

Asperger Syndrome with Highly Exceptional Calendar Memory: A Case Report

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Abstract

Some patients with pervasive developmental disorders develop unusual talents, which are characterized as savant syndrome. Herein we present neuropsychological examination and brain imaging (fMRI and brain SPECT) findings of an 18-year-old male with Asperger syndrome and highly unusual calendar memory. Neuropsychological evaluation of the case indicated mild attention, memory, and problem solving deficits, and severe executive function deficits that included conceptualization, category formation, and abstraction. Functional MRI findings showed activation above the baseline level ($P < 0.05$) in the bilateral inferior parietal lobule, precuneus, superior and middle frontal gyri, and medial frontal cortex. Brain SPECT findings, in comparison to rest-SPECT findings, showed that there was hypoperfusion in some brain regions, including the right frontal cortex and right parietal cortex. Baseline blood perfusion in the left frontal cortex was also observed, as well as hypoperfusion in the right parietal-occipital cortex and in the right basal ganglion (compared to the left side). The results of the present study and further research will contribute to our understanding of calendar memory and savant syndrome.

Key Words: Savant syndrome, Asperger syndrome, Calendar memory, fMRI.

INTRODUCTION

Asperger Syndrome (AS) was first defined as autistic psychopathy in 1944 by an Australian physician, Hans Asperger, based on his observations of 4 cases that had problematic peer relations. Today, severe language problems, and mutual relationship and social interaction deficits are accepted as the primary symptoms of AS (Ghaziuddin and Butler, 1998; Iwanaga et al., 2000). Excessive interest in specific topics is prevalent in AS. AS is defined by the DSM-IV and ICD-10 classification systems under the heading, "Pervasive Developmental Disorders" (WHO, 1992; APA, 1994).

Isolated unusual talents (savant syndrome) can be observed in patients with pervasive developmental disorders. One of the most prevalent examples of savant syndrome in autistic disorders is calendar calculation,

or calendar memory (Kahne et al., 2002; Mottron et al, 2006; Newport, 2006). Only a few studies have examined the relationship between AS and calendar memory, and none clearly explain the underlying neuronal mechanism of this ability (Heavey et al., 1999). Herein we present the clinical and neuropsychological characteristics, and brain imaging profile of an AS case with unusual calendar memory.

Case

ND was an 18 year-old male AS patient in the 10th grade. His first clinical interview included his father. ND claimed that he did not have any significant problem and therefore it was not necessary to consult a psychiatrist. His parents claimed that they consulted a psychiatrist because ND has always been introverted, does not often speak with others, and has poor academic performance.

Received: 02.09.2009 - Accepted: 29.01.2010

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In the examination room ND was reluctant to answer any questions and spoke in a monotone voice. He avoided eye contact and sometimes displayed mimics not fitting the content of the conversation. His developmental history revealed that he was the oldest child in the family and that his birth was unplanned. His mother was only 16 when he was born. Pathological signs were not observed during the pregnancy or delivery. In infancy he had been a quiet and calm baby; he slept well and did not cry when his parents left him alone. His parents reported that he did not react when strangers held him in their arms. His mother reported that he had acted as if he was not aware of the people around him.

His teachers observed that he did not initiate conversations, although he answered questions when he was asked directly. He also did not have any friends in school. His parents reported his teachers recommended that they consult a child psychiatrist. His parents reported that ND surprised people by correctly remembering details of historical events that his father and teachers could not.

ND's parents first consulted a neurologist when he was in the 4th grade, with the complaints of excessive thoughtfulness, excessive stillness, and reluctance to speak with others. Neurological examination and tests revealed no neurological problems, and the neurologists referred the family to the hospital's child psychiatry outpatient clinic. The family then presented to the child psychiatry outpatient clinic with the complaints of ND's excessive thoughtfulness and forgetfulness. ND would forget his school bag at home, go to school wearing regular clothing instead of his school uniform, or forget to wear his shoes. Moreover, he sometimes seemed unaware when people called him by name. Additionally, his parents reported that he would ask his parents weird questions, such as "is this piece of paper alive?" or "why doesn't water get into the eyes of fishes?" According to his past psychiatric records, his mean Wechsler Intelligence Scale for Children, Revised Version (WISC-R) score was 75. His verbal intelligence quotient (IQ) was 85 and his performance IQ was 75. His developmental history (according to past psychiatric records) revealed normal walking and speech development, and toilet training. The psychiatrists had considered a diagnosis of AS, but as ND did not have isolated and special interests he was diagnosed with pervasive developmental disorder.

Two years ago he began attending a new school and at that time his problems exacerbated. He stopped paying attention to how he dressed. He failed the 9th grade

and his new friends made fun of his excessive quietness and weird speech, which resulted in even less communication with his peers. During the last few years, he developed an excessive interest in calendars. He spent most of his time looking at old calendars that hung on the walls of his room and reading old newspapers. He began to write old calendar dates by heart. In his leisure time he read old newspapers on the Internet and read about past events that occurred on specific dates on old calendars. He memorized the details of the events and the dates of these events, including which day of the week. He became interested in history courses at school. He tried to establish friendships by sharing his knowledge of historical events and their dates, but failed to attract the attention of his peers.

Psychological examination revealed that ND had diminished self-care. Although he reported that it was not his decision to consult to a psychiatrist, his attitude toward the clinical interview was positive. He was conscious, cooperative, and had good orientation. He spoke clearly and understandably, but with a monotone voice and with little eye contact. His mood was slightly depressed. No psychiatric symptoms were noted, and his reality testing and judgment were both good.

Diagnosis

The most salient and consistent characteristic of ND's childhood history (even in the preschool period), was that he never achieved age-appropriate relationship patterns with peers. Moreover, ND developed unusual calendar memory and excessive interest in historical events. He also memorized the content and date-related details of historical events. Additionally, there was no speech delay or major problems with communication in his developmental history. These symptoms and case history details are consistent with the AS diagnostic criteria of DSM-IV. In the differential diagnosis his lack of speech problems, such as delayed speech development, and his current history supported the diagnosis of AS rather than autism (autism savant).

Calendar memory and neuropsychological examination

In order to assess ND's calendar memory we randomly selected calendars from the last 300 years with the help of a computer program. We asked the patient the weekdays of randomly selected calendar dates. The patient correctly answered all questions with a mean delay of 1 s. This procedure was repeated on subsequent days

and the results were consistent with his performance on previous trials.

We developed an evaluation sheet in order to assess ND's memory of the details and dates (day/month/year) of historical events. This evaluation sheet included dates of significant historical events that occurred during the past 100 years (e.g. death of significant people and general elections in Turkey). ND's performance remembering dates was good, though not as good as his calendar memory performance. For example, ND correctly remembered the dates and weekdays of the last 6 general elections and of the last 3 bye-elections in Turkey. He remembered the dates that significant people died and the dates of past newsworthy events of worldwide significance.

The patient's general intelligence level (intelligence quotient [IQ]) was assessed using the Wechsler Adult Intelligence Scale (WAIS), verbal memory functioning was measured using Rey's Auditory Verbal Learning Test (AVLT), verbal ability was assessed using the Verbal Fluency Test, visual memory skills were assessed using Benton's Visual Memory Test (Benton VMT), and executive functioning was measured using the Wisconsin Card Sorting Task (WCST).

WAIS verbal IQ, performance IQ, and general IQ scores were, respectively, 97, 89, and 93. ND scored lowest on the WAIS similarity subscale and highest on the WAIS general knowledge subscale. He performance was lower on the picture completion and object assembly subscales than on the other performance subscales. The results indicated mild attention and problem-solv-

ing deficits, and severe category formation and abstract thinking deficits. ND's WAIS intelligence scores were higher than his WISC-R scores, yet this difference was considered acceptable due to the nature of the WAIS.

ND's performance on Rey's AVLT showed that he had a normal short-term memory span; however, he scored low on the learning subscale. Moreover, he made pattern alteration and perseveration errors on Benton's VMT. ND appeared to have the ability to produce words with a phonological association with the given words on the Verbal Fluency Test. In other words, he could say words that had the same first syllable based on the similarity of the sound of the syllables.

ND was not able to complete any of the WCST categories. He had high perseveration scores based on WCST age and sex norms. These results indicated severe executive dysfunction, which is consistent with the results of other tests. His general performance on the neuropsychological tests indicated that ND had mild attention, memory, and problem solving deficits, and severe deficits in executive functioning, such as conceptualization, category formation, and abstract thinking ability.

Brain imaging findings

Functional magnetic resonance imaging (fMRI) and brain perfusion SPECT were used to assess the neurological characteristics that correlated with ND's odd symptoms. For fMRI gradient-echo planar (EPI-GRE) and BOLD (blood-oxygenation level dependent) images were obtained using a device with a 1.5 power magnetic field (Symphony, Siemens, Erlangen, Germany) (TR/

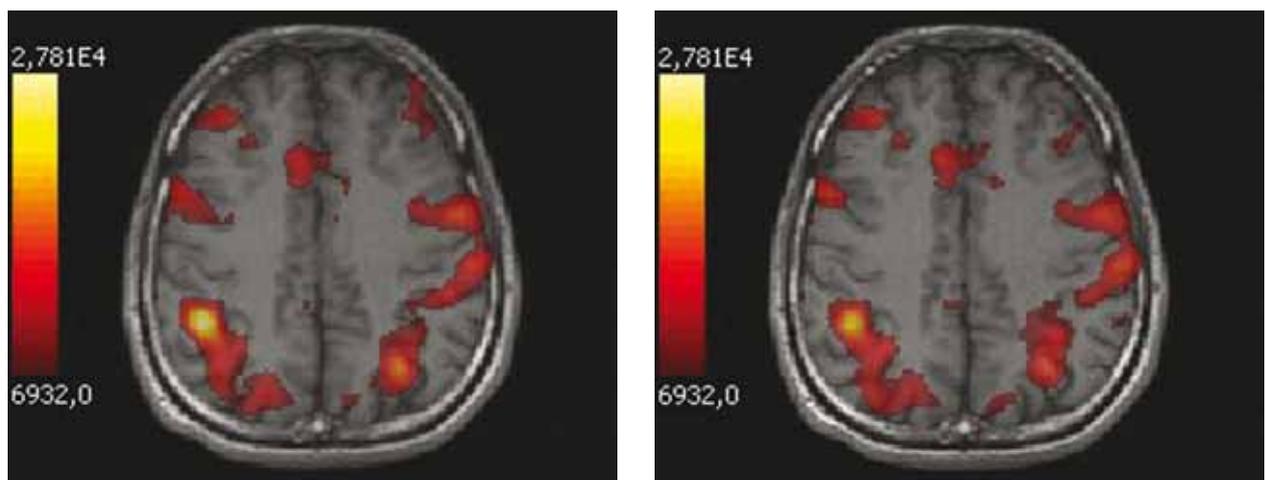


FIGURE 1A and B. Activation above the baseline level ($P < 0.05$) is observed in the bilateral inferior parietal lobule (Broca Area- BA 40), pre-cuneus (BA 7), superior and middle frontal gyri (respectively, BA 10 and 46), and medial frontal cortex with fMRI.

TE: 3700/50 ms). The paradigm of the evaluation included equal lengths of rest and active conditions (in which ND tried to remember the weekday of a given date) (20 dynamic evaluations) (block design per condition). Dynamic volume quantity was 120, including 3 rest and 3 active periods. ND's memory performance was assessed the following day outside of the magnet and the patient remembered the weekdays of all the given dates. The data were analyzed using Nordic Ice software (Nordic Imaging Lab, BOLD analysis, Norway). Activation over the baseline level ($P < 0.05$) was observed in the bilateral inferior parietal lobule (Broca Area- BA 40), precuneus (BA 7), superior and middle frontal gyri (respectively, BA 10 and 46), and medial frontal cortex with fMRI (Figure 1A, and B).

Using brain SPECT regional cerebral blood flow (rCBF) was examined in 2 stages:

1. **Rest-SPECT:** Tc-99m HMPAO (hexamethyl propylene amine oxime) was used as the radiopharmaceutical. The patient rested for 10 min in a dimly lit and quiet room before receiving an intravenous injection of 740 MBq Tc-99m HMPAO. Brain perfusion SPECT images were obtained 15 min after the injection using a double-headed SPECT gamma camera (Siemens, E-CAM, Erlangen, Germany).

2. **Active-SPECT:** Active-SPECT images were on another day subsequent to Rest-SPECT. ND was asked to remember the weekdays of given dates. ND received an intravenous injection of 740 MBq Tc-99m HMPAO. For 2 min following the injection the researcher continued to ask ND the weekdays of given dates in order to keep patient active and to enhance the spread of the radiopharmaceutical agent into the neurons. Brain perfusion SPECT images were obtained 15 min later.

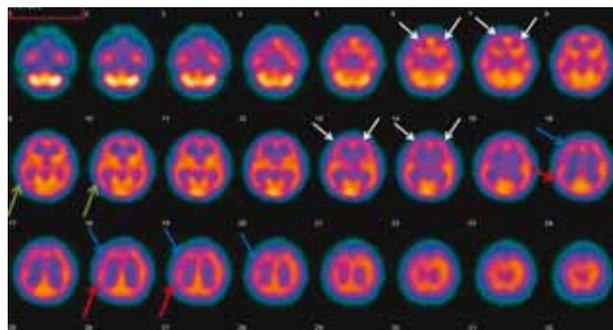


FIGURE 2. Rest-SPECT images of the patient. In the posterior segments of the brain hypoperfusion in the right frontal cortex (blue arrows) in the anterior regions, and hypoperfusion in bilateral frontal cortex (white arrows), right parietal cortex (red arrows), and right temporal cortex (compared to its symmetry) (green arrows) of the brain are observed.

rCBF evaluation was based on segments that were parallel to the orbitomeatal lines of the SPECT images. The distribution of radioactive matter in the cerebral cortex, cerebellum, and subcortical brain regions, increases or decreases in focal or global perfusion, and right-left anterior-posterior perfusion asymmetry were examined.

In the posterior segments of ND's brain hypoperfusion in the right frontal cortex (blue arrows) was observed. In the anterior segments of the brain, hypoperfusion in the bilateral frontal cortex (white arrows), right parietal cortex (red arrows), and right temporal cortex (compared to its symmetry) (green arrows) were noted. Perfusion in the other cortical and subcortical regions, and the bilateral cerebellum of the brain was normal (Figure 2).

Compared to the rest-SPECT images, hypoperfusion in the right frontal cortex, including the right parietal cortex (red arrows), was observed. Increased rCBF in the left frontal cortex (blue arrows) was noted. Hypoperfusion in the right parietal-occipital cortex (white arrows) and right basal ganglion (compared to the left basal ganglion) was observed. Perfusion in the other cortical regions, left basal ganglion, and bilateral cerebellum of the brain was normal (see, Picture 3).

DISCUSSION

The presented case had advanced calendar memory ability, which has previously been reported in autism disorder (Kennedy and Squire 2007). Although the literature suggests that differential diagnosis between AS and high functioning autism is difficult, the DSM states that the lack of language problems differentiates AS from high functional autism. The present case has had no lan-

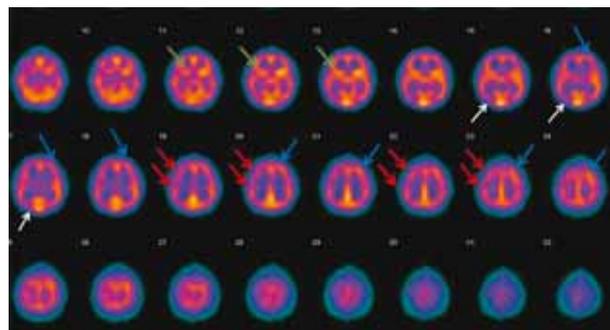


FIGURE 3. Active-SPECT images of the patient. Hypoperfusion in the right frontal cortex, including the right parietal cortex (red arrows), right parietal-occipital cortex (white arrows), and right basal ganglion (compared to the left basal ganglion), and increased rCBF in the left frontal cortex (blue arrows) are observed.

guage problems since early childhood and was therefore diagnosed with AS according to DSM-IV diagnostic criteria. Nevertheless, it has been suggested that there are no clear differences between AS and high functional autism, except in language development, which could be related to differences in the definition of language development (Noterdaeme, 2009). Hence, it was suggested that these disorders are 2 components of an autism spectrum

There are only a few imaging and neuropsychological studies on calendar memory ability (Kahne et al. 2002; Boddaert et al., 2005). In the presented case brain SPECT in the resting condition indicated low rCBF in the right posterior and bilateral anterior segments of the frontal and right temporal regions, and the right parietal cortex. Active SPECT imaging (in which the patient was cognitively activated with calendar memory tasks) showed hypoperfusion in the right frontal and parietal regions, and decreased rCBF in the right basal ganglion. Nonetheless, in the active condition rCBF in the left frontal cortex was higher, as compared to the rest condition.

Similar to our findings, SPECT and PET imaging studies on AS patients reported low rCBF in the frontal and temporal regions of the brain (Sugihara, 2007). A PET imaging study conducted with an autism disorder patient with advanced calendar memory ability reported increased rCBF in the left frontal cortex, including the hippocampus (Boddaert et al., 2005), which is similar to the increased rCBF in the left frontal cortex observed in the presented case. In addition, rCBF in the right frontal and parietal regions, and the right basal ganglion functionally decreased during the process of remembering. These findings indicate that the activity level in the right hemisphere decreased (suppression of right side of the brain), while activity in the left frontal cortex increased during the process of remembering.

The nature of calendar memory ability and whether or not it is related to the memory region of the brain are not fully understood. The presented case did not yield any useful information concerning how he accomplished the calendar memory task. He reported that the answers suddenly come to his mind because he often looks at calendars. He insisted that he did not perform any calculations in his mind, which was supported by his short response times (mean: 1 s). We think that he may have

memorized dates or obtained knowledge concerning calendar systems without any conscious awareness. Studies that compared memory skills in AS patients with those in normal subjects indicated that AS patients' episodic memory performance was lower than the memory performance of the normal subjects, but recognition task performance did not differ between the 2 groups (Bowler et al. 2000).

Similar to the IQ profile of AS patients, the presented case's verbal IQ performance was significantly higher than that of his performance IQ (Volkmar et al., 1996; Öktem, 1998). The presented case scored lowest on the WAIS similarity and picture completion subtests. This finding is similar to what was reported by Öktem (1998) and Devrimci Özgüven et al. (2001), but is in contrast to a report by Szatmari et al. (1990), who suggests that AS patients perform lowest on the comprehension subtest.

AS patients' difficulty forming social relations and understanding the emotional experiences of others might result from information processing problems. In particular, problems processing social cues might be due to executive dysfunction, and problems in conceptualization and abstract thinking, which indicate diminished frontal cortex functioning. This is supported by the SPECT results and neuropsychological profile of the presented case, and previous studies (Pennington and Ozonoff, 1996; Devrimci Özgüven et al., 2001), which suggest that frontal lobe dysfunction should be considered in the etiology of AS.

CONCLUSION

Brain SPECT imaging in the presented case indicates that the left frontal region of the brain was highly activated during the calendar memory task, and the right frontal and parietal cortex were functionally more dominant, as compared to their left counterparts. The presented case study contributes to our understanding of functional alterations in the brain in AS. It supports the idea that AS and savant characteristics, such as unusual calendar memory, are separate phenomena. Unusual calendar memory might be the result of a distinct neuronal circuit that develops with learning, but this idea should be investigated in greater detail with additional imaging studies.

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