HISTORY OF NEURO-OPHTHALMOLOGY

Optic Neuritis: Another Dickensian Diagnosis

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ABSTRACT

The clinical diagnosis and natural history of optic neuritis was established in the late 1880s by the ophthalmologists von Graefe and Nettleship. The earlier, accurate and insightful description of transient, bilateral visual loss of Esther, the main character in the Charles Dickens novel *Bleak House* (1852–1853), suggests optic neuritis as a Dickensian diagnosis. Remarkably, Dickens' observations, also preceding the earliest clinical description of multiple sclerosis by Charcot in 1868, described many clinical features such as a prodromal phase; a nadir; gradual recovery over weeks; glare disability; reduced contrast sensitivity; possibly Uhthoff's phenomenon; and visual fading. All this with an accuracy that, to quote Russell Brain, "would credit a trained physician."

Keywords: Charles Dickens, history, optic neuritis

INTRODUCTION

Charles Dickens (1812–1870) was an English writer who published his novels in monthly instalments. *Bleak House* was published in 20 such instalments between March 1852 and September 1853. Due to reforms at the institution described, the Court of Chancery, the narrative must have taken place before 1842. The year 1827 was suggested by Sir William Holdsworth (1871–1944) in his book *Charles Dickens as a Legal Historian*, published in 1928¹ because of the legal reforms referred to that were taking place at that time.

"Dickens looked on disease with the observing eye of expert clinician, and he recorded what he saw, and what the patient told him, so that he often gives us accounts which would credit a trained physician. [...] But I have said enough to show that, at a time when medicine itself was only just beginning to recognize the importance of physical signs, the characters in the world of Dickens's imagination are so real that they have

recognizable diseases of body and mind, described with the accuracy and insight of a great clinical observer."¹

It was in this article that Lord Brain introduced the term "Dickensian diagnoses."

CASE REPORT

The fictitious case is a lady from Dickens' novel *Bleak House*, called Esther Summerville (Figure 1). The text was cited from the open-access Project Gutenberg (available online at www.gutenberg.org).^{*} The selected citations were taken from subsequent text passages to maintain the fictitious chronology.

The prodromal phase of Esther's illness starts one evening:

"It was a great morning when I could tell Ada all this as she stood out in the garden; and it was a great evening when Charley and I at last took tea together in the next

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FIGURE 1 "Caddy's Flowers"—Showing Esther Summerville (holding flowers). Illustrating an episode from Chapter 17 of *Bleak House* by Charles Dickens.

room. But on that same evening, I felt that I was stricken cold."

Within one day her unwellness progressed:

"In the evening I was so much worse that I resolved to prepare Charley, with which view I said, 'You're getting quite strong, Charley, are you not?'"

This really marks the onset of Esther's disease:

"Charley promised, and I lay down, for I was very heavy. I saw the doctor that night and asked the favour of him that I wished to ask relative to his saying nothing of my illness in the house as yet. I have a very indistinct remembrance of that night melting into day, and of day melting into night again; but I was just able on the first morning to get to the window and speak to my darling."

Two days later Esther becomes blind:

"I believe it, my dear Charley. And now come and sit beside me for a little while, and touch me with your hand. For I cannot see you, Charley; I am blind."

There was an unaccounted number of weeks during which Esther remained sick:

"I lay ill through several weeks."

There is a reference to a visual hallucination during this period, whether febrile or due to visual deprivation alone cannot be determined with any degree of certainty from the text:

"Dare I hint at that worse time when, strung together somewhere in great black space, there was a flaming necklace, or ring, or starry circle of some kind, of which I was one of the beads!"

Subsequently Esther's vision returned:

"I was in this state when I first shrunk from the light as it twinkled on me once more, and knew with a boundless joy for which no words are rapturous enough that I should see again."

The recovery of Esther's vision was gradual:

"But now, my sight strengthening and the glorious light coming every day more fully and brightly on me, I could read the letters that my dear wrote to me every morning and evening..."

Interestingly, whilst being able to read, there appeared to be some residual visual defects:

"And I did read all the noble history, though very slowly and imperfectly then, for my eyes were so dimmed that I could not see the words, and I cried so much that I was many times obliged to lay down the long account she had cut out of the newspaper."

The next episode in which troubles with Esther's visual perception were described refers to an episode after some physical exercise:

"I was resting at my favourite point after a long ramble, and Charley was gathering violets at a little distance from me. I had been looking at the Ghost's Walk lying in a deep shade of masonry afar off and picturing to myself the female shape that was said to haunt it when I became aware of a figure approaching through the wood. The perspective was so long and so darkened by leaves, and the shadows of the branches on the ground made it so much more intricate to the eye, that at first I could not discern what figure it was. By little and little it revealed itself to be a woman's—a lady's—Lady Dedlock's."

And finally, it is repeated that she continues having trouble with prolonged reading, without the subjective experience of temporary loss of vision:

"[...] that my eyes were too often blinded to read much at a time."

DISCUSSION

Charles Dickens was a great observer.² The detailed description of some of Dickens' characters, such as the obese boy Joe, actually gave rise to an eponymous syndrome "Pickwickian syndrome."³ His detailed clinical descriptions continues to fascinate physicians of all disciplines.^{3,4} He was known to visit public institutions where he encountered blindness, deafness, and a large range of psychiatric illnesses.³ Dickens did not stop at observing patients, he promoted treatment for children and was, for example, supportive of the establishment of the Great Ormond Street Hospital for Sick Children in 1852.

All retrospective interpretation of the wealth of symptoms described by Dickens will remain speculative. Acknowledging this limitation, one may within reason assume that Dickens alluded in his many texts to diseases such as tuberculosis, restless legs syndrome, Parkinson disease, Tourette syndrome, torticollis, epilepsy, head injuries, cerebral atherosclerosis and stroke, diplopia, aphasia, supranuclear palsy, déjà vu, locked-in syndrome, paraplegia, dementia, Hutchinson-Gilford progeria syndrome, superior vena cava obstruction, cerebellar ataxia, cachexia, numerous psychiatric disorders, asthma, chronic bronchitis, sleep apnoea, alcohol-related problems, to name but few.²⁻⁶ To these many Dickensian diagnoses one may tentatively add bilateral simultaneous optic neuritis.

The text by Dickens provides a detailed description of Esther's visual symptoms, but remains more obscure on the possible aetiology: not unlike many cases referred to a neuro-ophthalmological service today. For this reason, the discussion will remain focused on the visual symptoms.

The narrated time frame suggests that Esther's visual symptoms started after an incubation period of days to weeks with a feverish prodromal phase of 1–2 days before bilateral loss of vision occurred. The association of optic neuritis with a range of infections is well recognised.⁷

Based on the change of seasons described with great detail by Dickens, Esther's symptoms started some time in spring or early summer and had already recovered before summer had passed. The incidence of optic neuritis is known to peak in spring.⁸

At the onset of her symptoms, there is no mention of retrobulbar pain, which is very common in optic neuritis.⁷ However, when the intracranial optic nerve is involved, pain is usually not experienced, whereas it will almost always be present when the orbital portion of the nerve is affected. As far as one can draw a conclusion from negative evidence, we may speculate that the optic neuritis was either bilateral involvement of the intracranial optic nerves or even a chiasmitis, which has often been misdiagnosed as bilateral

simultaneous retrobulbar optic neuritis. It is not exactly recorded for how long Esther remained blind, but her blindness lasted at least several weeks. Certainly, she had recovered in the summer months. This falls well within the time frame of recovery from optic neuritis.⁷

At the nadir of her disease, she describes some positive visual symptoms. It would be extremely speculative to interpret this as a reference to scintillations and or photopsias known to occur in optic neuritis.⁶ Visual hallucinations of unknown aetiology remain another possibility or a dream.

The description of the early recovery phase is intriguing in that Dickens mentions that Esther first shrunk from the light. Some patients with optic neuritis may notice glare disability under photopic conditions during the recovery phase. Daylight glare disability is not always enquired of, but if found to be a problem it is readily treated with dark glasses.

Visual recovery was gradual, just as patients with optic neuritis experience further improvement over a period of several months.⁷

The three remaining references to Esther's visual recovery are interesting. First, she cannot read for long as her eyes were "dimmed". Recently, the US neuro-ophthalmologist Laura Balcer has been instrumental in promoting low-contrast testing.⁹ Reduced contrast sensitivity is characteristics following optic neuritis.⁷ Typically, one would also expect reduced colour vision. It would be unusual for Dickens to have missed such a characteristic feature and one may speculate that this could have been because he described a case of bilateral simultaneous optic neuritis. Likewise, he makes no reference to an afferent pupillary deficit, also characteristic in optic neuritis, but more difficult to demonstrate in bilateral cases because a comparison cannot be made between the two eyes.⁷ Certainly, Dickens was aware of altered pupil responses, as Lord Brain alludes to a description of what may have been the Argyll Robertson pupil in Maggy (*Little Dorrit*).² In *Bleak House*, Dickens even describes the careful examination of the pupil response in Sir Leicester Dedlock.² Finally, no reference is made to Pulfrich's phenomenon, which describes difficulties with perception in depth following unilateral optic neuritis.¹⁰ Taken together, this is consistent with a history of bilateral optic neuritis in Esther.

Next, during her summer break in the lodgings of Mr. Boythorn, there is an intriguing episode of transient visual symptoms. Esther had just completed some physical exercise, because she rested after a "long ramble." Again, interpretation of her transient visual symptoms must remain speculative. One possibility, which may be related to a previous episode of bilateral optic neuritis, is Uhthoff's phenomenon. Following exercise, particularly in the "warmer" British summer months, can lead to transient,

conduction-block-related worsening of vision, a phenomenon attributed to Uhthoff.⁷ A reasonable alternative to this interpretation is that the unusual lighting conditions and shadows of the branches gave rise to problems due to reduced contrast sensitivity.

Finally, an exquisite account is given how vision dimmed after reading for longer periods. It is not uncommon that following optic neuritis patients complain about visual fading.⁷

An important limitation of the retrospective interpretation of a fictional character is the complete lack for evidence from objective assessments. Of note, objective assessment of the optic nerve head became only possible with the invention of the ophthalmoscope by Helmholtz (1851). This had then enabled the ophthalmologists von Graefe (1828–1870) in Germany¹¹ and Nettleship (1845–1913) in England¹² to characterise optic neuritis; to quote the latter author: “They are characterized by failure of sight limited to one eye, often accompanied by neuralgic pain about the temple and orbit and by pain in moving the eye; many recover but permanent damage and even total blindness may ensue; there is at first little, sometimes no, ophthalmoscopic change, but the disc often becomes more or less atrophic in a few weeks, and occasionally there are slight retinal changes.”

None of these objective assessments were available at the time Dickens was writing, and all interpretation of the true nature of the transient, bilateral blindness of his fictional character Esther remains essentially speculative. Nonetheless, the descriptions were sufficiently detailed to suggest a plausible chronology from known visual symptoms following optic neuritis including glare disability, reduced contrast sensitivity, Uhthoff’s phenomenon, and visual fading. Coincidentally, Dickens may have predated Uhthoff in his description of alcohol-related optic neuropathy in referring to another character of the book, Mr Krook, “He’ll have drunk himself blind by that time. He has been at it all day.” Mr Krook’s ultimate demise as a result of spontaneous combustion also provides an illustration of the extent to which Dickens researched the veracity of events portrayed in his novels: it is in the Preface to *Bleak House* (published in the Charles Dickens edition of 1867) that Dickens not only confirms the truth behind his account of Chancery but also provides evidence of attested occurrences of spontaneous combustion in response to critics.

CONCLUSION

Bilateral, simultaneous optic neuritis may have been a possible differential diagnosis of the fictive character Esther in Charles Dickens’ novel *Bleak House*. This report adds to the literature on Dickensian diagnoses first introduced by Lord Brain and extended ever since.^{2–6}

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