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YOUNG-ONSET DEMENTIA AND MRI CHANGES IN A PATIENT WITH SUBCLINICAL LIVER CIRRHOSIS DUE TO CHRONIC HEPATITIS C

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Abstract

Young-onset dementia (before age of 65) is relatively infrequent and presents a challenge in everyday neurological practice due to wide spectrum of clinical presentations and diversity of underlying etiology. When cognitive deficits are accompanied with liver dysfunction different etiologies should be considered. We present a case report of a young patient with subclinical decompensated liver disease due to underlying chronic hepatitis C, presented with the mildest form of hepatic encephalopathy spectrum, called minimal (subclinical) hepatic encephalopathy and characteristic MRI changes.

Keywords

• MRI • Cognition • Dementia • Hepatitis C

Young-onset dementia (before age of 65) is relatively infrequent and presents a challenge in everyday neurological practice due to wide spectrum of clinical presentations and diversity of underlying etiology. Since many of causes in this age group are treatable, their exclusion is mandatory. When cognitive deficits are accompanied with liver dysfunction different etiologies should be considered, such as alcohol toxicity, tumors, infections, Wilson disease and some rare hereditary diseases (e.g. hereditary hemochromatosis, mitochondrial diseases, porphyria) [1].

A 57-year-old right-handed highly educated man was referred to a dementia outpatient clinic due to progression of concentration and memory problems and slight changes in personality that had started insidiously a year earlier. His past medical history was unremarkable. There was no history of excessive alcohol intake. At the exam, his Mini Mental State Examination (MMSE) score was 18/30, he was disoriented in time and space with severely decreased verbal fluency (Benton verbal fluency test score 14), moderate attention (digit span, forward = 5, digit span, backward = 2) and episodic memory problems (immediate recall on Rey auditory-verbal learning test was 15/75), constructional apraxia, psychomotor slowing, difficulties in judgment and visual perception. Except for the moderately increased liver function tests (AST 128 U/L, ALT 138 U/L, GGT 143 U/L, total bilirubin 46 μmol/L), decreased thrombocyte count 67 x10^9/L and hypergammaglobulinemia (29.5 g/L), other tests were unremarkable (standard laboratory tests, thyroid hormone, vitamin B12, folic acid, serum and 24-hours urine copper, ceruloplasmin, Keiser-Fleisher ring, serum iron, serum and CSF antibodies to syphilis and *Borrelia burgdorferi*, tumor markers and paraneoplastic antibodies (anti-Hu, anti-Ri and anti-Yo). On T1-weighted MRI images bilateral hyperintensities in globus pallidum (Figure 1a and 1b) and substantia nigra (Figure 2) were revealed. Hepatitis B serology was negative; however hepatitis C antibodies (anti-HCV and anti-HCV RIBA) and serum ammonia levels (152.3 μmol/l, normal range up to 48.2 μmol/l) were markedly increased. Finally, enlarged liver and spleen with supracardial varices of third stage were shown on abdominal CT and oesophagogastroduodenoscopy. Patient was started with 20 mg of propranolol bid as prevention of varical bleeding, 10 mg of lactulosis bid and protein restriction diet. Interferon α was not introduced. On the last follow-up visit 7 months after patient's MMSE was 25/30.

This is the case of a young-onset dementia caused by subclinical decompensated liver disease with underlying chronic hepatitis C infection. Decompensated liver disease may by increasing blood ammonia levels cause both structural and functional brain abnormalities leading to hepatic encephalopathy of variable rate [2]. The mildest form of hepatic encephalopathy spectrum, called minimal (subclinical) hepatic encephalopathy, is characterized with subtle neurocognitive impairment that primarily affects information processing, attention and psychomotor skills and may lead to overt hepatic encephalopathy [3]. Patients with chronic hepatitis C infection, irrespective of the presence of cirrhosis, may show similar cognitive deficits. Although subclinical hepatic encephalopathy is frequent in patients with liver chirrosis, it is still largely underdiagnosed [4]. Hepatitis C infection is not uncommon in Croatian general population. The estimated prevalence of anti-HCV infection marker is 1.38% with around 200 newly infected cases reported every year to the Croatian Institute for Public Health [5]. Bilateral hyperintensities in basal ganglia (primarily globus palidus) and substantia nigra may be found on T1-weighted MRI images in patients with cirrhosis due to hepatitis C infection

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irrespective of their cognitive state [6]. Since ammonia plays a key role in the pathogenesis of encephalopathic presentation, ammonia-lowering agents may result in cognitive improvement. Interestingly, interferon therapy did not lead to any change in cognition in patients with chronic hepatitis C [7]. According to the American Academy of Neurology practice guidelines, blood ammonia testing is not a routine diagnostic test in patients with dementia [8], although it is generally used in diagnostic work-up of patients in dementia specialized centers. Since liver function tests might be unremarkable in around 30% of patients with hepatitis C infection despite inflammation on liver biopsy, blood ammonia is sometimes the only finding of decompensated liver disease and should be included in a diagnostic work-up of patients with cognitive problems [9].

Conflict of interest statement:

There is no conflict of interest or financial conflicts of interest.

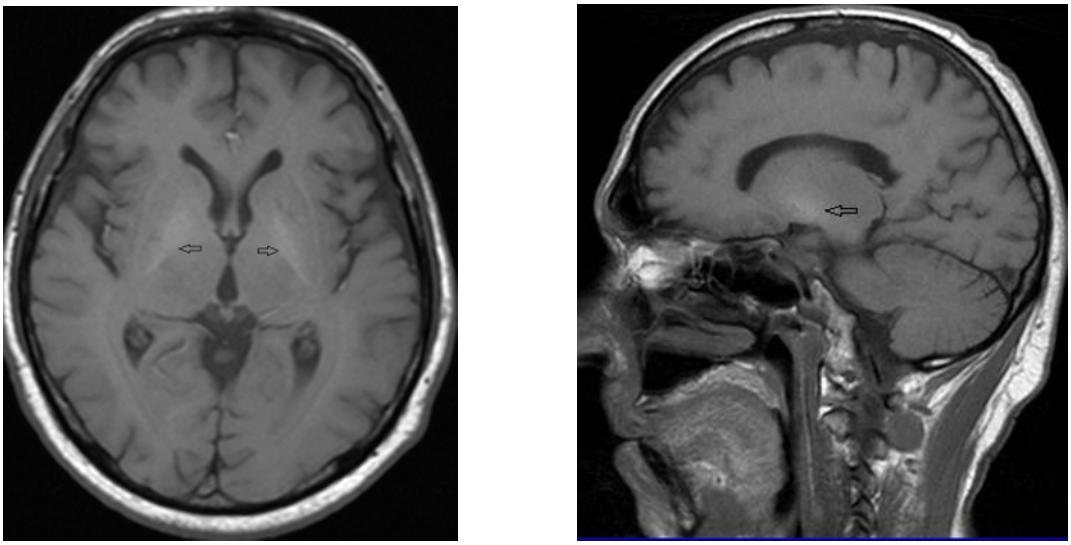

Figure 1. T1-weighted MRI image of brain (transversal section) showing bilateral hyperintensities in globus pallidus (arrows). 1b. T1-weighted MRI image of brain (sagittal section) showing hyperintensity in globus pallidus (arrow).

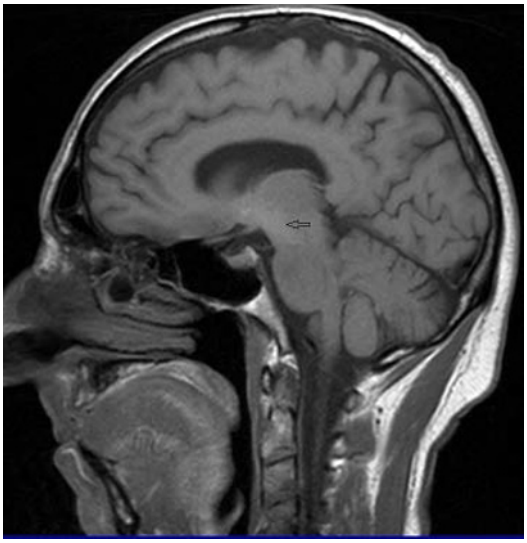

Figure 2. T1-weighted MRI image of brain (sagittal section) showing hyperintensity in substantia nigra (arrow).

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