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Posterior Form of Alien Hand Syndrome Associated with Idiopathic Hypertrophic Pachymeningitis

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Alien hand syndrome is usually secondary to either a corpus callosum lesion alone or in combination with a lesion in left, right or both medial frontal areas. Alien hand syndrome with non-dominant parietal lesions without callosal involvement has been rarely described. We report here a case of a 64-yr-old man who presented with alien hand syndrome accompanying hemispatial neglect and who was diagnosed with idiopathic hypertrophic pachymeningitis involving the right parietal cortex with the preservation of the corpus callosum.

Key Words: *Alien hand Syndrome, Neglect, Pachymeningitis*

INTRODUCTION

In 1908, alien hand syndrome (AHS) was first described by Goldstein as a feeling a woman's left hand had a will of its own[1]. The symptoms of AHS vary and some investigators proposed that these syndromes could be classified into at least four or five broad categories such as 1) diagonistic dyspraxia and inter-manual conflict, 2) alien hand sign, 3) syndrome of the anarchic hand or way-ward hand, 4) supernumerary hand, and 5) agonistic dyspraxia[2].

AHS is usually secondary to a corpus callosum lesion either alone or in combination with a lesion in left, right or both medial frontal areas, sometimes extending to adjacent cortical regions, thalamus or basal ganglia[3, 4]. AHS with non-dominant parietal lesions without callosal involvement has been rarely described[3-5] as "sensory or posterior form of alien hand sign", as opposed to the "motor or anterior form of alien hand sign". We report a case of posterior form of AHS associated with idiopathic hypertrophic pachymeningitis.

CASE REPORT

A 64-yr-old man who presented the feeling of strangeness of his left hand was admitted to our hospital. His past medical history was remarkable for hyperthyroidism and emphysema for 4 yr but they had been well controlled. He had graduated from primary school and his current job is a plasterer.

Two years ago, he experienced progressive headache and disturbed color vision with normal visual acuity. On admission, a gadolinium enhanced MRI scan of the brain revealed the right tentorium cerebelli to be thickened. Otherwise,

there was no parenchymal involvement. CSF exams including cytology were normal. Opening pressure was also within normal limits (120 mmH_2O). Additional serologic tests including rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibody, angiotensin-converting enzyme, VDRL, TPHA, FTA-ABS, HIV and tumor markers were negative. Meningeal biopsy revealed fibrosis and glial proliferation with lymphocytic infiltration, suggesting necrotizing lymphocytic pachymeningitis. All these evidence being considered, he was diagnosed as idiopathic hypertrophic pachymeningitis. He was given oral prednisolone 1 mg/kg per day for 14 days, gradually tapered and maintained 10 mg prednisolone. Subsequently, his headache and dyschromatopsia was improved.

Two months ago, he felt strangeness of his left hand with difficulties finding the way to his home after going out for a walk or shopping. Whenever he went to the public bathhouse, he paid extra-money (except scrubbing fee) to a body-scrubber because he needed the scrubber's assistance in dressing and undressing or bathing. One month prior to this admission, he could not drive his car any more, because he lost a sense of orientation or direction in the familiar places.

At this admission, he was given the regular medications (prednisolone 10 mg and azathioprin 50 mg and per day). He vehemently considered that his left hand was "silly, indiscrete and injudicious", useless and not in control. He said that "Once, when I tried to catch a door handle, my left hand gropingly reached to it earlier than my right hand. I was surprised and felt as if my left hand was silly, because I am a right-handed person." On neurological examination, he was alert, cooperative and fully oriented. Mini-Mental Status Exam (MMSE) was 20/30. On a Korean version of the Western Aphasia Battery (K-WAB), AQ was 86.4. On a Korean version of the Boston Naming Test (K-BNT), he was able to name 55 items out of 60 (>95 %ile). He was shown his left arm and asked to whom it belonged. He replied "It's mine." He had hemispatial neglect (Fig. 1). He managed to draw a clock, omitting the figures such as 7-10, and 11 on the left side. Several lines of writing were deviated to the right side of the page. He showed significant neglect whenever bisecting lines. Whenever an examiner showed the figures or written letters horizontally, he copied those only vertically. He showed perseveration during the Luria loop test and he could not correctly draw an interlocking pentagon. There were astereognosis in the left hand and agraphethesia in both hands. Optic ataxia, simultanagnosia, prosopagnosia or dyschromatopsia were not present. Inter-manual conflict and mirror movements were not observed. Although motor weakness was not present, he showed dystonic posturing of left hand (extended wrist, flexed and fisted fingers) with

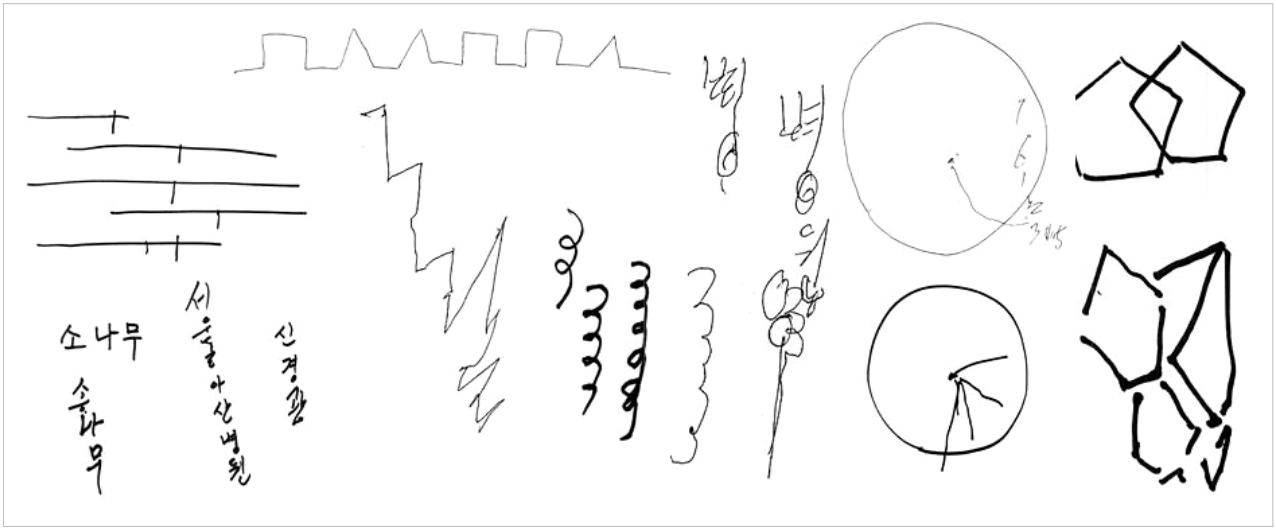

Fig. 1. Writing and drawing of the patient. This patient showed abnormal line bisection, perseveration in writing and drawing figures. There was characteristic vertical writing and drawing (such as a 'alternative square and triangle' and 'interlocking pentagon') as well as hemispatial neglect.

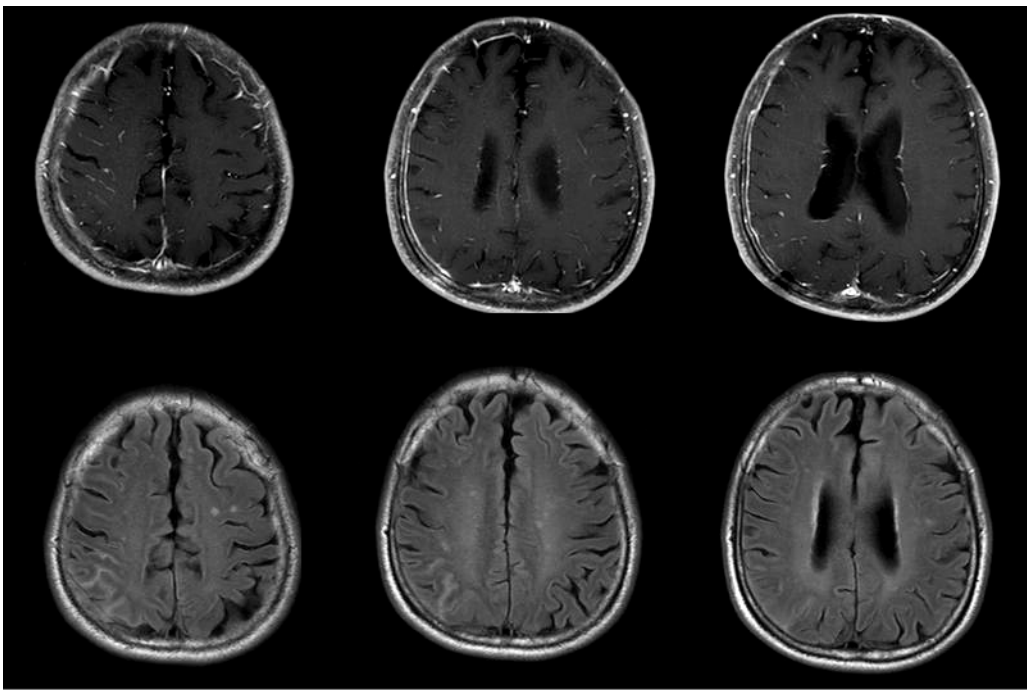

Fig. 2. The brain MRI of the patient. The pachymeningeal enhancement on the right fronto-temporo-parietal area (upper panel: gadolinium enhanced T1-weighted image) was evident and the abnormal high signal intensity was seen along the right parietal cortex (lower panel: FLAIR-weighted image).

intermittent pseudoathetoid movement. He had a positional sensory loss in left 2nd to 5th fingers and cortical sensory loss, but the other sensory modalities were normal.

Gadolinium enhanced T1-weighted MRI revealed diffuse dural thickening along the right fronto-temporo-parietal convexity and FLAIR-weighted imaging revealed localized high signal intensity in the right parietal cortex, suggesting parenchymal edema neighboring hypertrophic dura (Fig. 2). He showed normal CSF pressure (200 mmH_2O) and profiles. He was given oral prednisone 1 mg/kg per day and azathioprine 50 mg twice a day. His symptoms were gradually improved until 6 months from the last symptom onset.

DISCUSSION

Two major criteria for the diagnosis of AHS are 1) the feeling of foreignness; complaint of a foreign limb and 2) involuntary movements; complex, autonomous, involuntary motor activity that is not a part of identifiable movement disorders[3, 6]. At this admission, he had considered that his left hand was silly, uncontrolled and acted unintentionally. Therefore we think these phenomena can be described as an AHS and should be a kind of personification of affected limb and a feeling that his body part is foreign and autonomous.

In a clinical review of 20 reported cases of alien hand syndrome, the authors categorized AHS into two distinctive syndromes that frontal AHS and callosal AHS[7]. They suggested that frontal AHS is associated with reflexive grasping, groping, and compulsive manipulation of tools; and results from the damage to the supplementary motor area, anterior cingulated gyrus, medial frontal cortex of the dominant hemisphere and the anterior corpus callosum. On the other hand, callosal AHS is characterized by inter-manual conflict and an increased tendency for dominant limb exploratory movements coupled with release from an asymmetrically distributed, predominant non-dominant-hemisphere inhibition. Since our case had no corpus callosal lesion unlike their cases, these explanations cannot be applicable. Furthermore, the authors excluded from their analysis the cases associated with corticobasal degeneration which can

reveal repetitive, rhythmic and spasmodic involuntary movements with a tendency to levitate[4] and assume unusual posture. These excluded cases are more similar to our patient in terms of the characteristics of the involuntary movement. Interestingly, some authors suggested that the AHS is not always the expression of a disconnection syndrome and may result from a single strategically located lesion in the absence of involvement of the corpus callosum[8].

Some authors have classified AHS into a "posterior" or "sensory" form and an "anterior" or "motor" form[6]. In a previous study of 15 patients believed to have corticobasal degeneration, they reported that "alien limb" phenomena in addition to cortical sensory loss, apraxia and limb dystonia were characteristic of the disorder[9]. Autonomous movement and personification of the affected extremity which were ego-syntonic (positive feeling) in nature has been considered as characteristic of posterior variant AHS[10]. However our patient was not ego-syntonic in nature, he was some nervous with negative and strange feeling, although posterior-variant AHS has been classically described in corticobasal ganglionic degeneration[9].

The right parietal lesion seen on FLAIR-weighted MRI of our patient, suggesting vasogenic edema[11] along the thickened dural convexity might cause a loss of sensory integration and feedback interfering with motor function and release exploratory movements in the absence of appropriate stimuli. The positional sensory loss in the left hand was moderate, but it was not sufficient by itself enough to explain the foreignness of the hand. An alternative explanation could be the combination of a neglect syndrome[3] and loss of visual guidance of the affected limb (optic ataxia) that gives rise to AHS[12]. However our patient had neither an optic ataxia nor an occipital lesion including the splenium. It remains unknown why some small proportion of patients with a right parietal lesion exhibit an AHS, while most of the patients do not.

Another interesting finding of our patient is vertical writing and drawing with hemispatial neglect. Clearly, before this admission, he always had written letters horizontally. But at this time of admission, he could not write letters and draw figures horizontally any more. Also he could not copy the letters or figures horizontally. With this regards, a previous case study reported about horizontal versus vertical reading in neglect dyslexia[13]. The authors suggested that the neural processing of horizontal reading may be different from that of vertical reading, because the subjects with neglect dyslexia made less error in vertical reading than in horizontal reading. Despite of that, vertical writing of letters and vertical drawing of figures instead of horizontal writing and drawing as part of hemispatial neglect has not been reported yet. We think there are several possibilities that 1) partially compensated hemispatial neglect by unknown mechanism; subsequently he wrote the letters and drew figures vertically 2) the contribution of perseveration - writing syllabic units vertically on and on - "서울르르르" and 3) a rotated or distorted perceptional system as much as 90 degree.

This is a unique case showing posterior form of AHS and conspicuous vertical writing and drawing in that the underlying disease is idiopathic hypertrophic pachymeningitis involving the right parietal cortex with the preservation of the corpus callosum.

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