Thunderclap Headache: A Primary Symptom of a Steroid-Responsive Encephalopathy associated with Autoimmune Thyroiditis.

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Objective: Thunderclap headache is frequently associated with intracranial vascular disorders and is a frequent cause for emergency department admission. A randomized population-based prospective study estimated an annual incidence of Thunderclap headache of 43 cases per 100,000 adults. In contrast to thunderclap headache, steroid-responsive encephalopathy with autoimmune thyroiditis (SREAT) is a rare disease, affecting only approximately 2.1 in 100.000 people. The clinical presentation of patients with SREAT is highly variable, encompassing a broad spectrum of complaints and symptoms. A correlation of thunderclap headache as a primary symptom with autoimmune disorders, such as SREAT, is highly unusual.

Method: A 79-year-old female presented with a sudden onset of severe bifrontal headache. There were no preceding trauma and no history of migraine or other intermittent headaches. The initial neurological examination was normal. Brain CT-Scan showed minor microangiopathic and major macroangiopathic changes and strio-pallido-dentate calcifications but no other pathologies. Standard laboratory results revealed no abnormalities. Metamizole (2 g/d) was administered, which led to significant improvement and discharge. Within 24 hours of being discharged, the patient experienced difficulty in speaking. Within hours, the patient became delirious with partially aggressive behavior accompanied by optic hallucinations. Brain MRI showed minor microangiopathic changes along with small left temporoparietal postischemic defects. Under the suspicion of an infectious meningoencephalitis, broad-spectrum antimicrobial and antiviral therapy with ceftriaxone and acyclovir was initiated for a period of seven days, under which the patient's clinical state remained to a large extent unchanged. CSF analysis revealed moderate lymphocytic pleocytosis without evidence of infectious, paraneoplastic or metabolic causes. A follow-up brain MRI on the 10th day presented progressive leptomeningeal and sulcal T2/FLAIR hyperintensities parieto-occipital on both sides (Fig. 1) and temporoparietal on the left side along with an increasing unspecific subcortical and periventricular T2-FLAIR hyperintensities (Fig. 2).

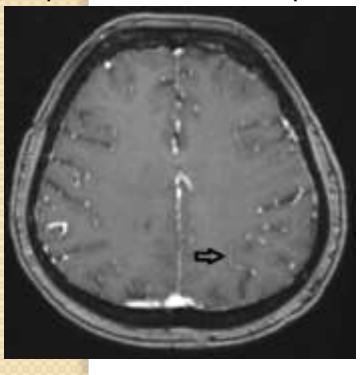

Fig. 1: MRI from June 2012 showing hyperperfusion of the leptomeningeal arteries (black arrow).

Result: Careful re-evaluation of the medical history revealed a preceding episode of aseptic meningoencephalitis which responded to steroids seven years before with comparable clinical findings. Although the patient had a known history of hypothyroidism, autoimmunological laboratory diagnostic regarding thyroid antibodies was never conducted. Here, the anti-TPO antibodies were markedly increased with normal values of TSH, T4, and the T3. Together with the history of steroid-responsive aseptic meningoencephalitis in 2012, diagnosis of SREAT was highly probable, and a high-dose immunosuppressive therapy with intravenous methylprednisolone was commenced. Under this treatment regime, neuropsychiatric symptoms showed clear and fast improvements. Visual and auditory hallucinations resolved completely within days.

Conclusion: SREAT is a rare but very serious illness. Importantly, no SREAT-specific clinical, laboratory, or radiological findings are existent, and the disease is often missed, underdiagnosed, or diagnosed with a substantial time delay. Clinically, both sudden onset of focal neurological symptoms and gradually increasing neurological complaints are described. Thunderclap headache in a patient without a history of migraine or other intermittent headaches being the primary symptom of a SREAT represents a peculiarity of our case. In SREAT, headache may occur in up to 80% of the patients, but it is normally mild or periodic, not a presenting sign, and is usually attributed to previous migraine and hypothyroidism.

To conclude, the present case underlines that awareness for SREAT is important when evaluating patients with acute onset of high-intensity headache associated with neuropsychiatric symptoms, particularly because most patients respond very well to steroids.

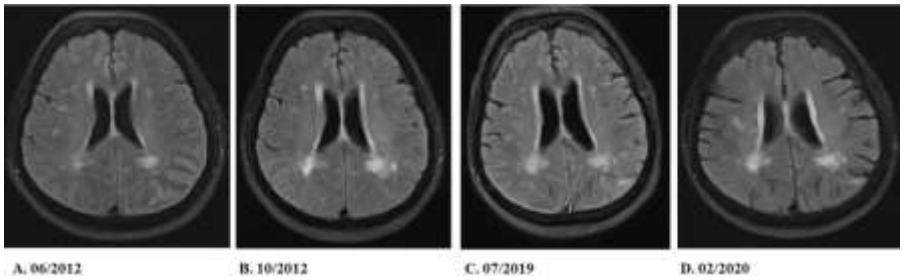

Fig. 2: FLAIR images (a–d) showing progressive diffuse signal abnormality involving the white matter of both cerebral hemispheres over 7-year period (a–c) with gradual resolving after steroid treatment (d). (a) June 2012. (b) October 2012. (c) July 2019. (d) February 2020.