Case Report

Atypical clinical manifestation of dementia: a progressive visuospatial deficit

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Abstract: Clinically and neuropathologically dementia is a very heterogeneous disease. Alzheimer’s disease (Alzheimer disease, AD), the most common cause of dementia, is an acquired cognitive and behavioral impairment of sufficient severity that markedly interferes with social and occupational functioning. This disease may be manifested as progressive memory problems, visuospatial deterioration, aphasia and/or deficits of executive functions. The patient we report here is a 73-year-old female patient with loss of ability of finding directions and understanding the shapes of materials. Her complaints increased progressively during the last two years. Even if she had retired from tailoring, she had deterioration in the visuospatial tests. Also there was occipitoparietal involvement in her cranial MR imaging, SPECT, and PET investigations. In this article a patient with a progressive visuospatial deficit, as an atypical presentation of Alzheimer’s disease, will be described.

Key words: Progressive visuospatial deficit, Alzheimer’s disease, dementia

Introduction

All primary degenerative diseases causing dementia are usually present with a prominent primary anatomical tendency (which is unchangeable) and a related clinical presentation. Alzheimer’s disease is a degenerative illness characterized by progressive cognitive deterioration, which typically starts with deficits in memory followed by affected executive, language, and visuospatial functions (1). Focal degenerative syndromes that are not typical for Alzheimer’s disease have been described. These are progressive visuospatial deficit, primary progressive amnesia, primary progressive aphasia, and primary behavioral changes (2).

In this case report we describe a 73-year-old female patient with atypical presentation of AD.
Case

Our patient was a 73-year-old woman complaining of losing ability to find directions and to understand the shapes of materials, which had been present for the previous two years. The patient reported that her complaints increased in the last 3 months; she reported having difficulty finding the bathroom at home, taking her trousers instead of her stockings from the cupboard, having difficulty to open the doors because of being unable to find the door handles, intending to watch TV but finding herself looking at the opposite wall, having difficulty finding the tap in the lavatory and the bathroom, and tripping and falling on the carpet. Her relatives reported that during the previous three months, she could not get out of her house alone, took the bread instead of the glass when she wanted to drink water, and started to have more falls. The patient did not report any visual problems. She had graduated from primary school (five years of education) and had worked as a tailor in the textile industry for about thirty years. She had been very good at designing dresses. Her previous medical history and familial medical history was unremarkable.

The somatic neurological examination of the patient was normal. Optimallogic examination was completely normal. Eye movements, direct/indirect light reflexes, and visual field examination were normal. Her speech was normal; she could state her complaints easily, coherently, and without any problem. She could recognize her fingers easily, her right-left orientation was normal. She had no Parkinsonism and her color recognition was normal.

The patient’s Mini Mental State Examination (MMSE) score was 23/30. Within this examination, writing was normal but copying figures was defective. The visuospatial tests conducted after the MMSE, such as cube drawing, flower drawing (Figure 1), Luria drawing and clock drawing tests (Figure 2), were affected. Bilateral cortical atrophy, especially prominent in the occipitoparietal areas, was seen in her cranial magnetic resonance (MR) imaging. Cortical hypometabolism of the occipitoparietal areas were more prominent in the single photon emission computed tomography (SPECT) and positron emission tomography (PET) investigations (Figure 3).

It was concluded that the patient had progressive visuospatial deficit and cholinesterase therapy (donepezil 5 mg/daily) was initiated. The response to the treatment was poor at the 1st and 3rd months. The patient died after a tragic traffic accident on the sixth month of follow-up.

Discussion

AD is an example of syndromes that neurologists, psychiatrists, and geriatricians make their diagnosis with the clinical approach. In fact, the proper term of this clinical diagnosis should be “Probable Alzheimer’s Disease”, as the definite diagnosis can be defined after pathological examination. The exact or relative frequency of the different or atypical forms of this clinical syndrome is not known. Galton et al. reported that 14% of their series of 180 patients were presented with atypical forms of AD. Six of these patients (24% of the atypical forms) had progressive
visuospatial deficit (3). Price et al. reported in their series of the consecutive autopsies of 20 Alzheimer’s patients, 2 cases in whom the visuospatial deficit was more prominent than memory and attention deficits (4). There is only one case in the literature with visual deficits that has been proven to be AD with pathologic evaluation, which is a rarer presentation compared to occipitotemporal and bilateral parietal syndrome (5). The principal feature of this syndrome is the deficit in visual processing. Neurodegeneration is more frequent in bilateral occipitoparietal regions. In our case there were similar unusual deficits in visuospatial functions that were reflected by the changes in neuroimaging. In the SPECT investigation, there was hypometabolism in the occipitoparietal areas, while there was atrophy in the MRI investigation, although widespread, it was more pronounced in the occipitoparietal cortical areas.

The first symptoms are getting lost even in familiar places (outer space orientation deficit), being unable to find an object even though it is right in front of the patient, having difficulty while reading (the lines becoming disordered), the deterioration of writing characters, and difficulties in daily functions like dressing up and eating. Visuospatial neglect may be one of the first findings. A vaguely described “visual cloudiness” may be the first complaint; approximately half of the AD patients have this
complaint. Ophthalmologic examinations and eyeglasses do not help (6). Our patient had difficulty identifying various objects in her visual field and difficulty in dressing up, yet her ophthalmologic examination was completely normal.

In the last 20 years it has become more evident that symptoms other than memory deficit are prominent in slowly progressive cortical syndromes. These focal cortical syndromes may include progressive aphasia and syndromes of visual perception or spatial syndromes. There may be cognitive deficits, revealing the closely woven relations between the parietal cortex areas responsible for orientation to place and space and areas for language and executive function. Benson et al. diagnosed their 5 patients with posterior cortical atrophy (7). In our case, the prominent deficit was that of the visuospatial functions rather than any memory problem; figure drawing (2 pentagons) in the mini mental status examination (MMSE), and cube drawing and flower drawing tests were affected as well as the clock drawing test. Defective Luria test may be explained by the pathology progressing to the frontal areas. Although there was a mild atrophy in the cranial MR imaging of the patient, there was no finding to explain this in the SPECT investigation. Additionally, her memory and insight were preserved, compared to the classical forms of AD (8).

The main characteristics of progressive visuospatial deficit are insidious onset and continuous progression of visual and spatial deterioration. This deficit presents itself with components of Balint's syndrome (optic ataxia, visual orientation deficit, and simultanagnosia), deficit in orientation to place and space, hemispatial neglect, and dressing apraxia (9). As a manifestation of bilateral parietal cortex involvement, some patients show a clinical picture of a combination of Balint's and Gerstmann's syndrome, while others have been reported with an asymmetric presentation (7). Our patient could recognize her fingers, her right-left orientation was normal; however, she had a dressing apraxia.

In contrast to typical Alzheimer's cases, there are unique areas of reduced metabolism in visuospatial deficit (10). In imaging studies, bilateral occipitoparietal atrophy, hypoperfusion, and hypometabolism are seen, fitting this profile. In the present case, hypometabolism and hypoperfusion of the occipito-parietal areas were demonstrated with SPECT and PET investigations. Even rarely, occipitotemporal cortical defects can be demonstrated in this group. When neurodegeneration is prominently in this area, object agnosia, prosopagnosia, and alexia are the primary findings. In that case, selective hypometabolism of these areas can be demonstrated with PET.

Warren et al. reported a case of a 60-year-old woman with a visual variant of the Alzheimer's disease; single photon emission computed tomography abnormalities were most marked in the parietooccipital regions of the brain. After treatment with donepezil, improvement was noted on neuropsychological testing and on brain SPECT, including increased perfusion (metabolism) in the occipital lobes (11). Our patient did not benefit from donepezil at the 1st and 3rd months. We did not have the chance of long term follow up because she had a traffic accident and died.

Progressive visuospatial deficit is a rarely seen presentation of Alzheimer's disease. The diagnosis can be reached through clinical and neurocognitive findings plus imaging methods and also through pathology. It is very educative; allowing the combination of neurocognitive information with the findings from the clinical examination and imaging methods.

References


