CASE REPORT

Color picture drawings without form and eye movements: A case report of visual form agnosia in a girl

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Abstract

The case of an 8-year-old girl who manifested cortical blindness and whose color drawings of faces and objects were without outlines is reported. Her birth was uneventful. When she was 10 months old, she fell down to the floor from a chair, resulting in a subarachnoidal hemorrhage. A repeat brain MRI revealed localized lesions in the visual cortices in the right and left hemispheres. As she grew older she was found to have visual imperceptions. She was found to have difficulties in learning visually the names of objects with form and letters, and in recognizing faces of her family. However, she was able to discriminate well the colors of faces and objects and learn easily the names of the objects with form by touching. She seemed to utilize subcortical vision for seeing colors of faces and objects.

Keywords: Cortical blindness, visual cortex, color discrimination, apperceptive visual agnosia

Introduction

It is very rare to encounter pediatric patients with cortical blindness caused by lesions in the bilateral visual cortices. Residual vision in cortical blindness is called visual agnosia and blindsight or subcortical vision. Cortical blindness is classified into apperceptive [1,2], associative [3], and integrative visual agnosia [4]. We had an opportunity to examine an 8-year-old girl with cortical blindness and we report on her residual abilities of vision and eye movements from the viewpoints of neuropsychology and neurotology.

Case report

The patient was an 8-year-old girl and the diagnosis was cortical blindness.

Her birth was uneventful and her growth was normal until she was 10 months old, when she fell down to the floor from a chair, resulting in a subarachnoidal hemorrhage. As she grew older, she was found to have visual imperceptions such as difficulty in learning visually the names of objects with forms or outlines and letters, and in recognizing faces. However, she was able to discriminate the colors of objects and faces and to learn the name of objects by touching and she was able to walk alone without bumping against objects. On the other hand, her development of speech and hearing was normal. She was referred to us from a children's hospital for further neuropsychological and neurotological examinations.

Brain imaging

Brain MRI performed at the age of 8 years revealed that slices of bilateral occipital lobes showed a low T1-weighted signal intensity with enhancement and a high T2-weighted signal with enhancement. These

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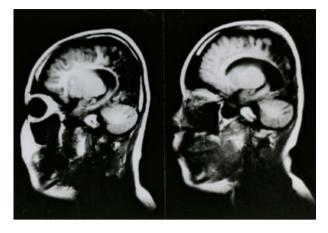


Figure 1. Brain MRI: the left image shows the left saggital slice for the right hemisphere and the right image shows the right saggital slice for the left hemisphere. Lesions in the bilateral visual cortices are detected as low signal intensity areas.

findings indicate severe atrophy of the bilateral occipital lobes and dilatation of the posterior horns in the lateral ventricles (Figure 1). Similar findings were obtained in the posterior frontal lobe and parietal lobe. These MRI findings suggest traumatic damage

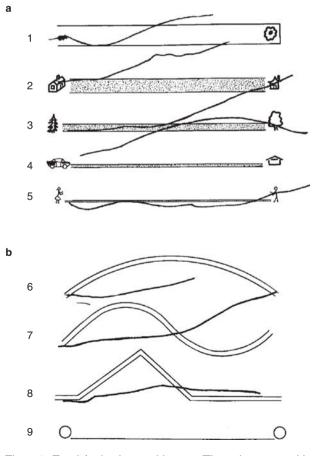


Figure 2. Frostig's visual recognition test. The patient was unable to draw correctly.

Table I.	Visual	cognition	and	touch	cognition	tests.	
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Object no.	Object	Visual cognition	Touch cognition
1	Ball	×	0
2	Cup	×	0
3	Scissors	×	0
4	Toothbrush	×	0
5	Spoon	×	0
6	Coin	×	0
7	Comb	×	0
8	Handkerchief	×	0
9	Fork	×	0
10	Pencil	×	0
11	Ring	×	0
12	Key	×	0

The patient was unable to recognize visually any objects with outlines but was able to recognize well objects by touching. \times , recognition not possible; \bigcirc , recognition correct.

of the bilateral occipital lobes after the accident at the age of 10 months.

Neuropsychological tests

Frostig's visual cognition test revealed that she was unable to draw lines well between parallel straight lines when she was instructed to draw correctly in the middle of two lines (Figure 2).

Table II. Results of color naming test.

Color no.	Color	Result		
	Strong contrast			
1	Red	0		
2	White	0		
3	Black	0		
4	Yellow	0		
5	Purple	0		
6	Blue	0		
7	Orange	0		
8	Pink	0		
	Weak contrast			
1	Brown	×		
2	Gray	×		
3	Light blue	\times (Blue)		
4	Yellow-green	\times (Green)		

The patient was able to discriminate any colors with a strong contrast but not those with a weak contrast. \times , recognition not possible; \bigcirc , recognition correct.

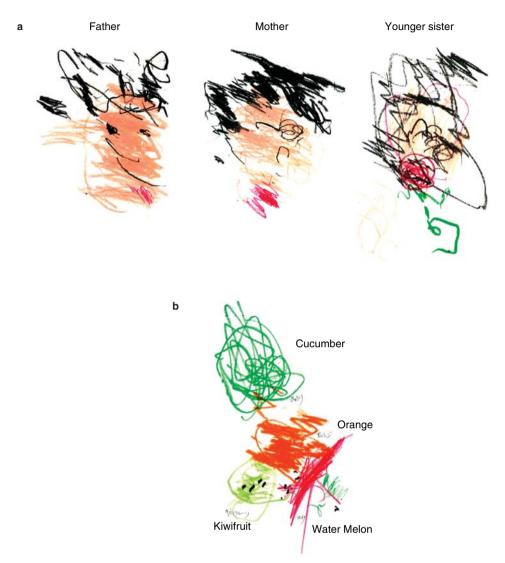


Figure 3. Color drawing test using color crayons. Use of colors is correct but the outlines of the objects are not drawn. (a) Drawings of the patient's father, mother, and younger sister. (b) Drawings of four kinds of fruit.

The visual cognition and touch cognition tests revealed that the patient was unable to recognize visually any objects with outlines and to copy pictures but was able to recognize well objects by touching (Table I).

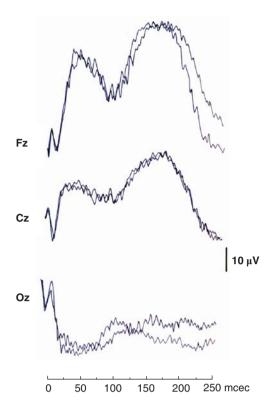
A color naming test revealed that she was able to discriminate any colors with a strong contrast but not those with a weak contrast (Table II).

She was instructed to draw her own face, her father's face, and four kinds of fruit using color crayons. The faces were drawn on the basis of color only without outlines (Figure 3A). The hair and eyes were black, the face was of skin color, and the lips were red. The other face with eye glasses was also drawn with the correct color but without the outlines. For four kinds of fruit, she was instructed to draw a cucumber, an orange, a kiwifruit, and a watermelon. At least, the color of each fruit was correctly painted even without the outlines (Figure 3B). The familiar face recognition test, using color photographs of herself, her parents, her speech pathologist, and her doctor revealed that she was unable to discriminate the faces.

Neurotological study

The flash-stimulated visual evoked responses recorded from Fz, Cz, and Oz showed responses from Fz and Cz but not from Oz. The absence of the late component of Oz is regarded as a result of atrophy of the bilateral occipital lobes (Figure 4) [5].

Her ENG recordings (Figure 5) revealed the following. Figure 5A shows the results of a smooth pursuit test. In this test, a laser spot that was projected on the screen in front of the subject moved clockwise at 0.25 Hz. She was instructed to follow the visual target and her eye movements were recorded by



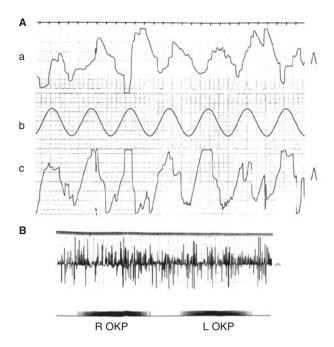


Figure 5. ENG recordings. (A) Smooth pursuit test. (a) Horizontal eye movements, (b) slowly moving target stimulus pattern, (c) vertical eye movements. (B) Eye movements in the optokinetic pattern test (R OKP and L OKP show stimulus patterns).

Figure 4. Flash-stimulated visual evoked responses. Responses at Fz and Cz are well elicited but not at Oz.

ENG. In Figure 5A, the middle recording is for the visual target, the upper recording is for the horizontal eye movements, and the lower recording is for the vertical eye movements. These recordings showed cogwheel patterns. Figure 5B shows the results of the optokinetic pattern test for provocation of optokinetic nystagmus, which demonstrated a very poor pattern. The damped rotation chair test that was performed to examine vestibular ocular reflex showed normal responses on the ENG recordings.

Discussion

The human occipital cortex is usually divided into three areas -17, 18, and 19 - according to Brodman (1909). Area 17, the primary visual cortex, is also known as the striate cortex and has clear-cut borders. Areas 18 and 19 contain multiple areas [6].

Up to the primary visual or striate cortex, the visual pathway represents a serial relay with a clear retinotopic arrangement. Beyond the striate cortex, the projections to extrastriate visual cortex proliferate into complex webs of parallel projections, back projections, and interconnections among a large number of specialized cortical modules. Early visual areas such as Areas 18 and 19 form a peristriate zone surrounding the primary visual cortex and receive direct inputs from the striate cortex. These regions respond differently to form, color, depth, and motion [6].

'Cortical blindness' is visual loss resulting from lesions in the bilateral visual cortices of the occipital lobes and is named visual agnosia, which is classified into apperceptive, associative, and integrative visual agnosia (Table III) [1–4]. However, the residual ability of visual sensation is called blindsight or subcortical vision [7–9].

The most common cause of cortical blindness is cerebral hypoxia or contusions due to a head trauma at the striate and parietal and premotor regions. The lesions in the striate cortex cause visual agnosia. Cerebral hypoxia can be caused by intoxication with carbon monoxide and inflammation such as meningitis, encephalitis, vascular occlusion, and trauma [6].

In this patient's cortical blindness, visual apperceptive agnosia, or form agnosia was caused by head trauma of the bilateral occipital lobes at the age of 10 months. The patient was able to discriminate colors but was unable to recognize visual forms of objects. She was able to draw color pictures with correct colors but without the outlines. However, it is noted that she was able to walk alone without bumping into objects. Her ENG recordings showed abilities of a good smooth pursuit but without optokinetic stimulation by an optokinetic pattern, which suggests that she could visually follow slowly moving objects but not fast-moving objects as in OKN stimulation.

Table III. Classification of cortical blindness.

No.	Reference	Туре	a. Form	b. Color	c. Light	d. Letter	e. Face	F. Copy
1	Benson & Greenberg, 1969 [1]	Apperceptive visual agnosia	×	0	0	×	×	×
2	Ruben & Benson, 1971 [3]	Associative visual agnosia	×	0	0	×	×	0
3	Riddoch & Humphreys, 1987 [4]	Integrative visual agnosia	×	0	0	×	×	

Perception and cognition: \bigcirc , correct; \square , partially correct; \times , not possible.

In this patient, cortical blindness is caused by lesions in the bilateral occipital lobes as demonstrated by brain MRI and visual evoked responses. Pathophysiologically, she was able to recognize color differences but not the outlines of objects. Probably, she has lesions in the striate cortex (Area 17), which were responsible for her not being able to recognize outlines of objects, but not in the other visual system, which enables her to recognize colors of objects and light.

In 1982, Ungerleider and Mishkin suggested a subdivision of the cortical visual system into the dorsal and ventral streams projecting from visual cortical areas: (1) a dorsal stream concerned with the analysis of the locations of objects and their movements in space and (2) a ventral stream responsible for the processing and storage of information about the identity of objects such as shape, color, and other salient physical properties [10–12].

Central visual loss or cortical blindness can occur owing to damage to the bilateral retrochiasmal visual pathways such as mitochondrial encephalopathy, head trauma accidents, and stroke-like episodes [6]. In this case after head trauma, color recognition ability is maintained but visual form recognition ablitiy is lost. Our patient's color drawings of faces and fruit are very unique because the outlines of objects are lost but the colors are correct. Although she was unable to recognize the form of objects visually, she was able to recognize them by touch immediately. This dissociation between visual recognition and touch recognition suggests that the lesions in her bilateral visual areas in the occipital lobe include the striate cortex and ventral or temporal system, as demonstrated by brain MRI. Finally, it is concluded that her cortical blindness is apperceptive visual agnosia or visual form agnosia with preservation of color recognition and loss of form recognition and copy ability.

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