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Manganese Encephalopathy in Setting of a Congenital Extrahepatic Portosystemic Shunt as Cause of Secondary Parkinsonism

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Abstract

Manganese has been shown to deposit in the basal ganglia. While its toxicity has a low incidence, it can result in Parkinsonian syndrome. We describe a case of a 43-year-old man, with medical history of congenital portosystemic shunt, who presented with parkinsonism in the setting of manganese encephalopathy. CTA of abdomen and pelvis showed findings of pelvic portosystemic shunting. Brain MRI with and without contrast showed T1 hyperintensity within the bilateral basal ganglia and anterior mid-brain. Patient was started on developed levodopa - carbidopa with mild improvement of motor symptoms but it was stopped due to the development of psychosis. This report emphasizes the importance of early recognition of congenital portosystemic shunt as an etiology of secondary Parkinsonism as early closure of it can be curative.

Keywords: Manganes; Parkinsonism; Levodopa Carbidopa; Portosystemic shunt

Introduction

Manganese (Mn) is an essential trace element necessary for physiological processes that support development, growth and neuronal function [1]. Manganese exposure has long been known to cause neurotoxicity. Mn has been shown to deposit in the basal ganglia, most commonly in the medial segment of the globus pallidus, and this can be detected on MRI of the brain as T1 hyperintensities [2]. While its toxicity has a low incidence, it can result in secondary Parkinsonism manifested by bradykinesia, rigidity, tremors, and early gait abnormalities [3-4]. Unlike other causes of parkinsonism, manganese neurotoxicity classically has a poor response to treatment with levodopa [5].

Normally, manganese is hepatically cleared and renally excreted however in patients with cirrhosis, it is not cleared by the liver and can be deposited as previously described [6]. This deposition can also occur in patients with congenital and surgical portosystemic shunts, as blood that has bypassed the liver is not cleared of manganese [7]. While case studies have been published on these depositions in children with congenital portosystemic shunts that were subsequently surgically treated with shunt closure, few studies have observed adult patients whose shunts were not closed [7].

Case Presentation

The patient is a 43-year-old man, with past medical history significant for congenital extrahepatic portosystemic shunt type II and multiple hospitalizations for hepatic encephalopathy, who was admitted to the gastroenterology department for suspected decompensated hepatic encephalopathy due to medication noncompliance.

Neurology evaluated the patient, as encephalopathy did not respond to lactulose. Upon physical examination, he was found to have bradyphrenia, hypophonic slow speech, hypomimia, reduced blinks, gait and postural instability, and significant bradykinesia for hand grips, finger taps, toe taps in upper and lower extremities.

The workup was significant for total bilirubin level of 2.1 and ammonia of 83. Reversible etiologies of encephalopathy, such as B1, B12, or zinc deficiency, hypothyroidism, and syphilis, were ruled out. Computed tomography angiography (CTA) of abdomen and pelvis showed distended mesenteric veins and dilated mesenteric veins extend down into the pelvis with markedly dilated vessels within the pelvis especially about the rectum (Figure 1). Brain MRI showed T1 hyperintensity within the bilateral basal ganglia and anterior midbrain (Figure 2). Wilson disease was ruled out in previous hospitalizations. Due to a history of congenital extrahepatic portosystemic shunt (CEPS), we favored the diagnosis of manganese encephalopathy induced secondary parkinsonism, even though serum manganese levels were found to be normal at 2ug/L.

The patient was started on a half-tablet of carbidopa-levodopa 100/25 TID for severe Parkinsonism. Interventional Radiology was consulted for shunt embolization, however, the procedure was deemed impossible due to the high number of shunts. One day after starting carbidopa-levodopa, the patient developed acute psychosis, requiring antipsychotics and restraints. Carbidopa-levodopa was discontinued, and psychosis resolved with improvement in gait instability and mild improvement in cognitive function and bradykinesia. The patient was discharged home and declined facility placement.

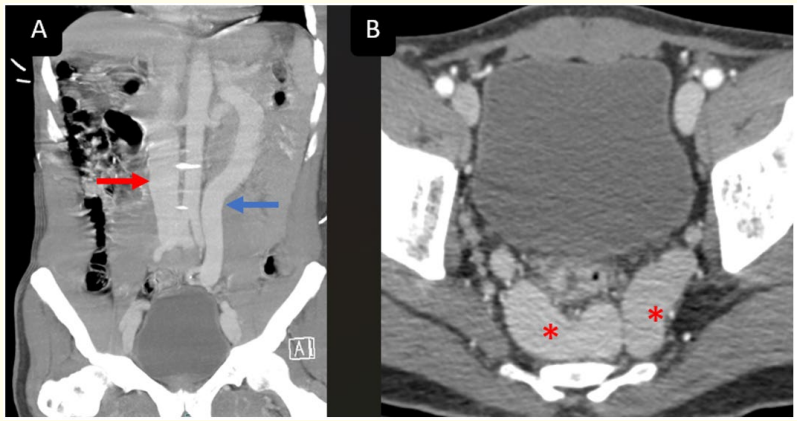

Figure 1: A: Computed tomography angiography (CTA) of abdomen and pelvis showing distended superior mesenteric vein (red arrow), and inferior mesenteric vein (blue arrow). B: Dilated mesenteric veins extending down into the pelvis with markedly dilated vessels especially about the rectum (red asterisks).

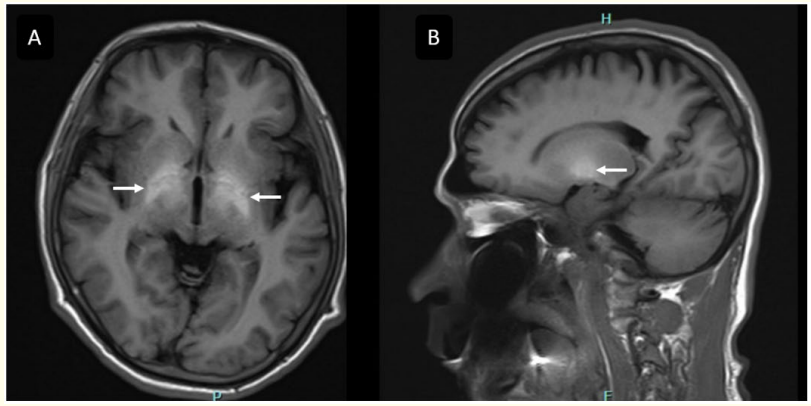

Figure 2: A: Brain MRI T1-WI axial plane showing increased signal intensity in cerebral bilateral globus pallidus (white arrows). B: Brain MRI T1-WI sagittal plane showing moderate signal intensity increase in globus pallidus (white arrow).

Discussion

CEPS is a rare condition where the portomesenteric blood drains into a systemic vein, bypassing the liver through a complete or partial shunt. Shunts commonly present with encephalopathy due to decreased hepatic clearance, leading to accumulation of ammonia and other toxins [8]. Similar presentation can be seen following a transjugular intrahepatic portosystemic shunt (TIPS).

Manganese is among the toxins that can contribute to encephalopathy. Excess Mn can precipitate nervous system toxicity, such as Parkinsonism [9]. Although our patient had normal Mn level, his clinical presentation is consistent with manganese toxicity. The diagnosis can often be made by clinical and radiological correlation with MRI showing increased T1 intensity in globus pallidus [10]. Manganese-induced Parkinsonism differs from idiopathic by bilaterality of symptoms, earlier onset, and poor response to conventional regimens, such as carbidopa/levodopa (C/L) [9-10].

Hepatic encephalopathy can frequently be managed with shunt embolization, which eliminates toxin deposition [11-12], however, our patient had extensive shunts that could not be embolized leaving us with medical therapy as the only option to treat his parkinsonism. Even though carbidopa /levodopa (C/L) is the gold standard therapy in idiopathic Parkinson Disease [3], previous cases of Parkinsonism precipitated by manganese encephalopathy did not have a proper improvement with C/L therapy [10]. Our case is unique in that the patient improved with a small dose of C/L although it initially precipitated an episode of psychosis.

Our case is an exceedingly rare example of Parkinsonism due to CEPS-induced manganese toxicity documented in an adult that partially responded to treatment with C/L. It provides evidence that C/L could be helpful in mitigating the disease course in patients who are unable to undergo shunt embolization.

Conclusion

Secondary parkinsonism due to CEPS is a rare cause of Parkinsonism that typically only responds to shunt closure. This case indicates a possible alternative method of Parkinsonism reversal in patients who cannot undergo embolization.

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