Anton syndrome as a result of MS exacerbation

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Visual anosognosia is the lack of awareness and denial of one's blindness. It is often referred to as Anton syndrome (AS), after one of the physicians who described the phenomenon of lacking insight into one's sensory deficit. There are limited numbers of case reports documenting the condition and many refer to the phenomenon in association with bilateral occipital infarcts. We describe a patient who developed visual anosognosia as a result of multiple sclerosis (MS).

Case report

A 24-year-old left-handed woman presented with right-sided hemiparesis and homonymous hemianopia. Head CT showed a hypodensity spanning the left temporo-parieto-occipital white matter. Brain MRI demonstrated a subcortical lesion with high T2/fluid-attenuated inversion recovery (FLAIR) signal, diffusion restriction, areas of apparent diffusion coefficient (ADC) dropout, no contrast enhancement, and mild mass effect on the lateral ventricle (figure, A). Serologic investigation (erythrocyte sedimentation rate, C-reactive protein, SS-A/B, antineutrophil cytoplasmic antibodies, antinuclear antibodies, angiotensin-converting enzyme, rheumatoid factor, aquaporin-4 antibodies, HIV, hepatitis A, B, and C, JC virus, cytomegalovirus, Epstein-Barr virus, human T-cell lymphotropic virus) was unremarkable. CSF glucose, protein, and cell counts were normal. CSF immunoglobulin G was elevated (>6 mg/dL) and isoelectric focusing reported presence of oligoclonal bands (>2 unique bands) in CSF with absence in serum. The patient was treated with IV methylprednisolone due to the high suspicion of a demyelinating process. She made gradual, incomplete recovery.

Follow-up MRI scans performed over the course of 1 year began to show small, asymptomatic juxtacortical and periventricular lesions in addition to one infratentorial pontine lesion (figure, B-D). The initial left-sided tumefactive lesion remained stable with ex vacuo dilation of the left lateral horn adjacent to gliosis in the left hemisphere.

Sixteen months after the first attack, the patient presented with global aphasia, left-sided sensorimotor deficiency, and cortical blindness. Brain MRI showed a new right-sided parieto-occipital white matter lesion with T2/FLAIR hyperintensity, restricted diffusion, dropout on ADC, and no contrast enhancement (figure, E). MRI of the total spine did not show any additional lesions. Laboratory studies were again unrevealing. The patient met the revised MacDonald criteria for MS with her 2 attacks and numerous T2 lesions throughout time and space.¹ Due to the aggressive nature of the patient's tumefactive relapsing-remitting MS, she was treated with methylprednisolone and plasmapheresis, and promptly started on alemtuzumab. All her symptoms improved over months except for cortical blindness.

Despite being unable to detect light or blink to threat, the patient insisted that her vision was intact and confabulated the presence and placement of objects when asked about her surroundings. Her vision recovered over 2 years; first she gained insight that she was blind, then was able

Practical Implications

While Anton syndrome is most commonly associated with bilateral occipital infarcts, consider a broader differential of pathology when patients present with visual anosognosia.

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Figure Demyelinating lesions on MRI of the brain over 2 years

T2/fluid-attenuated inversion recovery-weighted MRI brain sequences demonstrate (A) an extensive hyperintense subcortical periventricular lesion involving the left frontal, parietal, and occipital white matter, which caused mild mass effect on the posterior horn of the left lateral ventricle during the first attack, and (B-D) appearances of asymptomatic hyperintense periventricular, juxtacortical, and infratentorial lesions bilaterally in between attacks, followed by (E) a right hyperintense fronto-parieto-occipital subcortical periventricular lesion in addition to the previously known left posterior region of encephalomalacia and surrounding gliosis and ex vacuo dilation of the left posterior lateral ventricle during the second attack.

to identify light sources, and then was able to describe objects accurately despite having achromatopsia. Gradually, her color discrimination improved enough to self-apply makeup. Her vision plateaued with being able to describe scenes in magazine accurately, navigate her surroundings, and read with difficulty.

DISCUSSION

AS is rare; only 28 cases have been published between 1965 and 2016 (table). Of these, 78.5% (22/28) of reports described the condition in vascular pathologies (table). Visual anosognosia was most commonly associated with occipital infarcts (19 reports, 86.3% of reports relating to vascular etiologies) (table). Other vascular cases of visual anosognosia were in the setting of postpregnancy reversible cerebrovascular syndrome, systemic angitis, and hypovolemic shock (table).²⁻⁴

Our patient's case is an addition to scarce reports of AS in nonvascular etiologies. Prior to our description, the syndrome has been reported in association with 6 other conditions: progressive multifocal leukoencephalopathy in the setting of HIV, adrenoleukodystrophy, bisynchronous occipital seizures, mitochondrial encephalopathy and lactic acidosis, alcoholic withdrawal hallucinosis in a blind patient, and traumatic optic neuropathy in the setting of bifrontal contusions (table). It should be noted that the patient with AS and systemic angitis described in the report by Carvahal et al.³ also had a preexisting history of Hashimoto thyroiditis and MS. The onset of symptoms correlated to the patient's infarcts secondary

Table Case reports to date that describe Anton syndrome in relation to each patient's pathology

Case reports regarding visual anosognosia, reference no. ^a	Etiology	No. of patients (total n = 28)
Nonvascular etiologies		
5	Posttraumatic bifrontal contusions	1
6	Progressive multifocal leukoencephalopathy	1
7	Adrenoleukodystrophy	1
e1	Seizures	1
e2	Alcohol withdrawal hallucinosis	1
e3	Mitochondrial myopathy, encephalopathy, lactic acidosis, and strokelike episodes	1
	Total	6
Vascular etiologies		
2	Reversible cerebrovascular syndrome	1
3	Bilateral occipital infarcts	1
3	Systemic angitis	1
4	Hemorrhagic shock	1
e4	Bilateral occipital infarcts	1
e5	Bilateral occipital infarcts	1
e6	Bilateral occipital infarcts	1
e7	Bilateral occipital infarcts	1
e8	Bilateral occipital infarcts	1
e9	Bilateral occipital infarcts	1
e10	Bilateral occipital infarcts	1
e11	Bilateral occipital infarcts	1
e12	Bilateral occipital infarcts	1
e13	Bilateral occipital infarcts	1
e14	Bilateral occipital infarcts	1
e15	Bilateral occipital infarcts	1
e16	Bilateral occipital infarcts	1
e17	Bilateral occipital infarcts	1
e18	Bilateral occipital infarcts	1
e19	Bilateral occipital infarcts	1
e20	Bilateral occipital infarcts	1
e20	Unilateral occipital infarct	1
e20	Total	22

^aSee references and e-references (available at Neurology.org/cp).

to angitis and not as a result of MS. It should also be noted that the patient reported in the report by McDaniel and McDaniel⁵ had monocular blindness and alcoholism, and demonstrated global confabulation.

Given that our patient was able to recover her insight of blindness and recover some of her actual vision, we hypothesize that the syndrome is reversible with the recovery of white matter tract function. Visual evoked potentials (VEPs) may have been helpful to support this theory

Supplemental Data

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but were not obtained. In future cases, performing VEPs may be considered to test this hypothesis. We demonstrated that an MS exacerbation can result in visual anosognosia and improve. Our case expands the spectrum of visual phenomena associated with MS.

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Nina Kim: drafted and revised original manuscript, acquired and interpreted data. Deepti Anbarasan: edited manuscript, acquired and interpreted data. Jonathan Howard: edited manuscript, acquired, and interpreted data, provided case supervision.

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