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Case Report

Anton syndrome with bilateral occipital infarct: A case report ☆

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ABSTRACT

Anton syndrome results from damage to the visual cortex of the occipital lobes, where the anterior visual pathways remain intact. This damage results in the characteristic triad of cortical blindness, visual anosognosia, and visual confabulation. This case describes an 80-year-old male with a background of renal transplant 7 years prior, admitted to hospital with worsening transplant function, and soon after developed sudden onset cortical blindness. On examination, the patient was found to be in denial of his blindness and showed signs of visual confabulation, both of which pointed toward a diagnosis of Anton syndrome. Radiological investigation with computed tomography (CT) and magnetic resonance imaging (MRI) revealed bilateral ischemic stroke of the occipital lobes, which was later theorized to have occurred due to sirolimus-induced thrombotic microangiopathy (TMA). To the author's knowledge, this is the first case report of sirolimus-induced TMA, bilateral ischemic occipital lobe stroke, and Anton's syndrome, within the same diagnosis. This case highlights that a diagnosis of bilateral occipital stroke with denial of blindness should suggest a diagnosis of Anton syndrome. This report also discusses the importance of differentiating bilateral occipital infarction on CT imaging, from posterior reversible encephalopathy syndrome (PRES), using MRI.

Introduction

Anton syndrome is a rare and unusual form of blindness which characteristically features bilateral cortical blindness, visual anosognosia (denial of vision loss) and visual confabulation (emergence of events and experiences which never took place) [1,2]. Cortical blindness occurs where there is damage to the visual cortex of the occipital lobe, with sparing of the anterior visual pathways. Most commonly cortical blindness is caused by infarction [3]. In this case, we discuss a combination of Anton syndrome secondary to bilateral occipital infarction.

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Case

This case is of an 80-year-old male with a background of end stage renal failure (of undetermined cause) and a subsequent renal transplant 7 years ago. The renal graft was a human leukocyte antigen and cytomegalovirus mismatch, and the patient was started on Tacrolimus following the transplant. Graft function began to decline early in 2022. Biopsy late in 2022 revealed arteriolar hyalinosis of the transplanted kidney, a known chronic effect of Tacrolimus, which was switched to sirolimus in January in 2022. Only 2 months after the switch, the patient presented to the acute medical unit complaining of swelling in both of his legs and passing small volumes of urine. On physical examination, lungs were clear on auscultation and abdomen was soft and nontender. His calves were nontender but had significant pitting edema in both legs equally. There was mild bruising of the arms noted on further inspection. Pulse rate was 93/min, regular in rhythm; blood pressure was 128/62, which later had soon sharply to a systolic above 200. His respiratory rate, oxygen saturations, and temperature were normal. Initial differential diagnosis was worsening renal graft function (transplant related acute kidney injury) with possible associated urinary tract infection. The gentleman was started on intravenous antibiotics and blood tests were taken—Creatinine 329 (baseline creatinine pretacrolimus switch was 200), hemoglobin 82 (110 baseline) and platelet 43 (160 baseline). A repeat blood test showed the platelet count drop further to 27 and hematology opinion was requested.

After 3 days following admission, the patient's wife had raised a concern that he started to struggle with his vision, particularly reading the clock on the wall, which prior to this day was not a problem. The patient himself reported no complaint of visual changes. Subsequent ophthalmology review of the patient's vision revealed bilateral decreased visual acuity to hand movements only. Despite this, the patient maintained opinion that his vision was normal, a sign of visual anosognosia. When asked to identify a person in a striped shirt in the room, he pointed to someone present, even though no one was wearing a striped shirt, demonstrating visual confabulation. Pupil reflexes and fundoscopy were unremarkable, suggesting an intact anterior visual pathway. This combination of bilateral vision loss, anosognosia, and confabulation led to a diagnosis of Anton syndrome. Initial management was conservative with and response was minimal. Further investigations were requested to search for any potentially reversible diagnosis.

CT head was performed (Fig. 1) soon after which revealed hypoattenuation in the occipital lobes bilaterally, suggesting a possibility of 2 initial differential diagnosis. First was a subacute right occipital and right corona radiata infarct, plus a more established left occipital lobe infarct (due to differences in density of each lesion). This difference in density made it a more likely differential than posterior reversible encephalopathy syndrome (PRES), which also presents with this similar bilateral appearance in the posterior cerebral artery (PCA) territory. This was followed up with an MRI brain (Fig. 2) which confirmed the changes were due to infarction of both PCA territories. This was not treated with antiplatelet therapy, due to the low platelet count, following advice from stroke and hematology teams. Over the coming days, patient's vision began to slowly improve, but never fully recovered. As vision began to improve, anosognosia and confabulation also appeared to be resolving, likely due to the maturation of the infarct as edema settled and collateral blood supply started to form around the infarcted brain tissue.

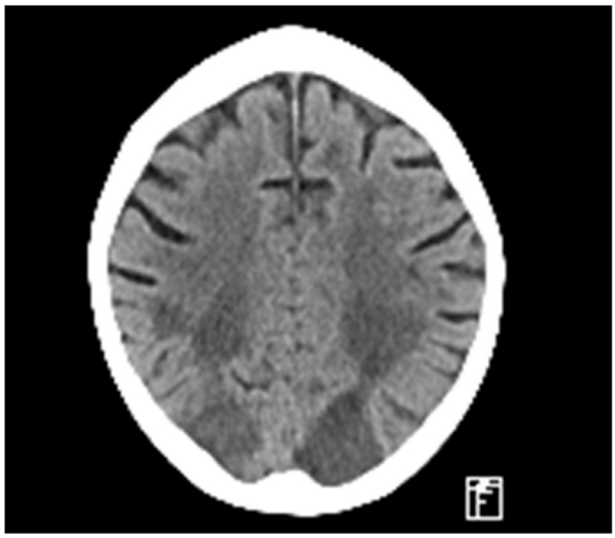

Fig. 1 – Computed tomography scan of the patient's brain, demonstrating ill-defined hypoattenuation within the right occipital lobe. There is a further focus of ill-defined hypoattenuation noted within the right corona radiata. There is more established hypoattenuation within the left occipital lobe which likely represents a maturing infarct, compared to the right.

Discussion between hematology, renal, and neuroradiology physicians took place to determine the cause of the bilateral PCA territory stroke. Due to the low platelet count, ADAMTS13 was conducted which ruled out thrombotic thrombocytopenic purpura (TTP). However, a peripheral blood smear found schistocytes and Burr cells, thus a provisional diagnosis of microangiopathic hemolytic anemia (MAHA) was made. The most likely explanation for the development of MAHA in this patient coincides with the drug switch from tacrolimus to sirolimus. When biochemistry was studied in retrospect, hemoglobin and platelets appear to start dropping at the time of this switch. A diagnosis of sirolimus-induced thrombotic microangiopathy (TMA) with secondary MAHA was made, which was theorized as the likely culprit for the PCA territory infarct, and subsequent development of Anton syndrome. Following the diagnosis of TMA, given the patients history of ESRF and frailty, a joint decision was made with the patient, family, and medical team to start palliative care. Further medical intervention was withheld. The patient through this period did not describe any significant discomfort, and as they deteriorated, palliative care treatments were helped guided by the observations of the family.

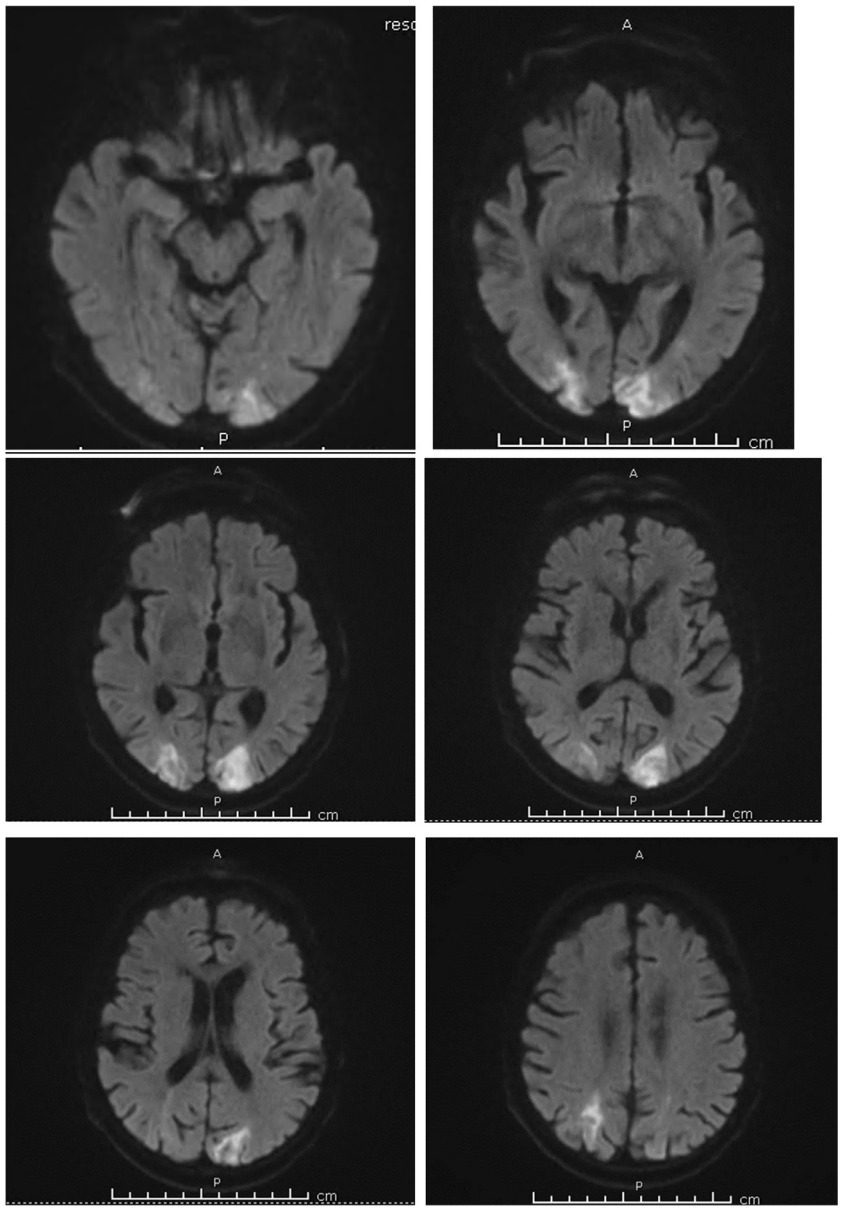

Fig. 2 – Magnetic resonance imaging of the patient's brain, demonstrating regions of cortical and juxta-cortical restricted diffusion both occipital and parietal lobes in posterior cerebral artery distribution bilaterally. These features are in keeping with multifocal infarction of both PCA territories.

Discussion

Gabriel Anton (1858-1933) was an Austrian neurologist and psychiatrist, known for his contribution to clinical neuroscience, particularly with studies of psychiatric conditions arising from damage to the brain cortex [4]. Anton syndrome is eponymous with Gabriel Anton. It is a rare condition which characteristically features 3 components: bilateral cortical blindness, visual anosognosia, and visual confabulation [1,2]. It is important to remember that for a diagnosis of Anton syndrome, there must be an absence of pre-existing memory impairment, such as dementia. This differentiates the syndrome from Dide-Botcazo syndrome, which also is known to include the presence of cortical blindness with visual anosognosia, but most commonly occurs in patients with significant dementia related memory loss [1].

What is thought to be the first case of Anton syndrome is when the Roman era stoic philosopher Seneca described visual anosognosia (denial of vision loss) and visual confabulation (emergence of events and experiences which never took place) in his case of Harpaste. Harpaste was a slave who became suddenly blind and used to deny her illness, complaining irrationally about a dark room, and asking to change quarters, in a room that was well lit [5,6]. In modern times, the number of cases on Anton syndrome per/epidemiology:

Anton syndrome is a very rare condition with the last long term report of the epidemiology stating only 28 published cases between 1965 and 2016 [5].

Cortical blindness arises where there is injury to the visual association cortex within the occipital lobes, with the visual pathways remaining intact. The most common cause of cortical blindness is ischemic stroke by occlusion of the PCA. When this is the case, it typically occurs unilaterally, resulting in a presentation of homonymous hemianopia. Bilateral occipital infarcts on the other hand, are not often seen in clinical practice, and can only be formally identified using diagnostic brain imaging modalities such as CT, MRI, or magnetic resonance angiography (MRA). Where there is a bilateral ischemia of the occipital cortex, it is possible that the occlusion lies more proximal in the PCA at the basilar artery bifurcation. Occlusion on of area should raise suspicion of a thrombo-embolic event [3,7]. In our case, a basilar artery embolism could not be ruled out; however, given the varying degree of hypoattenuation (Fig. 1) in each of the occipital lobes, separate PCA occlusive events are more likely to have occurred, secondary to the sirolimus-related TMA. Further MRA imaging of the brain would have been useful for assessing the basilar artery bifurcation; however, this would not have changed management plans and outcomes for the patient [8].

The main differential diagnosis that was considered in this case, given the presence of cortical blindness, was posterior reversible encephalopathy (PRES), previously referred to as reversible posterior leukoencephalopathy syndrome (RPLS). PRES is known to cause as bilateral involvement in occipital and parietal lobes. However despite its name, PRES can rarely be found in non-posterior distributions, including the frontal, cerebellar and brainstem regions [9,10]. PRES is known to be associated with a sudden rise in blood pressure, leading to hyperperfusion and resultant disruption of the blood-brain barrier, causing vasogenic edema and subsequently disturbances in vision [11]. As this case consisted of a renal failure patient with marked hypertension, it was important to differentiate the occipital changes found on CT with a follow-up MRI. PRES will typically nonrestrict on diffusion-weighted sequence, which is a hallmark feature of infarction, and what enabled PRES to be ruled out in our case (Fig. 2). PRES will also typically not show varying degrees of hypoattenuation throughout the affected areas, which further supports its negation versus infarction in this case (Fig. 1) [12]. Substantial recovery of visual function has been noted with PRES, when blood pressure is brought back under control and vasogenic edema settles. However, due to the nature of infarction, the patient in this case is unlikely to have a full recovery of vision, despite showing some initial improvement [13,14]. Given that PRES tends to have favorable outcomes for the patient's vision, it is important to make this differentiation from stroke, as it can impact the care pathway that the patient is sent on.

As a learning point, where there is a radiological diagnosis of ischemic stroke with suspicion of cortical blindness and visual anosognosia, Anton syndrome should be considered. Although a cerebrovascular event is the most common cause of Anton syndrome, PRES should also be considered, particularly in end stage renal failure patients or the presence of marked hypertension. Where the cause of Anton syndrome is vascular, only rapid recognition and treatment to help restore infarcted tissue will improve symptoms.

This case adds to the limited case literature involving TMA and Anton's syndrome. To the author's knowledge, there is no current case report identified within the literature which has described TMA, bilateral ischemic occipital lobe stroke and Anton's syndrome within the same diagnosis.

Patient consent

Written informed consent was obtained from the patient's next of kin for publication of this case report and the accompanying images.

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