ACTA MEDICA SALINIANA

CASE REPORTS

CASE REPORT: ANTON'S SYNDROME DUE TO ISCHEMIC CEREBROVASCULAR DISEASE

Ajla Muratović, Mirjana Vidović

ISSN 0350-364X

DOI: 10.5457/628

Ajla Muratović, Mirjana Vidović

ABSTRACT

Anton's syndrome (AS) describes the condition in which patient deny their blindness despite objective evidence of visual loss, and moreover confabulate to support their stance. It is a rare extension of cortical blindness in which, in addition to the injury to the occipital cortex, other cortical centres are also affected, with patients typically behaving as if they were sighted. This case presents 76-year-old patient with verified cortical blindness as a result of a stroke. The patient persistently denied vision loss. At the same time, she confabulated to see people and objects in her environment clearly. This syndrome may be unrecognized in routine neurological practice. A suspicion of cortical blindness and AS should be considered in patients with atypical visual loss and evidence of occipital lobe injury. Cerebrovascular disease is the most common cause of AS, as in our patient. Recovery of visual function will depend on the underlying aetiology, with cases due to occipital lobe infarction after cerebrovascular events being less likely to result in complete recovery. Management in these circumstances should focus on secondary prevention and rehabilitation.

Key words: Anton's syndrome, visual anosognosia, cortical blindness

INTRODUCTION

Visual anosognosia, that is, denial of loss of vision, associated with confabulation in the setting of obvious visual loss and cortical blindness is known as Anton's syndrome. Although the anterior visual tracts are intact, the visual association centres in the occipital cortex maybe compromised. Patients with Anton's syndrome strongly believe they can see what they cannot and behave and talk as though they were sighted. Attention to the possibility of the condition is, however, drawn when they walk into walls, fall over furniture and describe objects that are not present [1].

The French renaissance writer Montaigne (1533-1592) described in his second book of Les Essais the case of a nobleman who did not believe he was blind despite the obvious signs [2]. This was probably the first ever description of not perceiving one's own blindness in the absence of psychiatric illness or underlying cognitive impairment. A few hundred years later the Austrian neuropsychiatrist Gabriel Anton (1858-1933) described patients with objective blindness and deafness who showed a lack of selfperception of their deficits. He associated these with brain pathology [3]. Joseph François Babinski (1857-1932) later used the term anosognosia to describe the unawareness of the deficit in patients with hemiplegia [4].

We describe a case of a patient with Anton's syndrome and its associated features.

CASE PRESENTATION

A 76-year-old woman was admitted to the admission ambulance of Neurology Department of University Clinical Center in Tuzla, due to a sudden visual loss after examination by an ophthalmologist. Previous medical history revealed long standing diabetes, permanent atrial fibrillation and multiple ishemic strokes. The first stroke occurred in 1996, with consequence of right homonymus hemianopsia. She used anticoagulant therapy in regular treatment. In October 2016 and April 2017 our patient underwent surgical treatment of high-leg amputation of both legs due to femoral artery occlusion. On admission she was awake and oriented, normotensive, eupnoic, with slightly turned head to the right side and sudden visual loss. Ophthalmologic exam confirmed a severe vision loss. The patient sustained that she was able to see, despite the objective evidence of vision loss. Ocular movements, as well as photo motor

Affiliations:

Received:
06.06.2022.

Accepted:
15.11.2022.

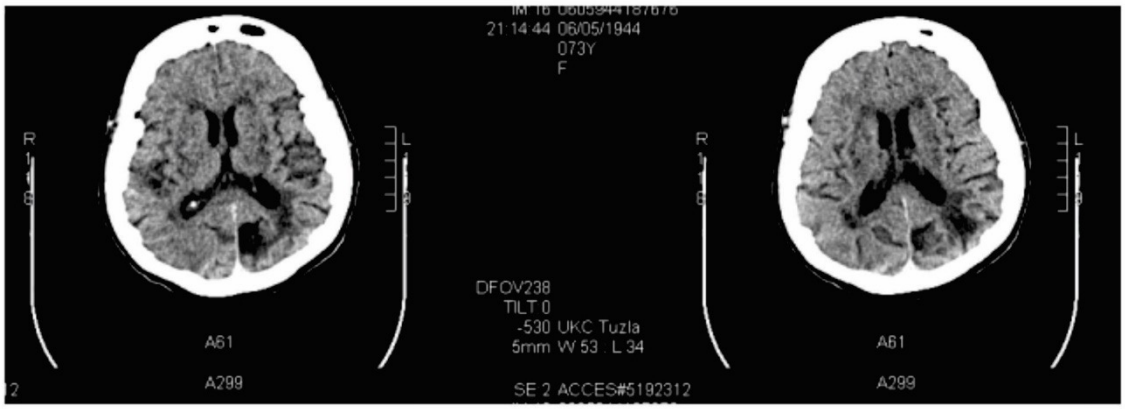
Corresponding author:
Ajla Muratović
Email: muratovic.ajlaa@gmail.com

Funding: none

Competing interests: none

reflex, were preserved. Fundoscopic examination revealed changes secondary to diabetes, inhomogeneous lens blur, non-proliferative diabetic retinopathy, condition after laser photocoagulation which was done in 1994 on both eyes, and subatrophy of papilla of the optic nerve in both eyes. Eye ultrasound showed the glassiness in the posterior corpus. The heteroanamnestic data were provided by the daughter and husband who take care of her. The night before admission, patient complained of blurred vision. When she got up, her husband noticed that she had difficulty taking the cup from the table. She complained that her table was far away and asked him to pass the cup into her hand. When she tried to drink coffee, she spilled it several times. She told her husband that she was sleepy and not in the mood to drink coffee. He noticed that she was behaving strangely compared to everyday life. During the day, she was sleepy. They measured her body temperature with a mercury thermometer, which was 38 degrees Celsius. An antipyretic was prescribed, to which she did not react. The measured blood sugar value was 13 mmol/L. She slept for 2 hours, and when she got up they offered her lunch which she refused, saying she was not hungry. She normally eats on her own, but that day they had to fed her. Furthermore, her family reported that she went occasionally agitated after the last stroke. She didn't vomit, nor was unconscious. She didn't complain of having a headache or visual impairment. She denied having difficulties of other organic systems. The dominant outburst in the neurological finding was vision loss. The patient was not aware of the sight loss. In particular, the sight loss was observed for the first time when the patient asked the doctor to turn the lights on, because the room was dark. Also when asked to describe the room, the patient provided a completely wrong visual description of the interior of examination room. In addition, she was unable to reach doctor's extended hand. During the examination, the doctor turned the lights on and off, asking the patient to say when it was on and when it was off. The patient speculated, hesitantly giving the answers. Despite this obvious blindness, the patient suffered a visual anisognosia, since she was unaware of her blindness and was confabulating about her surroundings when asked about it. Complete blindness was confirmed by ophthalmologist due to ischemic cerebrovascular disease.

Urgent computerised tomography (CT) of the brain revealed: In native axial brain scans, the left temporo-parieto-occipital lobe shows extensive low-density lesion which corresponds to chronic ischemia in the left posterior cerebral artery zone. In the white mass bilateral frontoparietal describes extensive confluent lesion in terms of atherosclerotic encephalopathy with isolated focal microischemic lesions. Multiple hypodense lesions of liquor which corresponds to lacunar ischemia in both basal ganglia. Mutually in the pons several similar lesions, with one lesion in the right hemisphere of the cerebellum with a diameter of 6 mm which corresponds to postischemic sequela. Endocranially, no visible intracranial or extracranial blood collections. CT findings of the ventricular system and extracerebral fluid spaces are neat. Bone hyperostosis reduces the skull. Visible calcified both vertebral arteries **[Figure 1].**



The drugs treatment, together with physical therapy, had no improvement of reduction in neurologic deficit. However, at the time of discharge, persistent elements of Anton's syndrome became more noticeable over time. Blindness remained permanent. Now patient has no orientation clue, occasionally agitated. She suffers from lack of perception of time and space. Family reports that she can often be upset because she spills food and fluids while eating. She refuses to be fed, because she states that she can do it on her one. When someone from the family enters the room, she gives compliments about clothes, talks about the weather outside, praises the hairstyle of the host of her once favourite TV show. Before she lost her sight, she always wore a watch on her hand. When the housemates talk to each other and ask about time, she always looks at the clock and answers. She needs a 24-hour care.

DISCUSSION

Cortical blindness matches the following clinical criteria [5]: a) loss of all visual sensations, including the perception of light and dark; b) loss of menace reflex; c) preservation of light and accommodation pupillary reflexes; d) a normal fundoscopic examination, and e) preservation of ocular movements. Visual anosog-

nosia, or Anton-Babinski syndrome (more frequently known as Anton's blindness), is a rare complication of cortical blindness, where the patients deny their visual deficit [6]. These patients also have damage on visual association cortex, causing the loss of the concept of vision and the awareness of their deficit [7,9].

Theories that try to explain the unawareness of deficit on the Anton-Babinski syndrome describe disconnection phenomena. The first theory describes a conscious awareness system (CAS) located on the parietal lobes, which monitors all the information received from the senses. This system connects with other, located on the frontal lobes, which integrates the information, in order to perform complex cognitive tasks. In Anton-Babinski syndrome, damage of association pathways between visual cortex and CAS would be responsible for the lack of awareness of the visual deficit [10]. Furthermore, the disconnection of the visual areas from other, such as language areas, makes the patients unable to describe the visual stimuli, and, because of that, fabulate an answer.

In addition the disconnection phenomena, other neuropsychologic mechanisms have been described, such as the signal transmission to the visual monitor (located on the visual association cortex) from a secondary visual system, located on the superior colliculus, pulvinar and temporoparietal regions [9]. In the absence of transmission on the geniculocalcarine pathway, this secondary visual pathway would allow blind patients to fabulate. Ischemic cerebrovascular disease, as a cause of cortical blindness, is the most common etiology of Anton-Babinski syndrome [8,11]. Other diseases described as causes are MELAS (Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes)[12], preeclampsia [10], obstetric hemorrhage [13], trauma [14], adrenloeucodistrophy [15], hypertensive encephalopathy [16] and angiographic procedures [17].

Neurological visual impairment, in which the visual disturbance is as a result of brain abnormality or damage rather than eye abnormalities, encompasses a broad spectrum of conditions. These include conditions such as cerebral visual impairment, visual neglect, visual agnosia, various visual perceptual disorders, homonymous hemianopia, lack of facial recognition, delayed visual development and cortical blindness. In patients with total cortical blindness secondary to bilateral damage to the occipital cortices, movement of objects may nonetheless be perceived, either consciously [18] (Riddoch's syndrome) or unconsciously (blindsight) [5]. Conversely, motion blindness, in which patients can see objects but cannot perceive the motion of these objects, has also been well described [19]. This may be accounted for by the presence of projections from the lateral geniculate nucleus, both to the visual cortex (V1) via the optic radiations and to the motion-selective middle temporal area (MT or V5), a cortical area not previously considered 'primary' [20]. Other manifestations compaired visual acuity may include Charles Bonnet syndrome, in which patients with visual loss from any cause may experience hallucinations, often very elaborate, with images of unfamiliar people or buildings, and so on, although with preservation of insight [21].

Anton's syndrome is the denial of loss of vision (visual anosognosia) associated with confabulation in the setting of obvious visual loss and cortical blindness. Frequently, patients with damage to the occipital lobes bilaterally also have damage to their visual association cortex, which may account for their lack of awareness [22]. Additionally, as suggested by Anton, damaged visual areas are effectively disconnected from functioning areas, such as speech language areas. In the absence of input, functioning speech areas often confabulate a response [9].

CONCLUSION

AS may be unrecognized in routine neurological practice. A suspicion of cortical blindness and AS should be considered in patients with atypical visual loss and evidence of occipital lobe injury. Cerebrovascular disease is the most common cause of AS, as in our patient. Recovery of visual function will depend on the underlying aetiology, with cases due to occipital lobe infarction after cerebrovascular events being less likely to result in complete recovery. Management in these circumstances should focus on secondary prevention and rehabilitation.

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