

Case report

Anton's syndrome: a rare and unusual form of blindness

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SUMMARY

Anton syndrome is characterised by visual anosognosia. It results from damage to both occipital lobes, while the anterior visual pathways remain intact. We describe four cases of Anton's syndrome. First case is that of a 73-year-old woman, who presented with two separate events of intraparenchymal brain haemorrhage, 4 years apart. Her first stroke affected the left and second affected the right occipital lobe. Bilateral occipital lobe damage resulted in cortical blindness. Second case is an 88-year-old man, who suffered from two ischaemic strokes, 2 days apart. Each stroke involved one posterior cerebral artery. This resulted in bilateral occipital and temporal lobe infarcts. Third case is a 64-year-old woman with chronic renal failure, who suffered bilateral occipital lobe infarction after haemodialysis, due to posterior reversible encephalopathy syndrome. Last case is that of an 80-year-old woman who suffered a basilar artery stroke, resulting in bilateral thalamic, temporal and occipital lobe infarction.

BACKGROUND

Anton Babinski syndrome is characterised by blindness, without the self-awareness of blindness. The anterior visual tracts are intact and lesion is in the posterior cerebral cortex, located in occipital lobes. Patients is in complete denial of visual loss. This lack of insight poses an increased risk of falls. Confabulation is a major part of the syndrome. In a bookish picture of Anton syndrome, area of damage is confined to occipital lobes only. In real life, parts of the brain either in close proximity to occipital lobe or those sharing a common blood supply are also affected. This results in a wide array of symptomatology ranging from but not limited to, cognitive impairment, somnolence and drowsiness. This may complicate an already challenging clinical picture. Loss of vision has a major impact on rehabilitation after a stroke.

CASE PRESENTATION

A 73-year-old woman was brought to the emergency department by with symptoms of a sudden onset headache, vomiting and increased confusion. Patient denied any visual symptoms. While taking history, clinician noted her lack of eye contact. On examination of visual field by confrontation method, it became apparent that she could not reliably see in any quadrant of visual field. She had no facial droop and was moving all four limbs.

Cardiovascular examination was unremarkable. Her vital signs were stable.

Her medical history was significant for hypertension, for which she was on a single agent Amlodipine. She also described having a 'bleed in the brain' 4 years ago, from which she made a good recovery and was living at home with her daughter providing support for some domestic activities. Her premorbid modified Rankin score was 2.

Looking through the records a CT head from 4 years ago showed an intraparenchymal haemorrhage in the left temporal and occipital lobe (figure 1). Subsequent MRI and MR Angiography failed to establish any underlying aetiology of bleed.

In the emergency department, a CT head was repeated, which demonstrated an acute intraparenchymal haemorrhage, this time in the right parieto-occipital lobes with extension into the lateral and third ventricle. This also showed high density foci within the left temporal and occipital lobe, likely representing parenchymal calcification at the site of previous haematoma (figure 2).

She was admitted to the stroke ward and over course of next week, developed insight into her visual deficit. Her vision improved to some extent over next 2 weeks, but only to facial and colour recognition on bedside examination. She was needing assistance with mobility, personal care and activities of daily living by the time of discharge. She was discharged home with support. Her modified Rankin score was 4 on discharge.

Our second case is an 88-year-old previously well man. He fell in his garden and subsequently suffered a fracture of distal part of his right femur. Prior to this he had been living independently, with premorbid modified Rankin score of 1. Patient subsequently underwent an open reduction and internal fixation of his femur. A week after the procedure, he was an inpatient undergoing rehabilitation, when he became increasingly confused. Initial investigation and management of delirium was focused on electrolyte correction, pain management and search for a possible source of infection. Patient was also started on empiric antibiotics. A CT head done at this stage was reported as normal though neuro-radiologist documented some suspicion of increased density within the right posterior cerebral artery (PCA). No associated brain infarction was noted (figure 3). He was reviewed by the ortho-geriatric team, who did not find any clear neurological deficit and hence advised that stroke was less likely to be the underlying cause of his delirium. Patient continued to receive prophylactic



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Figure 1 Old CT head showing left occipito-temporal haemorrhage.

dalteparin as routine for prevention of venous thromboembolism as an inpatient.

Unfortunately, over the next 36 hours he became increasingly confused and drowsy. His Glasgow coma scale (GCS) dropped to 12. CT head was urgently repeated and this time it showed bilateral occipital lobe infarction, a left temporal lobe parenchymal haemorrhage and infarction of right posterior temporal lobe. A possible increased density within both PCAs was reported, suspicious of bilateral PCA infarcts (**figure 4**). Patient was transferred to stroke ward. Well established infarcts and an uncertain time of onset meant acute measures like thrombolysis and thrombectomy were not appropriate. Over the course of a few days, patient's agitation settled but he remained disoriented in time, place and person. Due to occipital lobe involvement, a formal visual field assessment by orthoptics team was organised, which confirmed complete bilateral visual loss. Patient remained adamant that he could see. Unfortunately, the accompanying severe cognitive impairment also remained static over coming weeks and patient, who previously was independent with a pre-morbid modified Rankin score of 1 on admission, was now discharged to a nursing home with postevent modified Rankin score of 4.



Figure 2 New CT head showing right occipital haemorrhage.

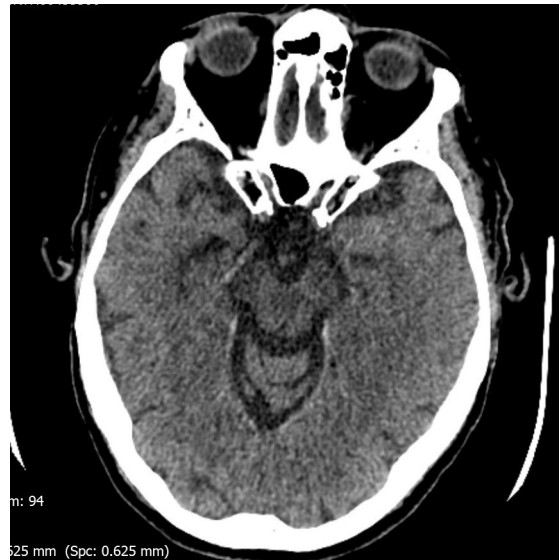


Figure 3 Hyperdense right posterior cerebral artery.

Our third case is of a 64-year-old type one diabetic with end stage renal failure. She routinely underwent haemodialysis three times a week. She lived in a bungalow with her husband, helped by carers visiting three times a day (pre-morbid modified Rankin score 4). She became confused after a dialysis session, so she was referred to medical admissions. On arrival, she was hypertensive with a blood pressure of 250/130 on presentation. Patient was not oriented in time place or person but there was no other motor or sensory deficit. It came to light on thorough neurological examination, including visual field assessment by confrontation method, that she had loss of vision in all fields. Patient denied any loss of vision and was confabulating. CT head revealed a hypodensity in the parieto-occipital lobes bilaterally, suggesting a possibility of posterior reversible encephalopathy syndrome (PRES) (**figure 5**). Other differential was stroke, so an MRI head and MR cerebral angiogram was attempted but abandoned mid procedure, as patient was not able to tolerate it. Patient was managed by lowering blood pressure with

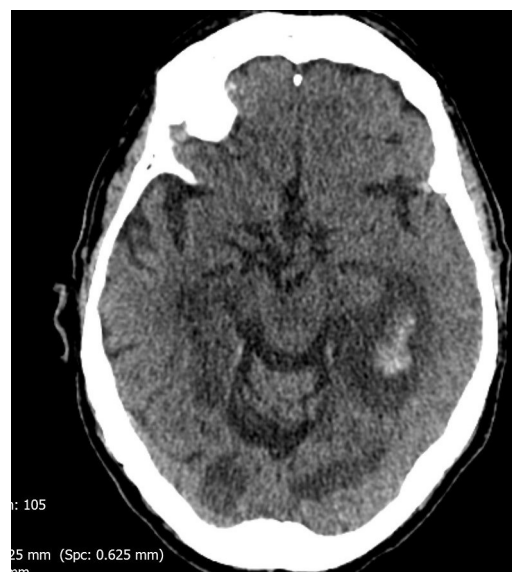


Figure 4 Bilateral posterior cerebral artery infarcts 24 hours later.



Figure 5 Bilateral hypodense occipital lobes due to posterior reversible encephalopathy syndrome.

intravenous nitrate infusion. Her vision failed to recover. After a period on the ward, she was discharged to a nursing home.

Our last case is that of an 80-year-old woman, brought to emergency department after a collapse. Husband described that she was well on waking up. An hour after waking up, she had symptom of dizziness, and then collapsed. She had medical history of hypertension and stroke in 2013, from which she made a good recovery and was now independent with a premorbid modified Rankin score of 1. On examination her GCS was 6. She was breathing spontaneously and maintaining oxygen saturation. Her pupils were fixed and dilated, and dolls eye reflex was absent. After securing airway, CT head and CT cerebral angiogram were performed that confirmed a thrombotic occlusion of a distal basilar artery (figure 6). As she had presented within thrombolysis window of 4.5 hours, intravenous alteplase was administered. Imaging had confirmed a proximal vessel occlusion, so case was referred to intervention neuro-radiologist. Mechanical thrombectomy of basilar artery was performed, with good visualisation of flow (figure 7). In next 24 hours patient made little if any recovery and next day CT head revealed bilateral PCA territory infarcts involving the thalami, medial temporal and occipital lobes infarcts, more extensive on right. We started post stroke care and nasogastric tube (NG) feeding. Over the next week patient made a slow recovery and regained consciousness. Detailed assessment at this stage confirmed gross visual loss in both eyes though patient kept denying this. Unfortunately,

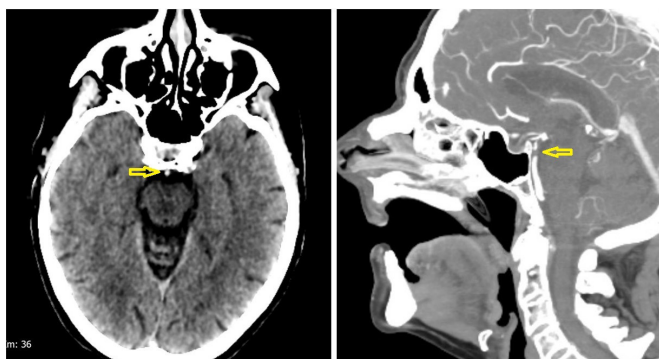


Figure 6 (A) Hyperdense basilar artery (arrow), (B) Filling defect in basilar artery on CT (arrow).

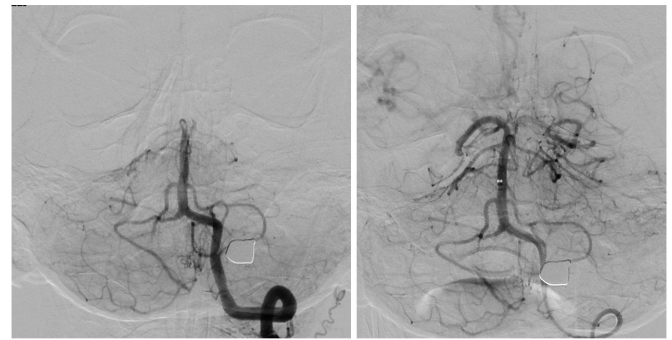


Figure 7 (A) Pre-stent and (B) post-stent thrombectomy and clot retrieval from basilar artery.

recovery was made difficult by a large neurological deficit and recurrent chest infections. Patient was eventually discharged to a hospice for end of life care.

OUTCOME AND FOLLOW-UP

All cases with Anton syndrome, had significant deterioration in functional status. At presentation, all four patients were living at home. Post event three out of four were institutionalised. Out of these, one was transferred to hospice for palliation. Discharged to home of one patient was made possible only with a big support package. All patients showed a large deterioration in their functional status as measured by modified Rankin scale.

DISCUSSION

Anton-Babinski syndrome is a rare neuropsychiatric disorder characterised by bilateral cortical blindness.¹ Its key features are visual agnosia, anosognosia and confabulation. It stands as one of the most striking examples of lack of insight.

The first ever description of a blind person, in denial of his blindness, was not by a clinician but by the French renaissance writer Michel de Montaigne (1533–1592). In the second book of his three-book series ‘Essais’, he mentioned a nobleman who loses his eye sight without taking a note of it.² The syndrome is named after Gabriel Anton (1858–1933), an Austrian Neurologist and Psychiatrist, best known for his descriptions of somatognosia and visual anosognosia.³

Anton’s syndrome should be considered in all cases of visual impairment, with bilateral occipital lobe lesions. Underlying pathology is injury to the visual association cortex in the occipital lobes. The anterior visual pathways remain intact. The most common cause of cortical blindness is PCA occlusion.⁴ When the terminal bifurcation of basilar artery is involved in a thromboembolic event, resulting stroke can be bilateral.⁵ This was the cause in our fourth case.

In the first case described here, two separate events of spontaneous intracerebral haemorrhage, involving each occipital lobe, first on left and then on right, led to complete blindness. The most common cause of spontaneous intracerebral haemorrhage is hypertensive vasculopathy.^{6–8} But in cases of lobar haemorrhages, cerebral amyloid angiopathy (CAA) should also be considered as a potential cause,^{9 10} especially when dealing with recurrent lobar haemorrhages in a patient above the age of 55.¹¹ CAA tends to favour areas of brain supplied by posterior circulation, meaning areas perfused by the vertebra-Basilar system. Hence, it more commonly affects cerebellum, occipital and temporal lobes, than the frontal and parietal lobes.¹² This might be due to failure of posterior circulation vessels to eliminate beta amyloid peptide efficiently.¹³ There are also reported

Reminder of important clinical lesson

cases of bilateral occipital lobe haemorrhages due to PRES causing sudden onset blindness.¹⁴ Interesting feature of the first case presented here, is that the two haemorrhagic strokes are 4 years apart, favouring CAA.

While Anton syndrome is characterised by visual loss with visual agnosia and confabulation, cognitive impairment is typically not associated with it.¹⁵ PCA supplies the occipital lobe through the lateral and medial occipital cortical branches. It also supplies the uncus and anterior part of the fusiform gyrus (through the Anterior temporal branch) and inferior temporal gyrus (through the Posterior temporal branch). These parts control recognition, visual processing, memory and perception.^{16 17} Damage to these can lead to impairment of executive functioning and memory.¹⁸ As main motor and sensory pathways remain mostly intact, despite damage to an extensive area of brain, these patients escape without having major motor or sensory deficits. This can fool clinicians especially when dealing with elderly patients where causes of confusion can be manifold. As cerebral infarction may not be visible on CT scan in acute phase, the initial work up of delirium may not yield a positive diagnosis. This is true for the second case described here. A thorough clinical assessment, including visual fields by confrontation can help in cooperative patients. In uncooperative patients or if suspecting that patient is confabulating, visual threat can be used to detect if patient can truly see.

Bilateral temporal lobe involvement is a unique pattern with a limited differential diagnosis.¹⁹ Occlusion of the basilar tip and bilateral posterior cerebral arteries can cause infarction of both temporo-occipital regions.¹⁹ These patients present with abrupt onset of confusion, agitation and visual disturbance. Agitation can settle within days, but visual and cognitive impairment can persist for years.²⁰ Further symptoms can arise when both thalami are affected. Although presentation of bilateral thalamic infarct can be visual, sensory or motor,²¹ the last patient described, suffered from increased somnolence and drowsiness. These features have been described with thalamic lesions. The proposed mechanism is disruption of the activating impulses from brainstem reticular formation.²²

As mentioned above, PRES is also known to involve bilateral occipital and parietal lobes. Other than visual loss, it can present with headache, confusion and seizures. It has been described with sudden rise in blood pressure in cases of renal failure.²³ By definition, PRES should be reversible and neurological deficit should improve on lowering the blood pressure.²⁴ Although delay in diagnosis can lead to permanent neurological deficit, which may have been the case with our third patient described here. This could be due to secondary processes than can occur with PRES like focal cerebral vasospasm causing infarction or

haemorrhage. In appropriate cases, MRI can be helpful in differentiating stroke from PRES. PRES will present with hyperintense signals on T2 and fluid attenuation inversion recovery (FLAIR) sequences of MRI, while it will not restrict on diffusion weighted sequence, which is a feature, typical of infarction.²⁵

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Learning points

- ▶ In the time of face, arm and speech test assessment, clinicians can miss important aspects of a cerebrovascular disease, especially in a confused elderly patient, often leading to misdiagnosis.
- ▶ Extensive brain damage, especially in posterior circulation territory, giving deficits like visual defects and cognitive impairment, can be missed on initial assessment if clinicians are not vigilant.
- ▶ Visual and cognitive impairment can have both short-term and long-term implications on rehabilitation and prognosis.

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