pyramidal syndrome characterized by incomplete hemiparesis on the right hemibody. Myotatic exalted reflexes in the right hemibody and plantar-cutaneous reflex were indifferent.

DISCUSSION

Reviewing the relevant literature worldwide, Labrune syndrome was considered as a possible diagnosis for the present case[2]. Despite the presence of cysts, calcification, and edema of the white substance found in our patient, suggesting neurocysticercosis, equinecocosis[3] or neoplasia, there was no serologic or histopathologic confirmation.

This syndrome is characterized by calcifications, leukodystrophy, and formation of parenchymal cysts[1]. Its onset can occur during childhood or adolescence, in an average of 12 years (7 months – 59 years), but there was not one in adults in Brazil, with neurological signs such as cognitive decline, seizures and pyramidal, extrapyramidal or cerebellar signs[1,2]. Our patient, unlike the other cases reported in literature, presented its first neurological manifestation at the age of 24. CT and MRI seen in our case were similar to the cases reported in literature, showing increased signal intensity of the white matter on MRI (T2 and FLAIR), basal ganglia calcification, and development of cysts[1-3].

Labrune reported the results of histopathology with rearrangement involving the microvessels, whereas perivascular foci of calcifications, hyaline deposits, and formation of Rosenthal fibers seem to be compatible with this change[1]. The histopathological findings of our patient were consistent with LCC. According to them, the likely primary pathologic feature is a rearrangement involving the microvessels and the formation of Rosenthal fibers[1,4].

In conclusion, the etiology of LCC remains unknown. In spite of relatively characteristic findings in imaging and histopathological examination, there is no uniformity in the clinical findings noted in the published articles. It can be speculated that the later age of onset, normal intelligence and slow progression, like in our patient, may indicate the shape of this rare disease in adults.

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Metamorphopsia associated with topiramate for migraine prevention

Metamorfopsia associada ao uso de topiramato para prevenção de enxaqueca

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A 33-year-old woman (registration 70-247/ IPUB-UFRJ) has had recurring headaches since the age of 18. She described more right-sided than left-sided throbbing, which could be severe and associated with nausea, light and noise sensitivity, and vomiting. She has had a visual aura, seeing "zig-zags", lasting for 20 to 30 minutes before the headaches. The only preventive measure she had taken in the past was divalproex sodium, with good efficacy. Because of the weight gain as a side effect, she refused to take it again. Her body mass index (BMI) was 35.3. She was started on preventive therapy with topiramate at 25 mg daily for two weeks that was to be increased weekly

to 100 mg daily. After 13 days of treatment, she developed continuous visual illusions upon waking in the morning; when looking at human faces, they were distorted and swollen. On many occasions, objects in front of her appeared to be either nearer or farther away. These visual phenomena persisted for approximately 12 hours and gradually disappeared with the discontinuation of topiramate. Thereafter, she never had similar experiences. None of these events was accompanied by the loss of consciousness or headache. The patient's impressions of reality and self-recognition were preserved. The neurological and psychiatric examination was normal, and a complete examination by a neuro-ophthalmologist was normal. An EEG, with activation procedures (hyperventilation and photic stimulation), and the MRI of the brain were normal.

Metamorphopsia is a visual illusion affecting the perception of the size, shape or inclination of objects[1]. Although this condition occurs in migraine aura, topiramate has been reported to induce other visual illusions, such as palinopsia (the illusion of a persistent or recurrent visual images following the removal of the exciting stimulus)[2] and alterations in body perception ("Alice in Wonderland syndrome") in patients with migraines[3].

The mechanism by which topiramate may cause these visual illusions in migraineurs is unknown. However, because it may occur in the aura of migraines, these visual illusions are likely to be a result of the migraine.

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Behavioral changes on amyotrophic lateral sclerosis (ALS): a case of ALS/FTD TDP-43 proteinopathy

Deterioração comportamental na esclerose lateral amiotrófica (ELA): um caso de proteinopatia TDP-43 associada à ELA e demência frontotemporal

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The frontotemporal dementia (FTD) is the second most common form of dementia in patients younger than 65 years, and its behavioral variant (bvFTD) is the most prevalent form[1,2]. During the last years, the overlapping between FTD and amyotrophic lateral sclerosis (ALS) has been frequently recognized, with symptoms of FTD preceding ALS and vice-versa. Herein, we report a case of ALS, which afterwards presented psychotic and behavioral symptoms, whose neuropathological diagnosis was compatible with bvFTD-ALS with TAR DNA-binding protein 43 (TDP-43) inclusion[3].

CASE REPORT

A 58-year-old man was admitted in our Emergency Unit with a one-year history of progressive weakness of limbs, associated with dysarthria, dysphonia, difficulty to close mouth and hands atrophy. Four days before admission, he developed dyspnea and acute respiratory failure. In the hospital, it was seen generalized weakness, global hyperreflexia, fasciculations on right arm, no sensory abnormalities, and the electroneuromyography showed chronic and acute denervation signs in cranial, cervical