

Short Communication

Huntington's Disease Complicated by Traumatic Subarachnoid Bleeding and Subdural Hematoma

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Abstract

Though Huntington's Disease (HD) is frequently complicated by falls and consecutive Traumatic Brain Injury (TBI) with Subdural Hematoma (SDH), traumatic Subarachnoid Bleeding (SAB) has not been reported as a complication of HD. A 67yo female with HD due to a CAG-repeat expansion of 43 repeats, diagnosed 18 months earlier, was admitted after a fall with a Glasgow-Coma Scale (GCS) score of 3. Cerebral CT on admission revealed bilateral SDH and SAB. No aneurysm could be detected on conventional angiography. After extubation, she was non-responsive to verbal requests and typical choreatic movement recurred. Tiaprid, olanzapine, and tetrabenazine were of limited effect. This case shows that falls in HD may not only cause SDH but rarely traumatic SAB. TBI in HD may worsen the phenotype and may increase the risk of a poor outcome.

Keywords: Traumatic brain injury; Huntington's disease; Dementia; Movement disorder; Huntingtin; Ataxia; Chorea

Short Communication

Huntington's disease (chorea Huntington, HD) is an autosomal dominant, exonic trinucleotide disorder caused by a protracted CAG-repeat >35 in the Huntingtin gene (HTT) on chromosome-4.1 HD becomes clinically manifest between ages 30 and 60y if there are 35-60 repeats and between ages 13 and 20y if there are >60 repeats [1]. There is anticipation in case of paternal transmission. The prevalence ranges between 4 and 10/10000 [1]. HD is clinically characterized by progressive choreatic hyperkinesias, cognitive decline, dysarthrophonia, dysphagia, falls, and immobility. Subdural Hematoma (SDH) is well recognised as a complication of HD [2-5] but traumatic Subarachnoid Bleeding (SAB) has not been reported as a complication of HD. The legal patient's representative consented with the publication in anonymous form.

The patient is a 67yo female, height 165cm, weight 44kg with a previous history osteoporosis and smoking who developed progressive memory deficits and cognitive impairment together with recurrent falls starting at age 65y. Since age 66y she developed slowly progressive choreatic hyperkinesias of all four limbs. At age 66y she was admitted for exertional dyspnoea and fever. Clinical exam at age 66y revealed short stature, facial dyskinesias, bilateral ataxia, bilateral dysmetria, ataxic stance with a tendency to fall, and choreatic movements on the upper limbs. Blood tests revealed iron deficiency and folic acid deficiency. Densitometry revealed osteoporosis. MRI showed susceptibility artefacts within the caudate nucleus bilaterally. Abdominal ultrasound revealed a liver cyst. Genetic work-up revealed a CAG-repeat expansion of 43 repeats in the HTT gene why HD was diagnosed. Tiaprid (300mg/d), olanzapine (5mg/d), and tetrabenazine (75mg/d) were given with limited effect. At age 67y she was admitted after a fall in her flat with consecutive coma requiring intubation and artificial ventilation. On cerebral CT scan bilateral acute and chronic SDH and SAB were diagnosed. CTA did not reveal an aneurysm.

After extubation and discontinuation of sedatives on hospital day she did not follow instructions, but presented with orofacial dyskinesias, choreatic limb hyperkinesias, and positive pyramidal signs.

Traumatic Brain Injury (TBI) is a comprehensible complication of HD, given the choreatic movements and the progressive cognitive decline destabilizing HD patients and thus giving rise to recurrent falls. In a study of 192 HD patients from Finland, seven patients with chronic Subdural Hematoma (SDH) were identified [5]. The incidence of chronic SDH was thus 63.4/100000/a and thus higher than in the general populations [5]. In a retrospective study of 58 patients with chronic SDH, four (6.9%) had HD [6]. There are also reports showing that chronic SDH precipitates chorea [2,4], why it is conceivable that chorea in HD may be increased by TBI. There are even indications that the first occurrence of chorea in HD can be triggered by chronic SDH [3]. The reason why the index patient experienced traumatic SAB and SDH remains elusive as it occurred unwitnessed but it can be speculated that another fall resulted in a TBI with rupture of a subarachnoid artery. The bilateral SDH presented with acute and chronic portions.

Overall, this case shows that falls in HD may not only cause SDH but rarely traumatic SAB. TBI in HD may worsen the phenotype and may increase the risk of a poor outcome.

Author's Contribution

JF: Design, literature search, discussion, first draft, critical comments, MK: Literature search, discussion, critical comments. Informed consent: verbal informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

Ethical Approval

The study was approved by the local institutional review board in the Hospital.

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