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Cortıcobasal Degeneratıon and Posterıor Cortıcal Atrophıa: a Case Report

Degeneração Cortıcobasal e Atrofıa Cortıcal Posterıor: Relato de Caso

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Abstract

Corticobasal degeneration is an atypical Parkinson's syndrome characterized by cerebral cortical findings such as apraxia or loss of cortical sensation. The aim of this study is to show the effect of occupational therapy interventions in a patient followed up with a diagnosis of corticobasal degeneration and posterior cortical atrophy. The study included by a 59 year old female patient. After the initial evaluation, she received an Occupational Therapy intervention for one hour, once a week, for eight weeks in our department. A final assessment was made at the end of eight weeks. As a result of the evaluations, significant improvements were observed in the functional independence scale, mini mental state test, and tinetti test. There is a decrease in the geriatric depression scale, but it is still at the border of depression. Progress in sensory evaluations is not significant despite the intervention, but it should be noted that the person has a progressive chronic disease. In conclusion, although corticobasal degeneration and posterior cortical atrophy are progressive neurodegenerative disorders, slowdowns in the course of the disease can be observed with planned person-centered rehabilitation interventions. The importance of early rehabilitation in such degenerative disorders can be emphasized and it is recommended to add occupational therapy interventions to these treatment programs.

Keywords: Corticobasal Degeneration. Occupational Therapy. Case Reports.

Resumo

A degeneração corticobasal é uma síndrome de Parkinson atípica caracterizada por achados corticais cerebrais, como apraxia ou perda de sensibilidade cortical. O objetivo deste estudo é mostrar o efeito de intervenções de terapia ocupacional em um paciente acompanhado com diagnóstico de degeneração corticobasal e atrofia cortical posterior. O estudo incluído por uma paciente do sexo feminino de 59 anos. Após avaliação inicial, recebeu intervenção de Terapia Ocupacional por uma hora, uma vez por semana, durante oito semanas em nosso serviço. Uma avaliação final foi feita ao final de oito semanas. Como resultado das avaliações, foram observadas melhorias significativas na escala de independência funcional, mini teste do estado mental e teste de tinetti. Há uma diminuição na escala de depressão geriátrica, mas ainda está na fronteira da depressão. O progresso nas avaliações sensoriais não é significativo apesar da intervenção, mas deve-se notar que a pessoa tem uma doença crônica progressiva. Em conclusão, embora a degeneração corticobasal e a atrofia cortical posterior sejam distúrbios neurodegenerativos progressivos, abrandamentos no curso da doença podem ser observados com intervenções planejadas de reabilitação centradas na pessoa. A importância da reabilitação precoce em tais distúrbios degenerativos pode ser enfatizada e recomenda-se adicionar intervenções de terapia ocupacional a esses programas de tratamento.

Palavras-chave: *Degeneração Corticobasal. Terapia Ocupacional. Relatos de Caso.*

Introduction

Corticobasal degeneration(CBD) is a rare and slowly progressive sporadic neurodegenerative disease. Corticobasal degeneration is an atypical Parkinson's syndrome characterized by cerebral cortical findings such as apraxia or loss of cortical sensation[1]. In addition, the main characteristic features of corticobasal degeneration include progressive clinical course, asymmetric involvement, rigidity. Currently, there are two clinical phenotypes of corticobasal degeneration. The first is a classic motor presentation with asymmetric Parkinson's symptoms, apraxia or loss of cortical sensation; the second is the presentation of dementia, which usually has focal cortical findings with or without Parkinson's characteristics[2].

Posterior cortical atrophy (PCA) syndrome is a rare clinical manifestation of several neurodegenerative diseases affecting the parieto-occipital cortex. The most common underlying pathology is caused by Lewy disease, progressive subcortical gliosis, corticobasal degeneration or prion diseases. The most prominent clinical feature of PCA is complex visual disorders such as object agnosia, simultaneous agnosia, optic ataxia and oculomotor apraxia, while the basic visual functions remain intact. These deficits lead to multiple impairments in activities of daily living that require visual control[3].

The aim of this study is to show the effect of occupational therapy interventions in a patient who was followed up with a diagnosis of corticobasal degeneration and posterior cortical atrophy.

2 Case Report

AK, who was followed up in the Occupational Therapy department of Hacettepe University, was included in the study. Functional Independence Measurement (FIM), The Mini Mental State Test, Tinetti Balance and Gait Assessment, Geriatric Depression Scale (GDS-15), Loewenstein Occupational Therapy Cognitive Assessment(LOTCA) and Sensory evaluations tests were used in the study. After the initial evaluation, she received occupational therapy intervention for eight weeks, one hour a week, in our department. At the end of eight weeks, the final assessment was made.

AK, a 59-year-old female patient, lives with her son. She provides her care with the help of caregiver. She lost his wife five months ago. She was referred to occupational therapy due to problems in visual-spatial skills, visual perception and neglect of the left side.

Since this study is a case report, an ethics committee number was not obtained.

2.1 Assessments

2.1.1 Functional Independence Measurement

Functional Independence Measurement (FIM) was used to evaluate the patient's activities of daily living. FIM was organized by the American Congress of Medical Rehabilitation and the American Academy of Physical Therapy and Rehabilitation in 1983 and published by Hamilton et al. in 1987, and adapted into Turkish by Küçükdeveci et al. in 2001[4,5] It is a test that includes 13 motors and 5 cognitive components. These components consist of self-care, eating, hygiene activities, bathing, toilet, transfers, locomotion, communication, social communication and dressing activities. The measurer scores all parameters on a scale of 1-7 (1: fully dependent on completing the activity, 7: completing the activity independently.)[4].

2.1.2 Mini Mental State Test

The Mini Mental State Test was first published by Folstein et al. (1975). It consists of eleven items gathered under five main headings as orientation, recording memory, attention and calculation, recall and language, and is evaluated out of a total score of 30[6].

2.1.3 Tinetti Balance and Gait Assessment

Tinetti Balance and Gait Assessment evaluates balance ability and gait under 2 main headings: the first 9 questions are about balance and the next 7 questions are about walking. Calculation of the survey score; The total score of the first 9 items gives the balance score, the total score of the next 7 items gives the walking score, and the sum of the balance and walking scores gives the total score. Item 16 is the whole of the actions done during ADL. As a result of the evaluation made by observation, the scoring is as follows: 2 points; correct execution of the indicated movement, 1 point; performing the specified movement with adaptations, 0 points; failure to move. If the total scale score is 18 and below, it indicates a high risk of falling, a score of 19-24 indicates a moderate fall risk, and a score of 24 and above indicates a low risk of falling[7].

2.1.4 Geriatric Depression Scale (GDS-15)

The GDS-short form, used in this study to determine the depression level of individuals, consists of 15 questions questioning the mood of the patient. Answers are given based on feelings from the past week; The answers are "yes" or "no", just like in the long form, and 1 point is given for a "yes" answer or a "no" answer, depending on the question. While scoring the scale, 1 point is given for each answer given in the direction of depression and 0 point is given for positive answers [8].

2.1.5 Loewenstein Occupational Therapy Cognitive Assessment(LOTCA)

This test includes four areas. These areas are; orientation, perception, visual motor organization, thinking processes. The test battery has 20 subtests[9]. In our case, we used only the spatial perception and praxis subtests of the sensing area of the test. Due to the physical and mental condition of the patient, other test parameters could not be applied. The tests used in the study are the two subtests of the perception test. Spatial perception subtest: Evaluated in three parts. In the first of these, the person is asked to make a right-left distinction on himself. In the second, the person is asked to do it by giving instructions about the pen and box in front of him. and in the third, the person who evaluates by sitting opposite to the person is asked to give instructions on the right-left distinction. Praxis subtest: Evaluated in three parts. these; motor imitation, demonstrating the use of objects, and symbolic events.

2.1.6 Sensory evaluations

Light Touch: Tested with a small piece of wool. It should be tested at different points of the arm and hand. Superficial Pain: It is evaluated by performing the pointed-blunt test with a pin. Proprioception: The extremity is brought to a position by closing the eyes and the person is asked to bring the opposite side to the same position, if the person cannot do the opposite side, she is asked to bring the same extremity to the same position again. Sterognosis: While the patient's eyes are closed, it is observed that the patient recognizes the shape by touching it and opens her eyes and shows the shape. Barognosia: The barognosy test is performed by blindly placing two objects with an estimated difference of 1-2 kilograms between the hands of the person and asking her to say which one is heavier. Grafestesis: In the test, their perception of simple patterns drawn on the palms is evaluated. Figures drawn with closed eyes are drawn on the palm of the patient, and after the eyes are opened, the patient is asked to

draw the same shape.

2.1.7 Intervention

Praxis skills were studied by simulating the person's ADL activities (eating, hand washing, bathing, dressing, self-care) in one-hour sessions for eight weeks. The intervention program is given in Table 1.

Table 1 - The intervention program

Sessions	Applied Interventions
Session 1	Motivational interview was held. Bathing and dressing activities were simulated and studied. Visual follow-up study was done. The person has no vertical plane tracking, the right side is followed on the transfer axis, but the left side is neglected.
Session 2	Bath and hand washing activities were simulated and studied. She does not use her left hand while performing overhead positions in activities. Person tracking was tried by walking in the corridor, and it loses track when turning to the left.
Session 3	The walking activity was practiced by placing single and colored steps. Since her depth perception was poor, she could not fully realize where the steps ended. Passing was practiced with an exercise ball. She had little strength in the step of taking and throwing the ball by leaning, and she could not find the ball when it went to the left side. Hand washing activity was studied by simulating and completing the activity with less physical stimulus.
Session 4	Memory and place-time orientation were studied by asking various questions about her daily life and family. Hair combing activity was studied. It doesn't scan the left side. Coffee making activity was studied. Since she could not do it herself, she was asked to explain the steps of the activity and the materials and dimensions. Upper dressing activity was studied.
Session 5	Hand washing activity was studied in daily living activities in self-care. Differentiation of objects such as chickpeas, beans, pasta on the felt texture, grip and hand manipulation were studied. Dance therapy was performed for balance and bilateral motor coordination.
Session 6	The balls of different colors, sizes and weights were used to distinguish these features. Due to the neglect of the left side, brushing and approximation was performed on the left arm. Walking and step work was done. It was tried to distinguish with objects such as felt textured apples and flowers.
Session 7	Symbolic events, right-left distinction, use of objects, position in space, motor planning work was done. Eating activity was simulated and studied.
Session 8	Eating activity was studied. Differentiation of objects such as chickpeas, beans, pasta on the felt texture, grip and hand manipulation were studied. Walking and step work was done.

Source: the authors.

The ADL activities studied in the interventions were selected from the problem areas identified in the FIM assessment. Sensory activities, especially tactile, visual and proprioception senses, were studied. Balance and motor coordination skills are studied. Left-sided awareness and bilateral motor coordination skills were studied due to left-sided neglect.

2.3 Results

The first and last case assessments are shown in the tables. As a result of the assessments, significant improvements were observed in the functional independence scale, mini mental status test, tinetti test in Table 2. In the functional independence scale, 12 points between the first and last assessments were achieved by the increase in the parameters of nutrition, dressing, toilet, walking, understanding, expression, and social interaction. The highest increase was in walking and expression parameters, with an increase from 2 points to 5 points. In the mini mental state test, our case progressed from 1 point to 8 points. In the first assessment, she received 1 point from the name of the city where she was located, while she received one point each from the name of the place and sentence repetition parameters, as well as the city in the last assessment. It received 2 points from the immediate recall parameter and 3 points from the motor function parameters. In the Tinetti test, 1 point increase in walking parameter was the route; The increase in the balance parameter was provided by the increase in the rotation sub-parameters.

Table 2 - Result of the functional independence scale, mini mental status test, tinetti test

Assesments	First assessment	Last assessment
Functional Independence Measurement	27/126	39/126
Mini Mental State Test	1/30	8/30
Tinetti Balance and Gait Assessment	Walking: 4/12 Balance: 11/16	Walking: 5/12 Balance: 13/16
Geriatric Depression Scale	8/15	6/15

Source: the authors.

There is a decrease in the geriatric depression scale, but it is still at the border of depression. Spatial perception and praxis, which are LOTCA sub-parameters, are shown in Table 3. While there was no difference in spatial perception in the first and last assessment; In the praxis parameter, 1 point with the symbolic events item decreased in the last assessment.

Table 3 - Result of the LOTCA sub-parameters

LOTCA	First Assessment	Last Assessment
Spatial perception	0	0
Praxis	1	0

Source: the authors.

Sensory assessments are shown in Table 4. It is seen that the sensory differences between the right and left sides of the case show parallel results with left side neglect due to corticobasal degeneration.

Table 4 - Result of the sensory assessments

Sensory Evaluations	First Assessment		Last Assessment	
	Right	Left	Right	Left
Light Touch	0/5	0/5	1/5	0/5
Superficial Pain	9/10	6/10	6/10	4/10
Proprioception	0/5	0/5	0/5	0/5
Sterognosis	4/7	2/7	4/7	3/7
Barognosia	+	-	+	+
Grafestesis	0/5	0/5	0/5	0/5

Source: the authors.

In the first assessment, when we say that the mistakes made in superficial pain are pointed to the blunt side; In the last assessment, the number of errors made in the superficial pain parameter increased. While there was no change in the assessments of cortical senses propriception and grafesthesia, weight awareness was provided on the left side in the assessment of barognosia. Progress in sensory assessments is not significant despite the intervention, but it should be noted that the person has a progressive chronic disease.

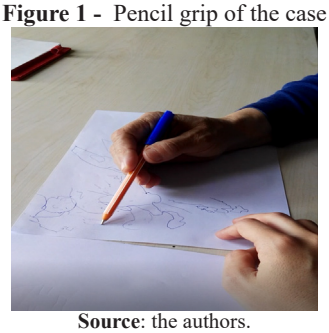
2.3 Discussion

Corticobasal degeneration is a rare disease characterized by a progressive clinical course, asymmetrical involvement, rigidity and apraxia, and minimal cognitive impairment is expected in the early stages of the disease, however, cases with dementia onset have been reported in the literature. Speech problems and swallowing difficulties can only be seen in the advanced stages of the disease [10,11] Our case has speech difficulties and advanced cognitive problems, and our case is at an advanced level.

The most common symptom associated with corticobasal degeneration is apraxia. Limb apraxia is almost always asymmetrical and ideomotor apraxia; tool use and imitation of tool use are deteriorated, while generally the recognition of actions is relatively preserved[12,13]. In our case, a decrease was observed in the praxis parameter, one of the LOTCA parameters, and at the same time, deterioration was observed in the use of spoons in the eating activity and in the use of soap in the handwashing activity. It has been shown that improvements in tool use in these activities after the intervention increased with the results on the Functional Independence Scale. In addition, in a study by Graham et al., the importance of tactile input in facilitating motor performance in tool use was emphasized[14]. Accordingly, we used the approximation and brushing technique in the intervention program of the study. Although there were increases in motor performance in activities, no specific increase was observed in sensory evaluation.

Structural apraxia and impaired handwriting are common in corticobasal degeneration. This finding is consistent with the marked apraxia of the extremities exhibited by most patients. Structural apraxia, as evidenced by drawing and copying tests, is mainly clinically defined[15]. In the watch drawing and writing activity with the case, the pencil grip is weak but the grip position is correct. It cannot produce a meaningful text as shown in Figure 1. The results of one study suggested that handwriting problems in patients with corticobasal degeneration may not be due to apraxia alone. In this study by Graham et al. reported that it may cause difficulty in remembering the shapes of letters[14].

Figure 1 - Pencil grip of the case



Source: the authors.

Findings regarding episodic memory function in corticobasal degeneration are inconsistent and variable, possibly depending on the stage of the disease as well as the patients[16]. Since the case was advanced level, there were memory problems and a motivational interview and memory study were performed.

In the literature, aphasia in corticobasal degeneration is frequently mentioned, but speech is not characterized in detail due to pathological or motor symptoms[10,15]. When we started the study, in our case, the words came out singly and were not intelligible. At the end of the sessions, the patient began to express herself better and to use more than one consecutive word.

Corticobasal syndrome is a progressive neurological disorder. Although patients lose more brain neurons than normal individuals, it has been reported that repetitive facilitation exercises can cause a decrease in the loss of neurons. However, it was stated in this study that more research is needed to elucidate the functional motor recovery mechanisms in patients with corticobasal syndrome during repetitive facilitation exercises and daily living activities training[15,17]. In our study, we achieved improvements in motor functions by simulating activities of daily living, but the development of proprioception sense did not appear significantly in the results during this period.

Posterior cortical atrophy (PCA) is a degenerative syndrome that manifests itself with progressive visual and spatial disorders. In a case with posterior cortical atrophy, specific therapeutic management enabled the achievement of functional goals responsive to the patient's wishes, and improvements were seen in activities of daily living[3]. In line with this case, we made progress in activities of daily living in our case study.

3 Conclusion

In conclusion, although corticobasal degeneration and posterior cortical atrophy are progressive neurodegenerative

disorders, slowdowns in the course of the disease can be observed with planned person-centered rehabilitation interventions. The importance of early rehabilitation in such degenerative disorders can be emphasized and it is recommended that occupational therapy interventions be added to these treatment programs.

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