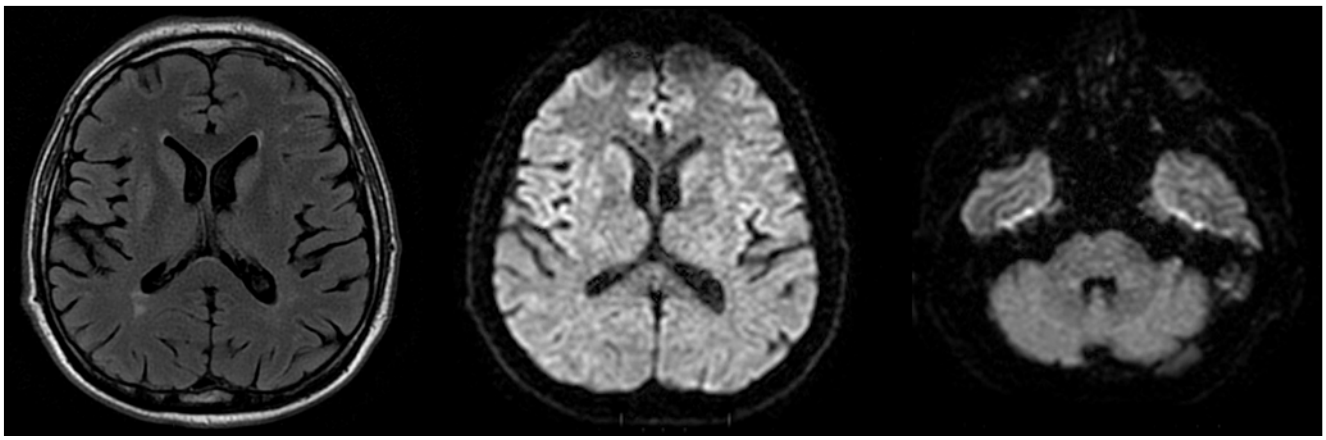

**Fig. 3.** Brain MRI after 2 months. T2 weighted scans with less hyperintensities involving basal ganglia especially on the left side (Right), DWI scans without pathology in the basal ganglia (Middle) and pons (Left).

After ten days the patient significantly improved his clinical condition with 1000 mg/100 mg of levodopa/carbidopa daily.

A follow-up visit after 2 months of the onset of EPM his speech improved; resting tremor disappeared, hypersalivation receded and mobility and gait were without problems. MRI findings were improved (Figure 3). The patient was able to walk quickly for about 3 km. At a follow-up 6 months after the hospital discharge, the patient stated even further improvement of the condition. He was able to walk for 10 km; he swam twice a week for 1 hour in a pool. A follow-up visit after one year he had only slight residual rigidity with mild masked-like face. Beta-CIT SPECT imaging of the dopamine transporter was normal. Levodopa/carbidopa dose was decreased; currently, it is set to 300 mg/30 mg/day and clinical status remained fully compensated.

## DISCUSSION

The most common causes of ODMS are hypoosmolal conditions, mainly hyponatraemia with rapid correction. In asymptomatic chronic hyponatraemia, there is a risk of ODMS even in slow correction (Adams *et al.* 1959). ODMS has usually a biphasic course. The first phase includes encephalopathy with serum level of hyponatraemia. These signs will disappear with natraemia correction. The second phase begins 2-7 days after natraemia correction. The symptoms are various and depend on the affected brain areas (Martin, 2004). In EPM, parkinsonian symptoms may appear (Seah *et al.* 2002); however, EPM without central pontine myelinolysis is extremely rare (Iman *et al.* 2012). In our patient, the natraemia correction values were above the upper border of the recommended range. After the first phase of ODMS he developed severe acute isolated parkinsonian syndrome with prompt improvement after starting high dose of dopaminergic treatment (Iman *et al.* 2012). No specific ODMS treatment is usually available. Only symptomatic and supportive therapy is recommended (Martin 2004). Appropriate hyponatraemia substitution is necessary as ODMS prevention.

The main diagnostic method in ODMS is magnetic resonance imaging (Chua *et al.* 2002). Abnormal MRI finding showing hyperintensive lesions on T2-weighted images is apparent usually within 10-14 days after clinical symptoms, but the extent of affection may not correlate with the extent of symptoms. For early diagnostics, diffusion weighted images (DWI-MRI) are very important (Chu *et al.* 2001). In our patient, restrictive diffusion on DWI sequence has been demonstrated in basal ganglia areas with palladial sparing and with normal findings at the pontine level.

In this report we present a rare case of isolated extrapontine myelinolysis with acute development of severe parkinsonian symptoms with a very good clinical response to levodopa substitution. We emphasize the necessity of slow correction of ion imbalance and point out to unfavorable factors like a history of alcohol abuse (Hagiwara *et al.* 2008). We underline the need for urgent monitoring including repeated MRI scans with high importance on DWI scans. Levodopa is recommended as a highly effective symptomatic therapy.

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