Atypical presentation of sporadic Creutzfeldt – Jacob disease

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Creutzfeld – Jacob disease (CJD) is a progressive, degenerative, and fatal disease of the central nervous system. It is caused by abnormal accumulation of prion proteins and is characterized mainly by progressive dementia, cerebellar, pyramidal, extrapyramidal findings and with myoclonus. Most commonly are psychiatric symptoms, which are often the first signs of the disease. The incidence of CJD is approximately 1 in 1,000,000.

We report a case of a 63-year old patient with acute paroxysmal and repeated vertiginous symptoms accompanied with vision difficulties. The symptoms started 2 months before hospital admission. In previous history the patient was conservatly treated due to lung carcinoma 10 years ago. Brain CT scan doesn't show any acute changes, but chronic cerebro-vascular changes suitable for age. Also, trans cranial Doppler examination of brain vessels and EEG was no specific.

After 2 months she develops a progressive neurological symptomatology. Repeated EEG and standard brain MR imaging were not typical for CJD, but the development of the clinical image, differential-diagnostic thinking and specific investigations of cerebrospinal fluid (CSF) , and additionally made specific MR neuroimaging of brain lead us to clear diagnosis.

Definitive diagnosis of disease is brain biopsy or post –mortem brain analyses but in many countries for many reasons (cultural, technical, familiar) are not welcomed and preformed, as with our country.

Therefore, we emphasize the importance of differential – diagnostic thinking when methods of investigations are unspecific, especially at atypical clinical presentation of sporadic type of disease, as with our case.