0–10, she rated her memory “5” and vision “7” (ie, superior to memory).

AST was reviewed 6 months later. She was well orientated in time and place. She stated that her vision was “very good, my eyes seem to be a lot better than before…” She estimated her vision as being 60–70% (normal) and her memory 50% (normal). At home, she had reported seeing non-existent people (eg, a girl who had to be fed and a baby).

She failed to blink when threatened by a gesture towards her face. She was asked to indicate when a very bright light directed on her face was turned “on” and scored 8/15 (correct). By contrast, she scored 8/8 (correct) when asked to indicate whenever she was touched on her hand. She said “I can see a little girl, vases of flowers, tea cups… “, these images are “distinct” and the flowers “are blue”. She then tried to touch them by bending and stretching her arm towards them. At the end of this assessment, AST estimated her vision to be “very poor, 30%” and her memory “20%”, again implying that her vision was more efficient than her memory. Her oral spelling was found to be at an average level.

Our investigations demonstrated that AST had no light perception and had no residual conscious visual function. She lacked insight into her visual impairment. Unlike the great majority of previously reported cases of Anton syndrome, her intellectual skills, although not intact, were relatively well preserved. She did not present with amnesia, confirming that this is not a sine qua non for the development of the syndrome. Her executive (frontal lobe) skills were mildly impaired. Her lack of insight was specific to loss of vision. Only occasionally did she accept a “reduction” in her vision, which she attributed to “poor lighting”. She frequently experienced what appeared to be visual hallucinations and, at times, acted upon them. Despite extensive damage in the occipital, posterior, temporal and parietal lobes, she performed creditably on tasks of visual imagery, suggesting that intact visual cortex is not essential for this function (see also Abutalebi and colleagues’).

The pathology in AST is unusual in that cranial irradiation resulted in extensive damage to the posterior brain which was otherwise intact. Most cases of cortical blindness due to isolated bilateral occipital infarction have pathology limited to the supply of the posterior cerebral arteries while most reported cases of Anton syndrome have more extensive brain damage.

Visual hallucinations are visual perceptual experiences in the absence of corresponding visual stimulation. AST seemed to experience a vivid hallucinatory visual world. Generally, however, one cannot be certain that the experiences patients with Anton syndrome report are hallucinations per se, as opposed to products of imagination or confabulations. Clearly, the relationship between the often reported “confabulation” and the occurrence of hallucinations without insight in Anton syndrome merits further research (see supplementary material available online). We have no way of knowing that AST experienced conscious visual images whenever she denied blindness, nor can we say what the interaction might be between the development of visual hallucinations and anosognosia for blindness. However, we do consider that such an interaction may be of significance in this syndrome. The specific issue of lack of insight in blind patients can only be resolved with further detailed individual and group studies, with particular emphasis on cases such as AST who, despite the marked impairment of her vision, was cognitively relatively well preserved, with demonstrable clarity of thought.

REFERENCES


CORRECTION

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S Viegas, A Weir, M Esiri, et al. Symptomatic, radiological and pathological involvement of the hypothalamus in neuromyelitis optica (J Neurol Neurosurg Psychiatry 2009;80:679–82). In the legend for figure 3 it should read: “Detection of aquaporin 4 antibodies was positive (43 and 77, controls <25 FU).”