

# Cortical Blindness Following Aortic Arch Surgery

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**Purpose:** To report a patient with Marfan's syndrome in whom cortical blindness occurred after planned circulatory arrest during aortic arch surgery.

**Case:** A 31-year-old man underwent aortic arch surgery because of an acute aortic dissection due to Marfan's syndrome. He noticed poor vision after surgery, although his pupillary reflexes and fundi appeared normal. Magnetic resonance imaging (MRI) and positron emission tomography (PET) were performed 2 years and 9 months after his operation.

**Results:** The MRI revealed cortical atrophy in the occipital cortex, and PET scans with fluorodeoxy glucose revealed extreme glucose hypometabolism in the occipital cortex. The atrophy reflected cortical laminar necrosis that presumably occurred during the planned circulatory arrest to the brain during the surgery.

**Conclusion:** It is occasionally difficult to diagnose cortical blindness with MRI, especially at the acute stage. We could find significant hypometabolism of the occipital cortex using PET. **Jpn J Ophthalmol 2001;45:547-549** © 2001 Japanese Ophthalmological Society

**Key Words:** Cerebral, cortical blindness, glucose metabolism, laminar necrosis, positron emission tomography.

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## Introduction

Cortical blindness is defined as a loss of vision that is caused by bilateral occipital lobe lesions with normal pupillary reflexes and normal fundus. Etiological factors include cerebral infarction, cerebral embolization, aortic arch arrest, and head trauma. Neuroimaging techniques such as computed tomography (CT), magnetic resonance imaging (MRI), or positron emission tomography (PET) are necessary to diagnose cortical blindness, but occasionally MRI does not reveal obvious disturbances. In such cases, PET may be effective in making a diagnosis of the cause of the cortical blindness.

## Case Report

A 31-year-old man who did not have a past history of serious illnesses was hospitalized because of sud-

den chest pain on December 8, 1996. Chest x-ray and CT revealed acute aortic dissection of De Bakey classification type I, and annulo-aortic ectasia related to Marfan's syndrome. The patient's father had undergone surgery for annulo-aortic ectasia. The patient underwent a modified Bentall operation with hemiarch replacement. The total anesthetic time was 19 hours. Two general circulatory arrests of 1 minute and 11 minutes were required, and retrograde cerebral perfusion (RCP) was also performed twice (40 minutes and 72 minutes). In the latter procedures, oxygenated blood is back-infused into the femoral artery instead of the ascending aorta so that the brain and other critical organs can be perfused. Lowest body temperature was 18°C during surgery.

The patient regained consciousness 10 days after surgery on December 18, 1996. His family noticed that he did not fixate steadily on objects or direct his gaze at anything. No ocular abnormalities were observed when he was examined by an ophthalmologist. Magnetic resonance imaging of the head was performed but no apparent abnormality was found. He had had poor vision for more than 2 years with-

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out a reasonable explanation, before visiting us to obtain a second ophthalmological opinion on August 19, 1999. It was 2 years and 9 months after his surgery.

His corrected visual acuity was 20/500 in the right and 20/600 in the left eye. He had normal pupillary reaction to light and had almost normal fundus appearance. Goldmann perimetry revealed generalized depression with preservation of some central visual field detected by V-4 target indicating a bilateral hemianopia with poor fixation. Magnetic resonance imaging and PET scans were prescribed to determine the cause of his decreased vision.

### Results

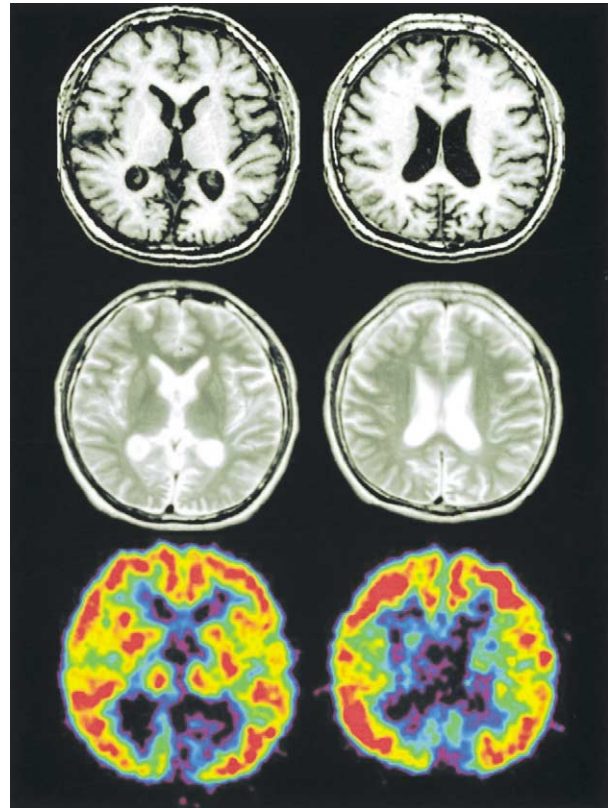
Magnetic resonance imaging revealed occipital cortex atrophy that extended over the entire occipital cortex, but there was no atrophy or infarction in other regions of his brain. The PET scan with fluorodeoxy glucose revealed extreme glucose hypometabolism in the occipital cortex and speckled hypometabolism in other areas of the entire cerebrum (Figure 1).

### Discussion

Based on the history of a possible hypoxic incident and the presence of atrophy and extremely low metabolism, there was a high probability of cortical laminar necrosis in the occipital cortex of this patient. Cortical laminar necrosis is a pathological change characterized by atrophic change of gray matter and is often caused by a hypoxic attack or hypoglycemia. Our findings led to the conclusion that the blindness in this patient was caused by cortical laminar necrosis of the occipital cortex.

During his aortic arch surgery, there were two planned circulatory arrests (one minute and 11 minutes) and RCP twice (40 minutes and 72 minutes). There is a possibility that the circulatory arrest and/or RCP led to the cortical laminar necrosis in his occipital cortex. There is also a possibility that dissection of the aorta caused occlusion or obstruction in his common carotid arteries and interfered with the blood flow of his brain. However, his consciousness was clear just before the operation, so there is less possibility of this. Microembolism of brain vessels by air or debris may have caused laminar necrosis during the surgery.

Retrograde cerebral perfusion is a popular method of cerebral perfusion and is often used during heart surgery, although the amount of cerebral perfusion is reduced. It is recommended that circula-



**Figure 1.** T1-weighted magnetic resonance imaging (MRI) (top), T2-weighted MRI (middle), fluorodeoxy glucose-positron emission tomography (PET) image (bottom). MRI: There is slight cortical atrophy and loss of gray matter in occipital cortex. PET image: There is extreme glucose hypometabolism in occipital cortex. Hypometabolism extends over the whole occipital cortex including primary and associated visual cortex.

tory arrest time should be less than 45 minutes and that the RCP time should be less than 80 minutes. Sato et al<sup>1</sup> reported that two patients suffered brain damage following RCP times of 101 and 130 minutes. Aomi et al<sup>2</sup> reported that a RCP time of longer than 80 minutes induced transient consciousness disturbance in 57% of their patients and proposed that the safe time limit was 80 minutes or less. If the duration of RCP exceeds this limit of 80 minutes ischemic lesions in the brain might be generated. The safe time limit should be shortened depending on the condition of the patient. The safe number of times for RCP is usually once during an operation, and if there is a second RCP there is a good possibility that the safe time limit should be shortened. In the present case, it is likely that the second RCP caused the cortical laminar necrosis in the occipital cortex leading to the cortical blindness. However,

laminar necrosis restricted to the visual cortex has not been reported frequently.

Cortical laminar necrosis occurs as a consequence of oxygen or glucose depletion, as in anoxia, hypoglycemia, status epilepticus, or ischaemic stroke. Takahashi et al<sup>3</sup> described the natural course and MRI findings as follows. In the acute stage, cerebral edema is seen, and in the early subacute stage, enhanced T1-weighted images show cortical laminar enhancement. Unenhanced T1-weighted images reveal characteristic laminar hyperintense lesions of the cerebral cortex in the late subacute stage. In the chronic stage, cortical atrophy and delayed but progressive white matter changes take place.<sup>3</sup>

The cases of cortical laminar necrosis reported in the last 10 years are summarized in Table 1. Laminar necroses after cerebral infarction, cardiac arrest, or respiratory arrest are relatively common. Komiyama reported that significant numbers of cortical laminar necrosis cases were found in the region of cerebral infarctions.<sup>4</sup> Ten viral encephalopathy-related cases in children have been reported.<sup>5</sup> Other causes include central nerve system lupus, immunosuppressive therapy, and radiation.

We studied the areas of laminar necrosis in the reports presented in the last 10 years. Laminar necroses generated after whole brain exposure to hypoxia or hypoglycemia are included, but localized infarction, degenerative diseases, and idiopathic diseases are excluded. Thirteen cases of laminar necroses

were reported to have occurred in the whole cerebrum. One case was only in the frontal lobe. Laminar necroses have been reported in many areas of the brain and no predominance in the visual cortex has been observed. As far as we have searched, no case has been reported in which the occipital cortex was selectively affected with laminar necrosis and resulted in cortical blindness without any other neurological deficits. These past reports suggest that the occipital cortex is not especially predisposed to laminar necrosis when the whole brain is exposed to hypoxia or hypoglycemia. Although we suspect that this patient is a rare case, we wish to alert cardiac surgeons and ophthalmologists to the fact that laminar necrosis can occur in the occipital cortex following aortic arch surgery and can lead to cortical blindness. In such cases, significant hypometabolism of the occipital cortex can be detected using PET.

## Conclusion

We report a case of cortical blindness due to laminar necrosis in the occipital cortex. Because it is difficult to diagnose this decreased vision as cortical blindness, especially at the acute stage, we recommend PET scans to study such cerebral dysfunction.

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**Table 1.** Causes of Cortical Laminar Necrosis (1991–2000)

Cerebrovascular disease	30
Encephalopathy	10
Respiratory arrest	5
Cardiac arrest	3
Hypoglycemia	2
Others	13
Total	63