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Medically unexplained visual loss in a specialist clinic: a retrospective case–control comparison

Éanna D. O'Leary [a,*], Benjamin McNeillis [b], Selma Aybek [c], Paul Riordan-Eva [d], Anthony S. David [a]

[a] Section of Cognitive Neuropsychiatry, Institute of Psychiatry, Psychology & Neuroscience, King's College London, London SE5 8AF, UK
[b] South London and Maudsley NHS Foundation Trust, London, UK
[c] Laboratory for Behavioral Neurology and Imaging of Cognition, Fundamental Neurosciences Department, Geneva University, Switzerland
[d] Department of Ophthalmology, King's College Hospital, London SE5 9RS, UK

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ABSTRACT

Purpose: To compare the clinical and demographic characteristics of adult patients with nonorganic or medically unexplained visual loss (MUVL) to those with other common conditions presenting to a neuro-ophthalmology clinic.
Methods: Case–control design: a retrospective review of medical notes on a consecutive case series of 49 patients assessed at the King's College Hospital neuro-ophthalmology clinic with unexplained visual loss and matched with the next assessed patient identified from clinic records. Patients presented post-symptom onset with a mean clinical course of 30 months (SD = 67 months) and standard clinical examination used to confirm diagnoses, alongside ancillary investigations if required.
Results: Seventy-two percent (n = 36) of MUVL patients were female. In comparison with patients with organic visual disorders, MUVL cases presented with significantly higher rates of bilateral (cf. unilateral) visual impairment (41%, n = 20), premorbid psychiatric (27%, n = 13) as well as functional (24%, n = 12) diagnoses and psychotropic medication usage (22%, n = 11). Medically unexplained cases were significantly more likely to report preceding psychological stress (n = 9; 18%).
Conclusions: Medically unexplained visual impairment may be regarded as part of the spectrum of medically unexplained disorders seen in the general hospital setting. Research is needed to determine long-term outcomes and effective tailored interventions.

1. Introduction

Many medical specialities frequently encounter individuals whose presentation cannot be accounted for by underlying organic pathology. Such medically unexplained symptoms are common across general/internal medicine and neurological fields, representing the most common diagnosis seen amongst some specialities [25]. Classification of disorders that have no known organic basis are included in the Diagnostic and Statistical Manual of Mental Disorders, DSM-V [1] under "somatic symptom and related disorders" (formerly "Somatoform Disorders") containing the subheading of "Conversion Disorder (Functional Neurological Symptom Disorder)" where psychological factors are judged to be associated with the presenting symptom or deficit. These are differentiated from "Factitious Disorder", by the fact that the deficit is not intentionally produced or feigned. This medically unexplained group present diagnostic uncertainty, management strain, high levels of distress/disability and risk resultant exposure to iatrogenic harm from misdiagnosis and over-investigation [9,10,16,19,30,31].

Amongst such functional deficits, medically unexplained visual loss (MUVL), characterised by the onset of visual impairment without evidence of contributory ocular or non-ocular pathology, represents a consistent presentation to primary and secondary care services. Typically, patients present with subjective visual complaints, but extensive investigation yields no apparent organic abnormalities, frequently resulting in neurologic or neuro-ophthalmic referral after no organic ocular cause is identified [12].

Villegas and Ilsen reported that between 5%–12% of patients presenting with visual loss to neuro-ophthalmology are subsequently diagnosed with medically unexplained/functional visual loss [34]. Further, simultaneous functional visual loss and organic visual loss can co-occur in the same individual, presenting as a "functional overlay" [17], with associated incidence reported between 16%–53% [27,29]. In idiopathic intracranial hypertension, functional visual loss has been reported to occur in 6% of patients with the potential to result in unnecessary surgical treatment [23]. Furthermore, authors report that up to 38% of patients with MUVL may present to other specialities with unexplained symptoms [13]. However, clinical descriptions of patients with

* Corresponding author at: Department of Psychosis Studies, PO Box 68, Section of Cognitive Neuropsychiatry, Institute of Psychiatry, Psychology & Neuroscience, King's College London, De Crespigny Park, Denmark Hill, London SE5 8AF, UK.
E-mail address: eanna.oleary@kcl.ac.uk (É.D. O'Leary).

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unexplained visual symptoms remain underrepresented within the neuro-ophthalmic literature and functional symptom medical reports [6,35].

Regarding nosology, the terminology of unexplained symptoms has evoked ample debate, with terms such as "nonorganic" [4,20] and "functional" [2,27] gaining adherents in some neuro-ophthalmic literature. We, alongside others, prefer the term "medically unexplained symptoms" as it is purely descriptive and makes no aetiological assumptions [13,36].

The purpose of the present study was to examine the characteristics of a group of consecutive adult patients with MUVL attending a specialist ophthalmology clinic; the characteristics of such patients were compared to a control-group of patients from the same clinic diagnosed with miscellaneous organic pathology.

2. Methods

2.1. Setting

UK National Health Service (NHS), King's College Hospital NHS Foundation Trust, London.

2.2. Participants

2.2.1. Cases

49 consecutive patients with a final designation of MUVL identified from the clinic records of a specialist neuro-ophthalmology clinic who were personally assessed by an experienced consultant (PRE) over an approximate 5 year period, 2003–2008.

2.2.2. Controls

Medical records were compared to those of the person next seen in the clinic by the consultant. Case notes were examined to determine demographic and clinical characteristics and patterns of visual impairment. MUVL was defined as reduced vision without any identifiable organic cause with concomitant one or more inconsistent feature(s) on visual function testing [13]. The primary clinical manifestations included unilateral or bilateral reduction of visual acuity with or without reduction of visual field, unilateral or bilateral reduction of visual field with normal visual acuity, functional overlay (see above) and convergence spasm. In many cases presenting with reduction of visual acuity the diagnosis of MUVL was suggested by results of visual field testing.

2.2.3. Assessments

Data were extracted on prior medical histories relating to neurological, ophthalmological, functional and psychiatric diagnoses. Duration of symptoms prior to diagnosis, documented psychotropic and analgesic medication usage, pain, headache and potential triggers (physical or psychological trauma) were also noted.

Demographic and outcome data of cases and controls were compared using SPSS (v. 22), and the relevant tests applied to analyse outcomes (independent samples t-test; chi-square). A two-sided p-value <0.05 was considered statistically significant.

All patients underwent complete neuro-ophthalmic assessment including tests of visual acuity, visual field, colour vision, pupil reactions, as well as ocular examination. Supplementary tests were dependent on the specific nature of the patient's visual impairment, and were chosen to investigate inconsistencies in the patient's performance.

The standard test of distance visual acuity was a Snellen chart at 6 m. The supplementary tests were:

a) Retesting of Snellen chart distance acuity at 3 and 1 m, the consistent result being ability to see lines further down the chart with decreased test distance whereas its absence indicates inconsistency.
b) Cardiff Acuity Test that utilises a single optotype, which removes clues to the patient of the level of acuity being tested, and is a preferential looking test, which allows the examiner to determine whether the optotype has been seen rather than relying on the patient to report if it has been seen, with comparison to Snellen chart distance acuity.
c) Mojon Optotype Chart in which angle of resolution is independent of optotype size such that the test distance is reduced until the patient sees the largest optotype and normally at that distance all the others also are seen whereas a need for the test distance to be reduced further for the smaller optotypes to be seen is inconsistent.
d) In patients with unilateral reduction of visual acuity, Snellen chart distance testing of visual acuity with both eyes (binocular) but with the good eye fogged with a high power lens should be the same as the visual acuity of the affected eye; better response indicates inconsistency in some cases to the extent that normal visual acuity may be shown to be present.

The standard test of visual field was Goldmann perimetry when the pointers towards inconsistency are progressively decreasing visual field as testing continues with the same size target (spiralling); same or very similar visual field to targets of widely different size (tubular visual field); boundary of visual field to a larger target crossing into the boundary of visual field to a smaller target (crossing isoptres); rapid oscillating fluctuation of the boundary of visual field to the same size target (star-shaped field); and wide fluctuation of results within a series of repeated tests. The supplementary visual field tests were:

a) Tangent screen test at 1 m and 2 m, the consistent result being increase with increased test distance whereas no change (tubular visual field) or decrease is inconsistent.
b) Confrontation test with a large object such as an A4 sheet of paper or even a white pillow when typically in MUVL the patient reports only a very small central field of vision.
c) Goldman or computerised perimetry with both eyes (binocular visual field) when a patient with a temporal hemianopia in one eye due to MUVL may have a complete hemianopia on the same side on binocular visual field test.
d) Comparison of the patient's navigational abilities to the results of visual field tests.

Normal imaging with MRI and/or CT of the head and orbits and normal visual electrodiagnostic testing were not required for diagnosing MUVL, but both usually were obtained.

3. Results

3.1. Patient characteristics

The mean age of symptom onset in cases was 37.5 years (SD = 14.3, range 13–61), significantly younger ($t = 2.2$, $p = .02$) than controls (45.5 years, SD = 19.6, range 14–81), with female predominance amongst the cases (72%; $\chi^2 = 5.9$, $p = .01$). Cases took longer (3.4 years, SD = 6.3) before diagnosis was reached, compared to controls (1.3 years, SD = 4.6) but not significantly dissimilar in the present cohort (all patients >18 years at diagnosis, $p = .08$; Table 1).

3.2. Outcomes

Amongst the range of primary clinical manifestations (Table 2), cases were markedly more likely to present with functional visual acuity reduction in both eyes (cf. unilateral; 41%; $\chi^2 = 6.1$, $p = .01$) compared to organic controls. No significant between-group differences were observed for presentations of field deficits, double vision, positive visual phenomena, ocular pain or headache. Functional cases had a lower