A case of progressive palsy with tiger-eye effect on MRI. A difficult diagnosis

Abstract:

The presented clinical case describes a rare occurrence of progressive palsy with a tiger-eye effect on MRI. The issues of the workup and differential diagnosis were discussed. There was an argument that along with other MRI criteria for the diagnosis of Progressive Supranuclear Palsy (PSP), the above-described symptom of the "tiger eye" may occur in some patients with this disease. This radiological sign serves as a pathological correlative, indicating the possibility of developing neurodegenerative disease according to a single universal mechanism of neuronal death in various parts of the brain that determine the specificity of clinical manifestations.

Keywords: progressive supra-nuclear palsy, diagnosis, MRI, neurodegenerative diseases

Introduction:

Richardson, Steele and Olszewski, in 1963, described a syndrome characterized by progressive Parkinsonism with frequent falls due to postural instability, supra-nuclear ophthalmoplegia, mainly in the vertical plane, pseudo-bulbar palsy, dystonic rigidity of the neck and upper arm muscles, and moderate cognitive deficiency in nine patients [1, 8, 11]. A further neuro-pathological post-mortem study demonstrated that in this condition, the basal ganglia, stem structures and the cerebellum are affected, with a predominance of pathological implants in the form of neurofibrillary tangles (NFTs), granules of degeneration, loss of neuronal cells and gliosis [7]. This syndrome is called progressive supra-nuclear palsy (PSP) and is regarded as the most common form of atypical Parkinsonism today [1, 11, 17]. The PSP frequency is 5-7 cases per 100,000 populations [8, 9, 11]. A recent UK study found that the peak incidence of PSP is at the age of 70–74 years with a prevalence of 18 cases per 100,000 populations [9]. In persons older than 80 years, the PSP occurrence rate is on average 14.7 cases per 100,000 populations [7, 9]. Japanese STUDY OF autopsy materials, however, suggests that the occurrence of the PSP frequency in elderly people may be even higher [15].

The aetiology and pathogenesis of PSP are not fully understood. The disease belongs to the tauopathies group (the same group includes Pick's disease, Parkinsonism-ALS-dementia, cortico-basal degeneration, primary age-related tauopathy (PART) / Neurofibrillary tangle-predominant senile dementia, chronic traumatic encephalopathy, fronto-temporal dementia, Lytico-Bodig disease, ganglioglioma and ganglio cytoma, meningoangiomatosis, post-encephalic parkinsonism and sub-acute sclerosing panencephalitis, etc.), in which certain hyper phosphorylated-protein forms are accumulated in neurons and glial cells [2, 4, 10, 12]. Several PSP clinical phenotypes are known, They include PSP with Richardson's syndrome (PSP-RS), PSP with predominant parkinsonism (PSP-P), PSP with pure akinesia AND gait freezing (PSP-PAGF), PSP with cortico-basal syndrome (PSP -CBS), PSP with predominant speech and/or language dysfunction (PSP-AOS and PSP-PNFA - PSP-progressive non fluent aphasia), PSP with predominant frontotemporal dysfunction (PSPFTD), PSP with cerebellar ataxia (PSP-C), PSP with primary lateral sclerosis (PSP- Pls) [14].

With the development of neuroimaging technologies, descriptions of a number of symptoms typical for PSP have appeared [3, 5, 8, 13, 18]. Conventional magnetic resonance imaging (MRI) in T2 mode at 1.5 T in patients with atypical Parkinsonism shows a low level of the signal in the putamen of the lenticular nucleus. This low signal is AS A result of an increased iron content [5, 13]. The MRI of approximately 70-80% of patients with PSP shows a decrease in the anteroposterior size of the midbrain with the formation of the Mickey Mouse symptom [13, 16, 18].

According to Boxer A. et al. atrophy of the midbrain and upper cerebellar peduncles is an important criterion for the differentiation of PSP-RS and other clinical forms of Parkinsonism [3]. It should be noted that the MRI signs of PSP are highly specific but less sensitive than clinical criteria [8, 13, 18]. “Hummingbird” and “morning glory flower” symptoms OF PSP WITH 100% specificity are described, but their sensitivity is 68.4% and 50%, respectively) [13].

Also known is the so-called MRI Parkinsonism index (MRPI) with a sensitivity of 100% and specificity of 99.2% –100% for PSP-RS []. The ratio of the sizes of the pons and the midbrain [13] is also used as a diagnostic

criterion. Further research with prospects for use of functional MRI, PET and other modern neuroimaging methods in PSP [4, 9] are being considered. Scientific databases OVID, PubMed, EMBASE, have published more than 400 scientific papers within the periods of 2009-2019, on the problems of diagnosing PSP. Nevertheless, only two of them mention the diagnostic role of the tiger eye symptom [1, 8], which is usually associated with a form of the neurodegeneration with brain iron accumulation - pantothenate kinase-associated neurodegeneration (PKAN) - (Hallervorden – Spatz syndrome) [6]. Particular for PSP, AND also occurring in Wilson-Konovalov disease, FOS poisoning, and some other pathological conditions [5, 7, 13]. With a high-resolution MRI (3 T or more), the sign of a "tiger eye" could occur even in healthy subjects [13].

Case Presentation

This study is devoted to the description of the case of PSP with a pronounced "tiger eye" symptom during an MRI scan.

Patient L., born in 1951, was treated in the Neurological Department of the Regional Clinical Hospital (Odessa, Ukraine) in December 2018. Her son reported that the patient had been suffering from this condition for 10 years. At its early stages; slow movements, lethargy, periodic episodes of dizziness were experienced, accompanied WITH A LOSS OF BALANCE FALLING FORWARD. As the disease progressed, gait worsened. AND ABOUT one and a half years ago symptoms as a frozen look appeared, speech deterioration, and swallowing disorders occurred in the form of choking when eating, memory LOSS and voice changes.

Neurological examination showed a decrease in cognitive functions (MMSE = 18 points), hypomimia, paralysis of the vertical gaze - there are no saccades, restriction in eyeballs movements vertically, slowing down of horizontal saccades, mild dysphagia, mild dysarthria. Pharyngeal and palatal reflexes were diminished. There were bilateral brisk tendon and Periostal reflexes, D = S. Positive hand pathological signs. Rough symptoms of oral automatism and resistance to passive movements affecting both agonist and antagonist muscles with periodic cogwheel-like modifications of muscle tone were detected. She had slow gait and was unstable in the Romberg position. Patient failed the finger-nose test in the sitting position. Convincing data for the violation of sensitivity is not available. Incontinence was experienced periodically. Comparing the MRI performed on admission,

changes (Fig. 1) in the region of the subcortical nuclei were traced to be a decrease in the signal intensity of the medial segments of the pallidum on T2-weighted axial images (due to excessive accumulation of iron) in the longitudinal hyper-intensive area (area of gliosis and vacuolization).

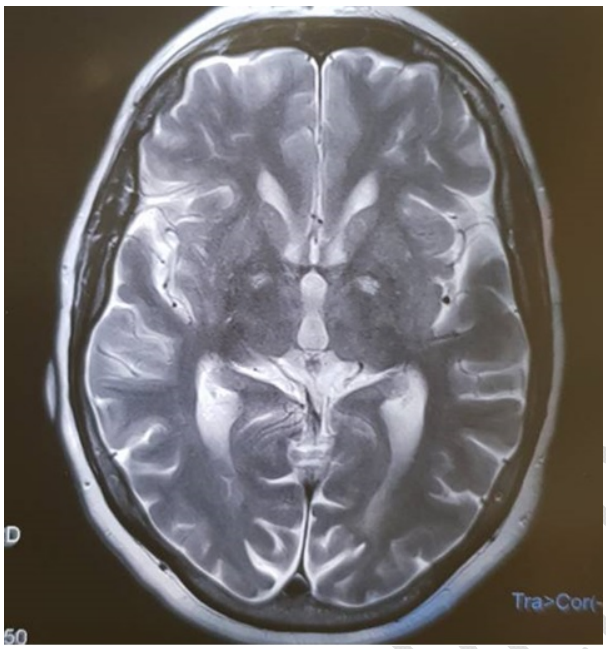

Fig. 1 Symptom "eye tiger" in a patient with PSP

The patient WAS diagnosed with progressive supra nuclear palsy (Richardson-Steele-Olszewski disease) with symptoms of atypical Parkinsonism, paralysis of the vertical gaze and mild subcortical-frontal dementia.

Discussion

Until recently, the “tiger eye” symptom was considered pathognomonic for Hallervorden – Spatz syndrome only. This is a rare autosomal recessive disorder which mainly affects the basal ganglia and is associated with the accumulation of iron in the brain. The most characteristic signs of the disease are a parkinsonian syndrome, various types of hyperkinesis, pyramidal signs, cognitive decline, pigmentary retinopathy and atrophy of

the optic nerves [6].

Davie C. et al. (1997) showed the results of MRI scanning and proton MR spectroscopy, performed in nine patients with PSP-RS [5]. Three of them showed characteristic signs of the "tiger eye" symptom. The authors believe that this symptom can be used for the differential diagnosis of various forms of atypical Parkinsonism.

Some authors consider that at the present time there is no reason to consider the symptom of the "tiger eye" as pathognomonic exclusively for Hallervorden – Spatz syndrome aka PKAN. In particular, in a number of observations, changes in the medial segments of Globus pallidus on T2-weighted axial images were due to multisystem atrophy and neuroferritinopathy [9].

Conclusion

Along with other MRI criteria for the diagnosis of PSP, the MRI sign symptom of the "tiger eye" may occur in some patients with this disease. This radiological sign serves as a pathological correlate, indicating the possibility of developing neurodegenerative disease according to a single universal mechanism of neuronal death in various parts of the brain that determine the specificity of clinical manifestations.

CONSENT

Not applicable.

ETHICAL APPROVAL

The manuscript was approved by LEC of Odessa Regional Hospital.

COMPETING INTERESTS

The author has declared that no competing interests exist.

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