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Neoplastic Masquerade Syndromes among Uveitis Patients

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Abstract

Purpose—To identify the demographic and clinical characteristics, along with the frequency, of neoplastic masquerade syndromes in a tertiary uveitis clinic.

Design—A retrospective observational cohort.

Methods—Demographic and clinical data on all patients presenting to the National Eye Institute (NEI) with uveitis between 2004–2012 were used to compare neoplastic masquerade syndromes and uveitis.

Results—A total of 853 patients presenting with uveitis were identified. Of these, 21 (2.5%) were diagnosed with neoplastic masquerade syndromes. The average age at presentation of masquerade syndrome patients was 57 years (median: 55, range: 38–78), for uveitis 42 years (median: 43, range: 3–98) ($p=0.0003$). There were 48% females in the masquerade syndromes group, compared with 59% females in the uveitis group. African American patients represented 9% of the masquerade syndrome patients and 36% of uveitis patients ($p=0.01$). Mean worse eye visual acuity was 0.89 (20/160) in neoplastic masquerade syndromes, and 0.66 (20/100) in the uveitis group ($p=0.21$). Ninety percent of masquerade syndrome patients had posterior inflammation, compared with 63% of uveitis patients ($p=0.006$). Forty-eight percent of the masquerade syndrome patients had unilateral disease, compared with 27% of the uveitis patients ($p=0.04$).

Conclusion—Patients with neoplastic masquerade syndromes were more likely to be older, male, or non-African American and to have posterior segment inflammation and unilateral disease. Patients with masquerade syndromes also had worse visual acuity than did uveitis patients. These differences in clinical characteristics may help raise the suspicion for neoplastic masquerade syndromes.

Introduction

The term masquerade syndrome is classically used to describe those conditions which include as part of their manifestation the presence of intraocular infiltrating cells, but that are

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not due to immune-mediated uveitis entities.^{1,2} The term “masquerade syndrome” was first used in ophthalmology by Theodore in 1967 to describe a conjunctival carcinoma that presented as chronic conjunctivitis.³ Hematologic malignancies, retinoblastoma, retinal detachment or degeneration, and intraocular trauma are just a few of the disorders that may masquerade as uveitis, with intraocular lymphoma representing the most common neoplastic masquerade syndrome.^{1,4} Neoplastic masquerade syndromes are known to represent a minority of cases seen in uveitis clinics, yet specific information regarding their prevalence or clinical characteristics is scarce, at best.⁴ The importance of accurate and timely diagnosis cannot be overstated, as making the correct diagnosis can be life-saving.

The goal of this study was to identify the proportion of patients with masquerade syndromes in a tertiary uveitis clinic and determine the baseline clinical characteristics of patients with masquerade syndromes.

Materials and Methods

All patients presenting to the National Eye Institute (NEI) between 2004–2012 with the preliminary diagnosis of ‘uveitis’ were identified using a database search of the NEI’s electronic medical records. A standard screening protocol for uveitis was followed on each patient, including erythrocyte sedimentation rate, complete blood counts, chemistries, HLA-B27 typing, determination of serum angiotensin-converting enzyme levels, urine analysis, tuberculosis testing, and serologic tests for syphilis. Because of the tertiary nature of our clinic, non-malignant masquerade syndromes such as retinal detachment or trauma are very rare and were not included in our search. The clinical, ophthalmological, and laboratory data at baseline for the identified patients were collected in retrospective chart reviews. All patients were seen under an Institutional Review Board (National Institutes of Health) approved clinical research protocol.

The baseline clinical characteristics of the neoplastic masquerade syndrome patients, and the uveitis patients were gathered with particular attention to sex and age at the onset of symptoms and presentation, anatomic site of uveitis, posterior segment findings, presumed initial diagnosis, definitive diagnosis, interval between the onset of symptoms and the time of final diagnosis, the diagnostic procedures performed, the presence and anatomic location of intraocular inflammation, and the initial and final best-corrected visual acuity. The anatomic location and the activity of the inflammation were determined by using SUN criteria.⁵ The initial best-corrected visual acuity was calculated by taking the mean of the logMAR visual acuity in better and worse eyes for bilateral cases and in the affected eye for unilateral cases.

Demographic and clinical data are presented with descriptive statistics. Univariate comparisons of characteristics between neoplastic masquerade syndrome patients and uveitis patients were made by using two-sample t-tests for continuous variables and chi-square tests for categorical variables. A two-sided p value less than 0.05 was considered statistically significant. Because of the small sample size in the neoplastic masquerade group, a multivariate analysis was deemed inappropriate.

Results

A total of 853 patients with complete data, and “uveitis” or “ocular inflammation” as the presenting diagnosis were identified. The average follow-up time for these patients was 5 years $\pm$ 4 years (median: 4, range: 0–27 years). The National Eye Institute (NEI) is a tertiary care center which accepts referrals from all over the World of all ethnicities and ages. Nevertheless, most of the patients in this study were either Caucasian (46%) or African

American (36%) and were referred from the Mid-Atlantic region of the United States. Also, patients aged less than or equal to 21 years old represented 17% of our cohort. This is reflective of the average uveitis population where only 10–15% are children.⁶ Less than 5% of patients were excluded due to incomplete data or follow-up.

Of the 853 patients, 21 (2.5%) were identified as neoplastic masquerade syndromes. Eight hundred and thirty two cases were diagnosed as nonmasquerade uveitis cases, of which 62 (7%) had infectious uveitis and 770 (93%) had non-infectious autoimmune uveitis.

For patients with neoplastic masquerade syndromes, the average length of time between the onset of symptoms and definitive diagnosis was 8 months (median 7, range 1–28 months). The average time to diagnosis from presentation to NEI was 2 months. The most commonly identified neoplasm amongst the masquerade syndromes was primary intraocular/vitreoretinal lymphoma (PIOL/PVRL) (17 of 21, 81.1% of the neoplastic masquerade syndromes; 17 of 853, 2.0% of all cases of “uveitis”). Other masquerade syndromes were identified as follows: mucosal associated lymphoid tissue lymphoma of the optic nerve sheath (n=1), chronic lymphocytic leukemia (CLL) (n=1), human T-lymphotropic virus (HTLV) T cell Leukemia (n=1), and Non-Hodgkin’s Lymphoma (n=1). Fourteen of the 21 (66.7%) patients were diagnosed using diagnostic vitrectomy and/or ocular tissue biopsy (optic nerve sheath biopsy (n=1) or chorioretinal biopsy (n=1)). Twelve of the 14 vitrectomies were performed immediately upon referral, while the other 2 were followed first on immunosuppressants due to initial favorable response and were biopsied after 12 months of follow-up. Four of the 21 (19.1%) patients were diagnosed from brain biopsy (2) or lumbar puncture and cytology (2), and the other 3 (Non-Hodgkin lymphoma (NHL), Chronic lymphocytic leukemia (CLL) and Human T-lymphotropic virus (HTLV) leukemia) were diagnosed systemically (Table 1).

When compared with the non-masquerade uveitis patients, patients with masquerade syndromes were more likely to be older, with an average age of 57 years (range: 38–78 years), as opposed to 42 years (range: 3–98 years) in the uveitis group (mean difference = 15.0; 95% confidence interval [CI] for the difference, 6.9–23.1; $p=0.0003$). The proportion of patients 50 years or older was 95% in the masquerade group, compared with 38% in the uveitis group (odds ratio [OR], 32.7; 95% CI, 4.4–244.5; $p=0.0001$). No patient aged less than or equal to 21 years old was diagnosed with a neoplastic masquerade syndrome. A total of 48% (10/21) of the masquerade syndrome patients were female, compared with 59% (489/832) in the uveitis group (OR, 1.6; 95% CI, 0.7–3.7; $p=0.19$). Among the masquerade syndrome patients, 13/21 (62%) were Caucasian, compared with 389/832 (47%) in the uveitis group (OR, 1.9; 95% CI, 0.8–4.5; $p=0.13$). There were significantly fewer African American patients among the masquerade syndrome patients (2/21; 10%) compared with the uveitis group (301/832; 36%) (OR, 0.2; 95% CI, 0.04–0.80; $p=0.01$) (Table 2).

The baseline visual acuity in the worse and better seeing eyes was not significantly different for the masquerade syndrome patients (logMAR 0.89 and 0.29 in the worse and better eyes, respectively), compared with the uveitis group (logMAR 0.66 and 0.36 in the worse and better eyes, respectively) (mean difference, 0.23; 95% CI, –0.58–0.13; $p=0.21$ and mean difference, 0.1; 95% CI, –0.19–0.33; $p=0.59$, respectively).

Nineteen of the 21 patients with masquerade syndromes (90%) as opposed to 522 of the 832 uveitis patients (63%) presented with intermediate, posterior or panuveitis (OR, 0.2; 95% CI, 0.0–0.77; $p=0.006$). The proportion of patients with active intraocular disease – determined by the presence of cells in the anterior chamber or vitreous haze on exam at the initial visit – was almost identical between the two groups, with 14/21 (67%) among the masquerade syndrome patients and 560/832 (67%) for the uveitis group. The proportion of

individuals with unilateral disease – as opposed to bilateral disease – was higher in the masquerade group (10/21;48%) than in the uveitis group (222/832; 27%) (OR, 2.5; 95% CI, 1.05–6.0; $p=0.05$).

Discussion

Masquerade syndromes represent a rare but important proportion of uveitis cases. In this study, the frequency of neoplastic masquerade syndromes among patients with uveitis in a tertiary ophthalmologic center was 2.5%. This is similar to the only such report from Europe in which the authors found that 2.3% of their patients with an initial presenting diagnosis of uveitis were eventually diagnosed with a neoplastic masquerade syndrome.⁴ In our cohort, the majority of patients with neoplastic masquerade syndromes had primary intraocular/vitreoretinal lymphoma (81.1%). These findings are consistent with the literature, which reports that masquerade syndromes are rare entities, and that the most common type is intraocular lymphoma.¹ Of the several different types of lymphoma that may masquerade as uveitis, primary diffuse large B-cell malignant intraocular central nervous system NHL (also known as primary CNS lymphoma, PCNSL) is the most common.^{7,8} Rothova et al reported that 13 of the 19 (68.4%) neoplastic masquerade syndromes had PIOL/PVRL, which is lower than the proportion in our cohort. However, this may be explained by the particular referral patterns to our institute. The characteristic clinical manifestations of intraocular lymphoma include bilateral painless vision loss resulting from tumor infiltration of the vitreous, retina, and subretinal space, predominantly in elderly patients^{2,7,9} which usually manifests itself as vitritis and chorioretinal lesions on examination.^{10, 11} Patients usually complain of floaters and a mild decrease in vision, and classically present with a refractory uveitis.¹² Whether the incidence of PVRL/PCNSL has been increasing over the past decade has been under debate, regardless, this type of lymphoma is associated with poor survival.^{13,14} Thus, the need to be vigilant in screening for this condition, and differentiating it from uveitis cannot be overstated.

We found that neoplastic masquerade syndrome patients were older, had lower visual acuity at presentation, and were less likely to be female than were uveitis patients.⁴ Uveitis is known to have female preponderance in general, and that is not seen in masquerade syndromes. Rothova et al also showed a similar proportion of females (42.1%) in their neoplastic masquerade cohort. There was a slight Caucasian predominance among masquerade syndrome patients in our cohort. Although the previous report did not address this question, we found that African American patients were less likely than other races to be diagnosed with neoplastic masquerade syndromes. This is likely driven by the fact that the majority of neoplastic masquerade syndromes were PIOL/PVRL, which primarily affects Caucasians.⁷ We cannot determine if this is unique to our center or is reflective of the overall uveitic masquerade syndromes since race and ethnicity were not previously evaluated.

In terms of the location of inflammation, neoplastic masquerade syndromes are more likely than uveitis to present in the posterior segment. Similarly, Rothova et al also reported 79% (15 of the 19 cases) posterior segment involvement among their neoplastic masquerade cohort. Unilateral presentation was much more common among masquerade syndromes than uveitis in our cohort. A posterior presentation is consistent with the behavior of PIOL/PVRL, as previously described.^{2,7,9} Infectious or neoplastic entities are known to be more likely to be unilateral.¹⁵ Though our non-masquerade “uveitis” cohort also included infectious uveitides, possibly diluting the difference between masquerade and non-masquerade cohorts, a difference in laterality was still detectable between the two groups. Rothova et al noted that 57% of the non-neoplastic masquerade syndromes and 47% of the

neoplastic masquerade cases were unilateral but a comparison to the non-masquerade uveitis cohort was not made.⁴

The variation in time to diagnosis among our patients with masquerade syndromes may be attributable to the difference in methods of diagnosis. In our masquerade patients various methods were utilized before reaching the ultimate diagnosis, reflecting the complexity of such cases and the reality of clinical practice. Clinical suspicion, examination, and history taking all play a critical role in making the diagnosis. A standardized, non-invasive method for ruling out masquerade syndromes would be desirable. The use of immunophenotyping, cytologic examination, and molecular techniques such as IgH gene rearrangement by PCR amplification for ocular specimens can aid in the diagnosis.^{4,15} Currently the gold standard for diagnosing primary vitreoretinal lymphoma is the detection of malignant lymphoid cells in the retina, vitreous, and/or the optic nerve.^{17,18,19} Such is commonly achieved through aqueous aspiration and/or diagnostic vitrectomy, retinal or chorioretinal biopsy.¹⁹ Advanced diagnostic techniques (e.g. microdissection, immunophenotyping, PCR) have been utilized at the NEI since the early 2000s and did not change substantively throughout the duration of this study. In this cohort, the overwhelming majority of the patients with neoplastic masquerade syndromes were diagnosed through ocular fluid or ocular tissue analysis.

Approximately 40% of all uveitis cases remain idiopathic despite extensive searches for the causes of their intraocular inflammation.²⁰ The consequences of missing malignant or systemic nonmalignant etiologies of uveitis can be severe. Thus, for patients older than 50 years with unexplained vitritis and chorioretinal lesions (especially those with a partial or poor therapeutic response) evaluation for a possible neoplasm is crucial. Several studies evaluated the clinical characteristics of *de novo* uveitis in the elderly (>60 years of age) and found that neoplastic masquerade syndromes constituted 1.5%-5.3%.^{21,22,23} Even though neoplastic masquerade syndromes are not the leading cause of uveitis in the elderly, the majority of these conditions occur in patients older than 50 years. It is important to note that almost all patients (95%) in our neoplastic masquerade syndrome cohort were 50 years or older.^{13,24,25} Additionally, as with most rare diseases, these patients may benefit from the expertise of a tertiary care center as highlighted by the shorter time interval from presentation to our tertiary care to ultimate diagnosis. It is the hope that if such cases are properly handled, an ophthalmologist may be the first to recognize systemic malignancy or accurately identify the nonmalignant systemic disease, thus preserving quality of life and preventing vision loss.

The results of this study should be reviewed with caution as with all retrospective studies. The major limitation of our study is the tertiary nature of our clinic, which may have led to potential biases in referral patterns. Due to specialized care and research interests, many patients are referred to the NIH for diagnosis and management of intra-ocular lymphomas, which may have led to an overestimation of the proportion of such cases among masquerade syndromes. Even if the proportion of patients who were ultimately diagnosed with intraocular lymphoma were disproportionately high when compared to the population at large, this would have little impact on the risk factors for the disease which are the focus of this paper.

Also, though the entire cohort is quite large, there were only 21 patients in the neoplastic masquerade group. More cases of neoplastic masquerade syndromes would have provided more power to detect clinical differences. It is possible that some of the associations we found could have occurred by chance due to multiple comparisons in a small cohort. Nevertheless, this study is the first study to evaluate the clinical characteristics of neoplastic masquerade syndromes in comparison to uveitis patients. And while the findings may not be broadly generalizable to the general population, these findings may be applicable to tertiary

uveitis clinics. Also, the referral patterns should not significantly affect the differences in clinical manifestations between masquerade and non-masquerade uveitis patients. Our referral pattern has a bias towards posterior segment uveitis, though such a bias would presumably underestimate – rather than overestimate – the difference in anatomic location of inflammation found between the two groups.

In conclusion, there exist valuable, distinguishable differences between the presenting characteristics of patients with neoplastic masquerade syndromes and those with uveitis. Patients with neoplastic masquerade syndromes were significantly more likely to be older, of non-African American race, and to have unilateral disease and posterior segment involvement at presentation. Diagnosis of masquerade syndromes is difficult, but necessary, and clinical characteristics may help raise suspicion and help differentiate neoplastic masquerade syndromes from uveitis.

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Table 1

Clinical characteristics of patients with ocular neoplastic masquerade syndromes at presentation

No.	Age	Sex	Visual Acuity (LogMAR)*	Time** to Final Diagnosis (months)	Final Diagnosis	Anatomic Location
1	57	F	0.30	14	PIOL/PVRL	Panuveitis OU
2	53	F	0.60	10	MALT Lymphoma	Panuveitis OD
3	50	F	0.10	2	CLL	Anterior OD
4	62	F	0.70	12	PIOL/PVRL	Panuveitis OD
5	56	F	3.00	5	PIOL/PVRL	Panuveitis OU
6	55	M	0.20	1	PIOL/PVRL	Panuveitis OU
7	50	F	1.60	17	PIOL/PVRL	Panuveitis OU
8	55	M	0.00	6	PIOL/PVRL	Posterior OS
9	79	F	0.50	9	PIOL/PVRL	Intermediate OS
10	54	F	0.20	1	HTLV Leukemia	Intermediate OU
11	53	M	0.10	15	Systemic NHL	Posterior OS
12	58	F	3.00	9	PIOL/PVRL	Panuveitis OU
13	52	M	0.00	8	PIOL/PVRL	Posterior OU
14	55	F	0.10	3	PIOL/PVRL	Intermediate OS
15	67	M	2.00	28	PIOL/PVRL	Panuveitis OU
16	71	M	0.50	1	PIOL/PVRL	Panuveitis OU
17	42	M	0.60	1	PIOL/PVRL	Panuveitis OD
18	68	M	0.90	5	PIOL/PVRL	Anterior OS
19	62	M	0.10	4	PIOL/PVRL	Posterior OD
20	78	M	1.40	4	PIOL/PVRL (T cell)	Intermediate OS
21	49	M	3.00	15	PIOL/PVRL	Posterior OU

PIOL/PVRL: primary intraocular/vitreoretinal lymphoma, MALT Lymphoma: mucosa associated lymphoid tissue lymphoma, CLL: chronic lymphocytic leukemia, HTLV: Human T-lymphotropic virus, NHL: Non-Hodgkin lymphoma, M= Male, F= Female, OD= Right Eye, OS= Left Eye, OU= Both Eyes

* Visual acuity of the affected eye was used in unilateral cases and the worse eye in bilateral cases.

** Represents the time elapsed between the onset of symptoms and the definitive diagnosis.

Table 2
Baseline characteristics of patients with ocular neoplastic masquerade syndromes and non-masquerade uveitis

	Masquerade Syndromes N (%)	Uveitis N (%)	Mean Diff. or Odds Ratio	95% CI	P value
Number of Cases	21 (2.5%)	832 (97.5%)			
Age at Onset (yrs) Mean (median; range)	57 (55; 38–78)	42 (43;3–98)	15.0	6.9, 23.1	0.0003
Female	10 (47.6%)	489 (58.8%)	OR: 1.6	0.7, 3.7	0.19
Visual Acuity mean logMAR (Snellen)	0.89(20/160)	0.66(20/100)	0.2	−0.1, 0.6	0.21
Location of Inflammation: -Anterior Segment -Posterior Segment *	2 (10%) 19 (90%)	310 (37%) 522 (63%)	OR: 0.2	0.041, 0.8	0.006
Active intraocular inflammation	14 (67%)	560 (67%)	OR: 1.0	0.4, 2.4	0.8
Unilateral disease	10 (48%)	222 (27%)	OR: 2.5	1.0, 6.0	0.045
Race/ethnicity: -Caucasian: -African American: -Hispanic and other:	13 (62%) 2 (10%) 6 (28%)	389 (47%) 301 (36%)** 142 (17%)	OR: 1.9 OR: 0.2 OR: 1.9	0.8, 4.5 0.043, 0.8 0.7, 5.1	0.13 0.01 0.24

N = number of cases; (%) = percentage of the whole patient population; OR = Odds Ratio; 95% CI = 95% Confidence Interval

* Includes intermediate, posterior, and panuveitis

** Reflects a comparison between the proportion of African American and non-African American patients in each group