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CASE REPORT

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Bilateral occipital lobe infarct neglect deficit (BLIND) syndrome

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ABSTRACT

Cortical blindness is characterized by loss of vision due to dysfunction of the visual cortices, most commonly secondary to bilateral ischemic infarcts of the occipital lobe. Other causes include surgery such as aortic valve replacement, laryngeal surgery, craniotomy, cerebral angiography, head trauma, and partial seizures. Visual anosognosia is a distinct feature of cortical blindness, wherein patients claim they can see and confabulate visual perceptions, despite loss of sight. We herewith present a rare phenomenon known as Anton Syndrome, an eponym named after the Austrian neurologist and psychiatrist, Gabriel Anton (1858–1933). There are a limited number of cases of Anton's Syndrome in the literature, with only 28 case reports published from 1965–2016. Although he was bestowed a neurologic eponym, Anton was an advocate of eugenics and racial hygiene. He publicly advocated for 'superior breeding' and 'selection' in order to 'build a brave and noble race.' We therefore propose replacing the eponym with Bilateral Occipital Lobe Infarct Neglect Deficit (BLIND) Syndrome, with intention of raising awareness of this unique presentation as well as of the widespread interest in eugenics in the early 1900s amongst physicians, notably Gabriel Anton.

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Anton syndrome; cortical blindness; visual anosognosia; blind syndrome; atrial tachyarrhythmia

1. Case presentation

A 76-year-old male was driving his car when he suddenly experienced bilateral blurred vision [1–4]. Unable to use his car or phone properly due to the abrupt onset of his visual deficits, the patient reportedly remained in his car for three days. He was found by police, who brought him to the emergency department. On initial evaluation, the patient reported that he had been driving to visit his daughter in another state when he suddenly needed to pull over to the side of the road. He then recalled, 'I was found by a priest who invited me into his home, took me in, and fed me a nice meal. The priest then called the ambulance to pick me up.' The patient reported several weeks of increasingly frequent 'lightening-like' bright flashes of green and yellow in his right eye, but denied the presence of floaters, curtain loss of vision, or diplopia. He reported that his visual acuity was 20/30 bilaterally at his last complete ophthalmology examination one year prior to presentation.

Physical examination revealed an irregularly irregular rhythm and tachycardia. EKG was remarkable for atrial flutter with 2 to 1 block and heart rate of 150 beats per minute (Figure 1). Ophthalmology evaluation demonstrated significantly diminished visual acuity (< 20/200 bilaterally with 360° constriction of confrontational visual fields), intact pupillary reflexes and ocular movements bilaterally, and no evidence of retinal, macular or choroidal pathology on dilated fundus examination. The patient was deemed cortically blind, prompting further evaluation with computerized tomography (CT) and magnetic resonance imaging (MRI) of the head. CT brain without contrast demonstrated findings compatible with bilateral subacute parieto-occipital infarcts with local sulcal effacement, without mass effect or hemorrhagic transformation (Figure 2). Magnetic resonance imaging (MRI) of the brain without contrast showed acute to subacute bilateral posterior cerebral artery territory infarcts with associated mild petechial hemorrhage and additional small subacute infarcts in the bilateral cerebellar hemispheres. MR angiogram of the head and neck without contrast showed no evidence of intracranial occlusion, stenosis, or aneurysm suggesting evidence of symmetric bilateral infarcts may be cardioembolic in nature in the setting of new onset atrial tachyarrhythmia. The patient also demonstrated signs of visual anosognosia: he occasionally reported seeing words/colors on the television in his room while it was turned off, and he remarked that he was able to recognize the facial features of the primary medicine intern who visited following him daily on the medicine floors, despite overt blindness.

2. Discussion

Anton's Syndrome has a distinct presentation, in which a patient is in denial of loss of vision, often confabulating visual perceptions in the setting of cortical blindness [1]. This phenomenon is referred to as

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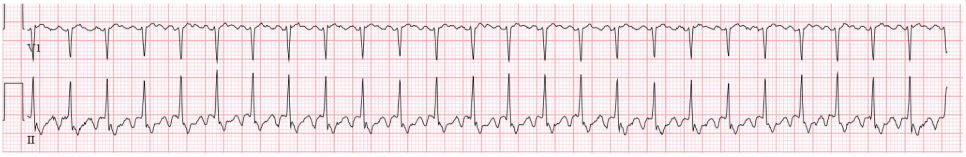

Figure 1. EKG was remarkable for atrial flutter with 2 to 1 block and heart rate of 150 beats per minute.

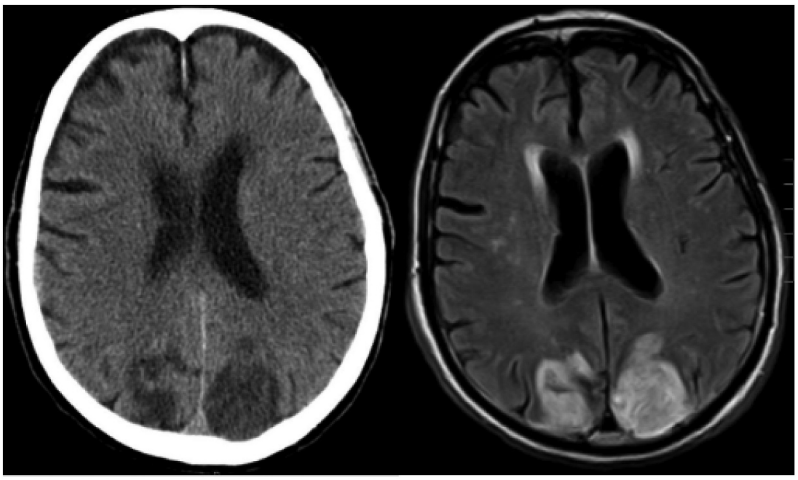

Figure 2. CT brain without contrast revealed bilateral subacute parieto-occipital infarcts (left). MRI of the brain without contrast on FLAIR sequence showed acute to subacute bilateral PCA territory infarcts (right).

visual anosognosia. Our patient presented with the classic description of Anton Syndrome. His imaging results are consistent with cortical blindness, likely cardioembolic in the setting of new onset atrial tachyarrhythmia. Paradoxically, the patient still perceived that his vision was preserved throughout his hospital course. Additionally, the initial presentation in which the patient described meeting a priest who picked him up and called the ambulance leads us to believe he may have been confabulating in the setting of his acute bilateral occipital infarcts.

Aldrich et al. notes that cortical blindness can be caused by multiple etiologies, including cerebrovascular infarct, surgery such as coronary artery bypass, aortic valve replacement, laryngeal surgery, craniotomy and cerebral angiography [1]. Mechanisms of cerebral dysfunction during surgery or angiography are usually described as an event of anoxia, hypoperfusion, or hemorrhage leading to infarction of the cerebral tissue [1]. Post-traumatic contusions, progressive multifocal leukoencephalopathy, adrenoleukodystrophy and seizures are nonvascular etiologies of cortical blindness that can present with findings consistent with Anton Syndrome [2]. For management, it is essential to distinguish vascular from nonvascular etiologies in patients with suspected Anton Syndrome. Vascular causes usually imply poor prognosis, while visual anosognosia and vision deficits due to nonvascular causes may improve and sometimes resolve completely [1,2,5].

Bilateral cortical blindness is most commonly due to vascular insufficiency of the region served by the distal branches off the posterior cerebral artery [5,6]. There are several theories to support the mechanism of visual anosognosia experienced in patients with cortical blindness. Anton postulated that damaged visual areas are effectively disconnected from functioning areas. Functioning speech areas produce confabulated responses due to the absence of input from disconnected speech-language regions [7,8]. Another neuropsychological hypothesis refers to destruction of the sensory monitors wherein images are interpreted incorrectly [7,8]. Presence of false feedback from another visual system has also been suggested. In the absence of visual input, false internal imagery from the superior colliculus, pulvinar and temporoparietal regions may transmit signals and convince speech areas to produce a confabulated response [7,8].

The unusual phenomenon of Anton Syndrome was first referenced more than 2,000 years ago when the stoic philosopher Seneca (c. 4 BC–AD 65) described the phenomenon in a letter to Lucilius,'This woman suddenly lost her sight … Incredible as it may appear … she does not know she is blind.'[9]. The French writer Montaigne (1533–1592) documented the description of a patient who portrayed unawareness of his blindness [10]. Three centuries later, neuropsychiatrist Gabriel Anton (1858–1933) described a cohort of patients with

blindness and deafness who displayed awareness deficits in association with their brain pathology [11]. The term 'anosognosia" was later conceived by Joseph François Babinski (1857–1932) to describe this unusual symptomatology [12].

Although he made contributions to medicine, Anton believed firmly in eugenics. Politically, Anton was described by his peers as a 'true German', and advocated for restoration and welfare of the German race [4]. He believed in 'selection' and 'superior breeding', in hopes of building a brave and noble race [11]. Similar to many scientists of his time, Anton believed that racial hygiene was the scientific approach to improving society [11,13]. In 1925, he gave a detailed speech demonstrating his beliefs in eugenics: 'In light of present knowledge, it is no longer reasonable to passively follow the hereditary decline of entire families and the hereditary inferiority and diseases as in an ancient Greek tragedy. [...] The successful individual's frequent performance at the highest level demands a mental concentration, a sacrifice also in a physical sense, which is often detrimental to the activities of reproduction. [The result is] a constant self-eradication of the successful individual. [We have to] take precautions in order to protect and improve [...] the quality of the race. [...] It should be our directive that [the physician's] service to the race is also the service to the individual and vice versa.' [11,14] Physicians beyond the German-speaking countries openly advocated for racial hygiene. Other racial hygienists honored by neurological eponyms existed such as Heinrich Lundborg (1868–1943; Unverricht-Lundborg disease) in Sweden and William Gordon Lennox (1884–1960; Lennox-Gastaut syndrome) [3]. What remains most noteworthy about Anton's beliefs is the similarity in ideology of the Nazis. Shortly following Anton's death, Adolf Hitler was appointed Chancellor of Germany (1933) and invoked the 'Law for the Prevention of Hereditarily Diseased Offspring' that prescribed compulsory sterilization for people with diseases such as schizophrenia, epilepsy, and Huntington's chorea [15]. Thereafter, involuntary euthanasia programs such as T4 (1939–1945) enforced the killing of thousands, including children, with mental diseases under the supervision of physicians [15].

Fortunately, our history has drastically transformed, where involuntary euthanasia is no longer practiced. However, physicians who advocated for racial hygiene, like Gabriel Anton, are still recognized in our literature. Jeannette et al. discourages use of eponyms as diagnostic terms in favor of terms that describe distinct features of the particular disease. She highlights the specific instance of renaming Wegener's granulomatosis to granulomatosis with polyangiitis', on the evidence that Dr. Friedrich Wegener was a member of the Nazi party [16,17]. We therefore propose using the term, Bilateral occipital Lobe Infarct Neglect Deficit (BLIND) Syndrome, to replace the eponym, Anton syndrome, for individuals who develop cortical blindness with visual anosognosia after experiencing bilateral occipital lobe ischemic infarcts. While still respecting Anton's contributions to neuropsychiatry, we believe BLIND Syndrome will encompass the phenomenon of visual anosognosia in cortical blindness, as well as pay homage to the thousands who lost their lives in the 1900s' euthanasia programs.

Disclosure statement

No potential conflict of interest was reported by the author(s).

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