Dide and Botcazo Syndrome—Syndrome of Bilateral Obstruction of Posterior Cerebral Arteries

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INTRODUCTION

Cortical blindness resulting from a bilateral occlusion of the posterior cerebral arteries is rare. Diagnosis itself sometimes is difficult, especially when some pupillary abnormalities are present. But if a certain constellation of symptoms accompanies the blindness, involvement of the territory of the bilateral posterior cerebral arteries becomes very probable, making the diagnosis of cortical blindness much easier. This symptom cluster has long been known as the Dide and Botcazo syndrome in French literature. An example of the syndrome will be presented.

The syndrome consists of (1) cortical blindness or cortical visual impairment, (2) spatial disorientation and (3) defect of memorizing.^{3 8}

A CASE REPORT

A 57-year-old man suddenly developed a generalized convulsive seizure with loss of consciousness during the night in September, 1975. He regained consciousness in about 15 minutes and left no neurological symptoms. He started having the same kind of attack about once a month. In January,

1976, while walking on the street, he tended to deviate to the right, which disappeared quickly. On the 10th of April, he experienced a fogginess of vision for a short duration. On the morning of April 13, headache and nausea started and around noon he suddenly became totally blind and was admitted to the Department of Ophthalmology, Kobe University Hospital, on the next day. By then he could detect hand motion bilaterally. The fundi were normal. Extraocular movement was full. Pupils were somewhat dilated and reaction to light was slow and incomplete. Visual field could not be measured. No other neurological signs were present. No abnormal mental symptoms were noted either. Lumbar puncture was performed with normal results. The patient was started on steroids with a diagnosis of retrobulbar neuritis. On the 22nd of April, he developed a generalized convulsion, from which he never fully recovered and remained confused.

Summary of Clinical State After the 22nd of April

The patient showed gradual recovery throughout his hospital stay but the essential symptoms remained unchanged.

The general physical status was within normal limits. The neurological examination was performed by one of us (A.Y.).

Received for publication Apr. 14, 1977.

The pupils were dilated and reacted to light slowly and incompletely. Fundi were normal. The vision was difficult to determine because of his mental status but was thought to be about light sense. Other cranial nerves were intact. The left knee jerk was brisker than the right. The plantar reflexes were extenser, bilaterally. There were positive grasp reflex of the left hand, positive snout and sucking reflexes. Muscle tone was slightly elevated, bilaterally. Muscle power was thought to be normal. There was no sensory change.

The psychological status of the patient was characteristic. He was awake and cooperative. He was disoriented to time and place. He was not able to tell day from night. He had no idea where he was and was totally at a loss as to the location of a toilet or his room. Even in his own room he could not orient himself correctly. He responded well to immediate tasks and questions but was not able to retain new information for long. Thus, he never learned the name of the hospital. He enjoyed visitors but forgot the visit next day. He also showed retrograde amnesia and could not recall his recent activities before he was admitted to the hospital. He thought he was in his home town which was far from Kobe. However, he was able to remember events in his younger days correctly. His language was essentially normal except for mild naming difficulties when objects were presented to him by hand.

Most remarkably, he was unaware of his blindness and was completely indifferent to the daily inconveniences brought by the defect. Occasionally he would complain that his eyesight was not as good as they used to be, but maintained that he could see well enough all the same.

Laboratory Examinations

Electroencephalogram—Right after the attack, an EEG was taken which showed bilateral 4 to 6 Hz. slow wave activities diffusely and continuously.



Fig. 1: Anteroposterior projection of a left brachial arteriogram. Both posterior cerebral arteries are occluded at the distal end of their ambient portion (arrows).

Angiographical study—A left retrograde brachial arteriography was performed by a neurosurgeon (N.T.). The posterior cerebral arteries were occluded at the distal end of its ambient portion, bilaterally. Calcarine, parietooccipital, posterior temporal and posterior choroidal branches were not visualized (Figs. 1 and 2). A left carotid arteriogram was also taken and was normal.

CT-scan study—About two months after the onset, CT-scan was performed. By contrast enhancement infarcted areas were clearly delineated. The medial portion of the posterior half of the temporal lobe and the occipital lobe was destructed, bilaterally. The lesions were consistent with the territory of the posterior cerebral arteries (Fig. 3).

Visual evoked potential study-No potential was evoked on the occipital lobe by



Fig. 2: Lateral projection of the same arteriogram. Point of occlusion is clearly seen (arrow).

visual stimuli.

DISCUSSION

The core symptoms of the present case include (1) cortical blindness, (2) spatial disorientation, (3) loss of recent memory, (4) retrograde amnesia and (5) unawareness of blindness. Of these five symptoms, combination of cortical blindness and unawareness of the defect has been well known since the publication of the classic paper by G. Anton in 1899.¹ Association of cortical blindness, spatial disorientation and amnesia (recent memory loss coupled with retrograde amnesia) was described three years later by Dide and Botcazo.⁸ The paper remained unnoticed in the English literature. Their patient showed bilateral hemianopsia with



Fig. 3: CT-Scan with contrast enhancement. At this level high-density area surrounded by low-density rim is clearly seen in the medial aspect of the occipital lobes.

preservation of central vision, had difficulty memorizing new materials, had good memory of his old occupation and completely lost topographical notion. Necropsy revealed old infarcts of the bilateral occipital lobes involving both lingual lobes and the left calcarine cortex. This so-called "Dide and Botcazo syndrome" was recently discussed in detail.^{3 5 14} Another variety of association of cortical blindness and amnesia was published lately.⁴ Most other papers on cortical blindness lack specification of mental symptoms.

Cortical Blindness

Cortical blindness is caused by a bilateral destruction of striate cortices or optic radiation underlying the cortices. Blindness may be complete but in cases of bilateral hemianopsia central vision may be preserved. Among various etiologies, occlusive vascular lesions are the commonest. Autopsy verified bilateral occipital lobe infarctions were studied by many authors.^{11 20} However, if we limit our attention to the status of posterior cerebral arteries that are responsible for those infarctions, descriptions become less. We could find that only four cases of autopsy verified bilateral occlusions of these vessels.^{3 20} Like ours there were also angiographically confirmed cases.13 14 18 Embolism has been held as a cause of simultaneous obstruction. Sources of these emboli may be cardiac or mural thrombus of the vertebral or basilar arteries. Anterograde extension of thrombus from the basilar artery was also demonstrated by Castaigne and others but their cases lack clinicopathological correlation.⁶ We don't have an etiological explanation as to the source of the probable emboli in the present case. The cardiac system did not show any abnormalities.

Mention should be made as to the state of the pupillary finding in the present case. Dilated or slowly reactive pupils in cortical blindness were described only occasionally. Among cases collected by Symonds and Mackenzie, one case had dilated pupils which were immobile, one had dilated but reactive pupils, and the other had mediumsized pupils with slow and incomplete light reaction.²⁰ There were other sporadic mentions.^{13 17 19} The mechanism is unclear but it is very likely that a small lesion in the midbrain is responsible for this type of atypical pupillary signs.

Spatial Disorientation

The present case was characterized by prominent difficulty in orienting himself in his immediate environment. Anton's case had the same type of difficulty.¹ This should be distinguished from the amnesic syndrome because in the latter spatial difficulty to his direct surroundings is not usually very conspicuous. This topographical difficulty may be due to the right parietooccipital involvement.⁵ ¹⁰ This aspect has been stressed only by the French authors.^{3 5 8 14}

Amnesic Syndrome

The present patient could attend to immediate verbal stimuli very well. He conversed But he could not remember new well. materials at all. He also showed extensive retrograde amnesia. This defect of memorizing has been well known to occur as an axis symptom of Korsakoff's psychosis with lesions of bilateral mammillary bodies and medial dorsal nucei of the thalamus.²¹ It is also well established that bilateral hippocampal involvement interferes with retention of new information.7 15 Some authors postulate the so-called Papez circuit of hippocampo-mammillo-thalamo-cingulate system as an anatomical substrate of memorizing.² ¹² Bilateral fimbriae were destroyed in one case.³ The most part of the bilateral hippocampi were involved in another.⁴ Our case showed bilateral medial temporal lobe involvement as detected by CT-scan. The amnesic syndrome, which accompanies some cases of cortical blindness, can thus be explained by lesions of the hippocampi or issuing fibers from them within the territory of the posterior cerebral arteries.

Unawareness of Blindness

As is well known, Anton was the first who described this peculiar attitude of a patient to his own defect. The patient was "unaware" of his blindness. This concurrence of blindness and anosognosia for it has since been much emphasized. Our patient was also indifferent to the defect and behaved as if he could see. But it is of interest to note that this "blindness to his blindness"¹ was not present at first in the present case. It became manifest only after confusion and other neurological signs developed. This indicates that lesions which produce cortical blindness are different from those which produce unawareness of the blindness. On the other hand, some authors postulated Korsakoff's psychosis as a basis of this type of indifference toward his own defect.16 However, this cannot be supported either, since some patients who were blind and amnesic seem to have had no Anton's syndrome.^{3 4 8 14} Thus the relationship between cortical blindness and unawareness of the defect is still open to question. We are inclined to support Gassel's statement that anosognosia is more likely to occur with mental clouding and confusion and frequently the awareness fluctuates with the levels of consciousness.⁹

This constellation of symptoms except for unawareness of blindness occurs by lesions of the portion of the cerebrum nourished by the posterior cerebral arteries on both sides. Thus it can duly be called "the syndrome of bilateral occlusion of posterior cerebral arteries" as has been proposed.³

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