Intraocular Schwannoma: Case Series of 28 Patients and Literature Review

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Research

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Abstract

Background: Intraocular schwannoma is a rare intraocular tumor, which is often misdiagnosed. We aimed to analyze the demographics and clinical characteristics of patients with intraocular schwannoma.

Methods: Retrospective case series were collected from May 2005 to July 2021 in Beijing Tongren Hospital. Then a literature review was also performed.

Results: A total of 28 patients were diagnosed with intraocular schwannoma histopathologically. The median age (range) of the included patients was 39 (12-64) years old, among whom half subjects were female. The most common symptom was visual loss (75.0%), followed by visual field loss (10.7%). Intraocular schwannoma presented as nonpigmented mass, which occurred mainly in ciliary body (42.9%), followed by choroid (32.1%) and ciliochoroid (25.0%). 16 patients (57.1%) were clinically misdiagnosed as uveal melanoma. Tumor excision was performed for all patients and increased light transmission was detected in half cases. In the consecutive follow-up (median: 6.0 years, range: 0.5-16.0 years), no recurrence or metastasis case was detected.

Conclusions: Intraocular schwannoma is a rare benign intraocular tumor. It usually presents as nonpigmented mass, which is easily misdiagnosed as nonpigmented uveal melanoma.

Background

Schwannoma, also known as neurilemmoma or neurinoma, is a benign tumor that originates from the peripheral-nerve sheath [1]. Schwannoma is a rarely seen intraocular tumor that occurs in ciliary body, choroid, and occasionally in the iris [2–3]. They are rarely pigmented, while usually amelanotic or pseudo-pigmented when covered by retinal pigment epithelium. However, it is difficult to distinguish schwannoma from amelanotic malignant melanoma by clinical examination alone. To our knowledge, malignant transformation of an intraocular schwannoma has not yet been reported. Although this type of tumor is cytologically benign and not life threatening, appropriate treatment is required to prevent visual loss and its progressive enlargement. In this study, we reviewed our experience dealing with 28 intraocular schwannoma patients, which contains the largest sample size so far. We aimed to describe the clinical features, relevant histopathology, and treatment options of intraocular schwannoma.

Methods And Materials

Data collection

We retrospectively reviewed the clinical data and histopathological materials from all patients with documented intraocular schwannoma, who were diagnosed in Beijing Tongren Hospital from May 2005 to July 2021. Patients’ age, sex, symptoms, systemic diseases, tumor locations, preoperative clinical diagnoses, and treatment procedures were all recorded. Hematoxylin-eosin-stained slides were available in all cases, as well as special stains such as periodic acid-Shiff (PAS) and Masson. Immunohistochemical studies were also performed based on formalin-fixed and paraffin-embedded tissue. The monoclonal antibodies used were against Leu-7, Vimentin, S-100, smooth muscle actin (SMA), Melan-A, human melanoma black-45 (HMB-45), Ki-67, and neuron-specific enolase (NSE). This study was approved by the Institutional Review Board of Beijing Tongren Hospital.

Literature review

A literature search was performed in electronic databases including PubMed, Embase, Web of Science, and Cochrane Library using the following terms: "schwannoma", "intraocular schwannoma", "neurilemmoma", "neurinoma", "uveal schwannoma", "choroidal schwannoma". The publications were limited for human studies and manuscripts written in English. The last search date was July 1st, 2021. All identified full-text references were reviewed by two authors (LD and HYL). After selection, references with relative information and available data were included in the literature review.
Results

The basic characteristics of the included patients were presented in Table 1. A total of 28 patients were histopathologically diagnosed as intraocular schwannoma. The median age (range) of all of the patients was 39 (12–64) years old, among whom half cases were female. Visual loss was the most common symptoms (75.0%), followed by visual field loss (10.7%). Almost all intraocular schwannoma (27/28) presented as nonpigmented mass (Fig. 1), which occurred mainly in ciliary body (42.9%), followed by choroid (32.1%) and ciliochoroid (25.0%). An adjacent retinal detachment was common (Fig. 2). Fundus fluorescein angiography and indocyanine green angiography revealed multiple hyperfluorescent areas in the neoplasm in the early phase followed by its marked staining in the late phase (Fig. 3). In magnetic resonance imaging (MRI), intraocular schwannoma often showed equal signal intensity in T1-weighted images and low signal intensity in T2-weighted images (Fig. 4), while in ultrasonography the tumor usually presented as mushroom mass with relatively medium internal reflectivity (Fig. 5). For tumors located in anterior segment, ultrasonic biological microscopy always revealed a medium internal reflectivity in ciliary body, with iris and lens affected (Fig. 6). 16 patients (57.1%) had history of clinically misdiagnosis as uveal melanoma. Tumor excision was performed for all patients by senior author WBW, and increased light transmission of the tumor was detected in 50.0% cases during the surgery. Consecutive follow-up was conducted for all patients, with the median of 6.0 (0.5–16.0) years. No recurrence or metastasis case was found in any included patients.
Table 1
Characteristics of the included patients.

| Case  | Year | Sex | Age (year) | Symptom (duration) | Tumor location | Clinical diagnosis | Treatment | Tumor size (mm) | Light transmission |
|-------|------|-----|------------|-------------------|----------------|-------------------|-----------|-----------------|-------------------|
| Case 1 | 2005 | M   | 28         | Visual field loss (4 mos) | Ciliochoroid | Uveal melanoma | Excision | 18×15×12 | Increased |
| Case 2 | 2008 | F   | 54         | Visual loss (6 yrs) | Choroid      | Unknown          | Excision | 10×8×7 | Unknown |
| Case 3 | 2009 | M   | 46         | Visual loss (5 yrs) | Choroid      | Uveal melanoma  | Excision | 11×8×5 | Increased |
| Case 4 | 2010 | M   | 22         | Visual loss (3 weeks) | Ciliochoroid | Unknown          | Excision | Unknown | Increased |
| Case 5 | 2010 | F   | 32         | Visual loss (3 mos) | Ciliary body | Unknown          | Excision | 8×6×5 | Increased |
| Case 6 | 2010 | F   | 49         | Visual loss (2 yrs) | Ciliary body | Unknown          | Excision | 8×6×5 | Unknown |
| Case 7 | 2011 | M   | 57         | Mass (1 mo)        | Ciliary body | Uveal melanoma  | Excision | 9×7×6 | Increased |
| Case 8 | 2012 | F   | 20         | Visual field loss (2 mos) | Ciliochoroid | Uveal melanoma  | Excision | 15×13×7 | Unknown |
| Case 9 | 2012 | M   | 34         | Visual loss (18 days) | Choroid      | Uveal melanoma  | Excision | 8×7×7 | Unknown |
| Case 10 | 2014 | M   | 45         | Diplopia (2 yrs)   | Ciliary body | Unknown          | Excision | 9×9×5 | Unknown |
| Case 11 | 2014 | M   | 32         | Visual loss (6 mos) | Choroid      | Uveal melanoma  | Excision | 9×8×6 | Unknown |
| Case 12 | 2015 | F   | 36         | Visual loss (2 mos) | Ciliary body | Uveal melanoma  | Excision | 15×8×7 | Increased |
| Case 13 | 2015 | F   | 36         | Visual loss (1 mo) | Choroid      | Uveal melanoma  | Excision | 10×8×5 | Increased |
| Case 14 | 2015 | F   | 50         | Mass (1 year)      | Sclera and ciliochoroid | Uveal melanoma | Excision | 16×15×8 | Increased |
| Case 15 | 2015 | M   | 46         | Visual loss (3 mos) | Ciliary body | Uveal melanoma  | Excision | 13×8×7 | Increased |
| Case 16 | 2015 | F   | 27         | Visual loss (3 mos) | Choroid      | Unknown          | Excision | 12×8×7 | Unknown |
| Case 17 | 2016 | M   | 32         | Visual loss (6 mos) | Ciliary body | Hemangioma       | Excision | 14×8×7 | Unknown |
| Case 18 | 2016 | F   | 40         | Visual loss (20 days) | Ciliochoroid | Unknown          | Excision | 13×11×9 | Unknown |
| Case 19 | 2017 | F   | 46         | Visual loss (2 mos) | Ciliary body | Uveal melanoma  | Excision | 15×11×10 | Unknown |
| Case 20 | 2017 | F   | 38         | Visual field loss (3 mos) | Ciliary body | Hemangioma       | Excision | 11×8×5 | Unknown |
| Case 21 | 2017 | F   | 23         | Visual loss (2 mos) | Ciliary body | Unknown          | Excision | 13×7×7 | Increased |
| Number | Year | Sex | Age (year) | Symptom (duration) | Tumor location | Clinical diagnosis | Treatment | Tumor size (mm) | Light transmission |
|--------|------|-----|------------|-------------------|----------------|-------------------|-----------|----------------|-------------------|
| Case 22 | 2017 | F   | 38         | Visual loss (6 mos) | Ciliochoroid | Uveal melanoma | Excision | 15×12×10 | Increased |
| Case 23 | 2018 | M   | 64         | Visual loss (3 mos) | Choroid | Uveal melanoma | Excision | 10×10×4 | Unknown |
| Case 24 | 2018 | M   | 45         | Visual loss (1 yrs) | Ciliochoroid | Uveal melanoma | Excision | 6×5×5 | Unknown |
| Case 25 | 2018 | F   | 12         | Mass (2 mos) | Ciliary body | Uveal melanoma | Excision | 19×15×9 | Unknown |
| Case 26 | 2019 | M   | 47         | Visual loss (1 mo) | Choroid | Unknown | Excision | 3×3×3 | Unknown |
| Case 27 | 2019 | M   | 43         | Visual loss (1 mo) | Choroid | Uveal melanoma | Excision | 7×6×6 | Increased |
| Case 28 | 2021 | M   | 48         | Visual loss (1 mo) | Ciliary body | Unknown | Excision | 10×7×5 | Unknown |

Immunohistochemical studies were performed in 26 cases. Most cases diffusely immunoreacted with antibodies to S-100 protein and Vimentin (Fig. 7). There was no immunoreactivity for muscle marker SMA or melanocytic markers (Melan-A and HMB45). Immunostaining with the proliferation marker Ki-67 labeled less than 1% of the neoplastic nuclei.

The literature search identified 36 references with 49 cases after removal of duplicates [2, 4–38]. A brief summary of the main findings in publications was presented in Table 2. The median age (range) was 33 (0.5–76) years old, and 32 patients (65.3%) were female. Visual loss was the most common symptom (67.3%). The neoplasma occurred mostly in choroid (53.1%), followed by ciliary body (28.6%), and ciliochoroid (18.4%).
Table 2
Summary of reported intraocular schwannoma

| Author      | Age (year) | Sex | Symptom                | Tumor Location | Tumor Size (mm) | Light Transmission | Treatment     | Follow-up |
|-------------|------------|-----|------------------------|----------------|-----------------|-------------------|---------------|-----------|
| Cho [4]     | 30         | F   | Elevated IOP           | Choroid        | 30×10×10        | N/A               | Excision     | N/A       |
| Damato [5]  | 28         | F   | Visual loss            | Ciliochoroid   | N/A             | N/A               | Enucleation  | N/A       |
|             | 37         | F   | Visual loss            | Choroid        | 6.9×5.8×2.1     | N/A               | Biopsy       | 11 years  |
|             | 33         | M   | Visual loss            | Choroid        | 7.5×6.9×3.9     | N/A               | Biopsy       | 11 years  |
|             | 45         | M   | Visual loss            | Choroid        | 18×18×10        | N/A               | Biopsy       | 1 year    |
|             | 15         | M   | Visual loss            | Choroid        | 12×12×3.6       | N/A               | Biopsy       | 6 years   |
| Donovan [6] | 68         | F   | Visual loss            | Ciliary body   | 6×10            | Increased         | Enucleation  | N/A       |
| Fan [7]     | 21         | M   | Visual loss            | Choroid        | 13×13×6         | Decreased         | Enucleation  | N/A       |
| Freedman [8]| 31         | M   | Visual loss            | Choroid        | 24×24×23.5      | N/A               | Enucleation  | N/A       |
| Goto [9]    | 19         | F   | Mass                   | Ciliary body   | 7×7×9           | N/A               | Excision     | 4 years   |
| Graham [10] | 11         | F   | Mass                   | Choroid        | 19×7×7          | Increased         | Excision     | 2 years   |
| Huang [11]  | 37         | F   | Visual loss            | Choroid        | 10×9×5          | N/A               | Excision     | 1 week    |
| Hufnage [12]| 57         | F   | Visual field loss      | Ciliary body   | 7.5×9.5         | Increased         | Excision     | 15 years  |
| Jajapuram [13]| 56        | M   | Visual loss            | Choroid        | 4.5×4.5×6       | N/A               | Enucleation  | N/A       |
|             | 33         | F   | Visual loss            | Choroid        | 10×6            | N/A               | Enucleation  | N/A       |
| John [14]   | 0.5        | F   | Intermittent esotropia | Choroid        | 5×12.5          | N/A               | Enucleation  | N/A       |
| Kalik [15]  | 58         | M   | N/A                    | Choroid        | N/A             | N/A               | Enucleation  | 15 weeks  |
| Kim [16]    | 39         | F   | Visual loss            | Ciliary body   | 17×17×22        | N/A               | Enucleation  | N/A       |
| Kiratli [17]| 11         | M   | Visual loss            | Ciliary body   | 15×15×7         | N/A               | Enucleation  | N/A       |
| Kuchle [18] | 26         | M   | Visual loss            | Ciliary body   | 5×5             | N/A               | Excision     | N/A       |
| Lee [19]    | 74         | F   | Visual loss and proptosis| Choroid       | 10×12.2×12.1    | N/A               | Enucleation  | 5 years   |
| Matsuo [20] | 73         | F   | Eye irritation          | Choroid        | N/A             | N/A               | Enucleation  | N/A       |
| McLaughlin [21]| 34        | F   | Mass                   | Intrascleral   | 2               | N/A               | Autopsy      | N/A       |
| Mortuza [22]| 58         | M   | Eye pain               | Entire globe   | 2.3×2×2         | N/A               | Enucleation  | N/A       |
| Nair [23]   | 12         | M   | Subconjunctival mass   | Entire globe   | N/A             | N/A               | Excision     | 26 months |

F: female, M: male, IOP: intraocular pressure, N/A: not available.
| Author          | Age (year) | Sex | Symptom                        | Tumor Location | Tumor Size (mm) | Light Transmission | Treatment    | Follow-up |
|-----------------|------------|-----|--------------------------------|----------------|-----------------|-------------------|-------------|-----------|
| Packard [24]    | 43         | F   | Visual loss                    | Choroid        | N/A             | N/A               | Enucleation  | N/A       |
| Pineda [25]     | 46         | M   | Mass                           | Ciliary body   | 2×1×1           | Increased         | Enucleation  | N/A       |
| Quintana [26]   | 76         | F   | Proptosis                      | Choroid        | 25×23×22        | N/A               | Enucleation  | N/A       |
| Rosso [27]      | 40         | F   | Visual loss                    | Choroid        | 15              | Increased         | Enucleation  | N/A       |
| Saavedra [28]   | 9          | F   | Mass and visual loss           | Iris and ciliochoroid | 6×3     | N/A               | Enucleation  | 7 years   |
| Shields [29]    | 70         | F   | Mass                           | Choroid        | 9×9×3            | Decreased         | Excision    | 6 months  |
| Shields [30]    | 14         | F   | Visual loss                    | Choroid        | 14×3            | Decreased         | Enucleation  | N/A       |
| Shields [31]    | 30         | M   | Ptosis                         | Choroid        | 7×6×3.5         | N/A               | Enucleation  | N/A       |
| Smith [32]      | 30         | F   | Visual loss                    | Choroid        | 15×11×15        | Increased         | Enucleation  | N/A       |
| Swan [33]       | 32         | F   | Visual loss                    | Choroid        | Cherry          | N/A               | Enucleation  | N/A       |
| Thaller [34]    | 28         | F   | Visual loss                    | Ciliary body   | N/A             | Increased         | Enucleation  | N/A       |
| Turell [35]     | 47         | M   | Visual loss                    | Ciliochoroid   | 17×15×11.4      | Decreased         | Excision    | N/A       |
| Udyaver [36]    | 19         | M   | Episceral mass                 | Ciliary body   | 13×13×11.5      | Increased         | Biopsy      | 4 months  |
| Xian [37]       | 63         | F   | Visual loss                    | Ciliary body   | 8×6×6           | N/A               | Excision    | N/A       |
|                 | 38         | F   | Visual loss                    | Ciliochoroid   | 10×10×10        | N/A               | Excision    | N/A       |
|                 | 36         | F   | Visual loss                    | Choroid        | 13×10×9         | N/A               | Enucleation  | N/A       |
|                 | 22         | F   | Visual loss                    | Ciliochoroid   | 16×13×11        | N/A               | Excision    | N/A       |
|                 | 19         | F   | Visual loss                    | Ciliochoroid   | 15×13×12        | N/A               | Excision    | N/A       |
|                 | 37         | F   | Visual loss                    | Ciliochoroid   | 27×15×12        | N/A               | Excision    | N/A       |
| You [2]         | 23         | F   | Visual loss                    | Choroid        | 12.6×15.5       | Increased         | Enucleation  | 11 months |
| Yu [38]         | 25         | M   | Visual loss                    | Ciliochoroid   | 10.1×8.8        | N/A               | Excision    | N/A       |
|                 | 48         | F   | Visual loss                    | Ciliary body   | 16.7×11.2       | N/A               | Excision    | 3 months  |
|                 | 30         | M   | Visual loss                    | Ciliary body   | N/A             | N/A               | Excision    | 16 months |
|                 | 18         | F   | Visual loss                    | Iris           | N/A             | N/A               | Excision    | 6 months  |

F: female, M: male, IOP: intraocular pressure, N/A: not available.

**Discussion**

Intraocular schwannoma is a rare and benign peripheral nerve neoplasma that usually appears as solitary and amelanotic lesion in the ciliary body or choroid. So far, a total of 49 cases of intraocular schwannoma have been reported, and we have collected another 28 cases, which contains the largest sample size in a single center. Among all these 77 cases, intraocular
schwannoma occurs more often in female than male with a ratio of 2:1. This trend is also consistent with schwannomas that occur in other parts of human bodies [39].

In our study, choroidal schwannoma was often clinically misdiagnosed as amelanotic melanoma, because of the similar shape and color. Even MRI and ultrasonography cannot provide distinguished characteristics exactly. However, we still considered they were more likely to be benign tumors such as schwannoma, neurofibromatosis, or hemangioma. Therefore, we chose to excise the mass with vitrectomy, and the histopathologic findings confirmed our speculation.

Considering with the experience of previously reported cases, it is very difficult to differentiate intraocular schwannoma with amelanotic melanoma, hemangioma, and metastatic carcinoma. Clinical features, ultrasonography, and MRI findings in schwannoma can only provide limited clues to identify those tumors. It is often impossible to make a definite diagnosis before histopathologic examination, which makes it difficult to choose treatment strategies. Surgical excision or tissue biopsy may be a better choice when diagnosing an amelanotic neoplasm. For those cases with huge intraocular mass and severe vision loss, enucleation can also be considered. Immunohistochemistry based on histopathological slide is helpful in the diagnosis of intraocular schwannoma, especially in differentiating with uveal melanoma and neurofibroma. Schwannoma diffusely immunoreacts with antibodies to S-100 protein and frequently expresses Vimentin and Leu-7 [40].

Generally, the prognosis of intraocular schwannoma is favorable. Malignant change of schwannoma is rare, so that it has not been reported in any intraocular location. Only one case was reported with recurrence after local excision [12].

In conclusion, intraocular schwannoma is a rare and benign tumor, which is often misdiagnosed as uveal melanoma. Therefore, cautions should be taken when diagnosing an amelanotic intraocular mass.

**Conclusion**

Intraocular schwannoma is a rare benign intraocular tumor. It usually presents as nonpigmented mass, which is easily misdiagnosed as nonpigmented uveal melanoma.

**Abbreviations**

PAS: periodic acid-Shiff, SMA: smooth muscle actin, HMB-45: human melanoma black-45, NSE: neuron-specific enolase, Melan-A and HMB45: melanocytic markers.

**Declarations**

**Ethics approval and consent to participants**

Ethics approval and informed consent were not required for this study because of public accessibility to the data.

**Consent for publication**

Not applicable.

**Availability of data**

Not applicable.

**Competing interest**

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Authors’ contributions

W.B. Wei, Y.M Li and L. Dong designed the study, Y.M Li, L. Dong and X.L Xu wrote the manuscript. H.Y Li, Q.Yang, and R.H Zhang collected the data and conducted the analyses, W.B. Wei edited and revised the manuscript. All authors have approved the submitted version and agreed with the contributions declarations.

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Figures
Figure 1

Appearance of intraocular schwannoma of Case 10 (A), Case 20 (B), and Case 25 (C) that located in ciliary body. Blue arrow: tumor.

Figure 2
Ultra-widefield fundus image of intraocular schwannoma of Case 24, and an adjacent retinal detachment was also noted.

Figure 3

Fundus fluorescein angiography (A) and indocyanine green choroidal angiography (B) of Case 24.

Figure 4

Magnetic resonance imaging (MRI) showed equal signal intensity in T1-weighted images and low signal intensity in T2-weighted images of intraocular schwannoma of Case 27.
Figure 5

Ultrasonography showed a mushroom mass with relatively medium internal reflectivity of Case 21.
Figure 6

Ultrasonic biological microscopy of case 28 revealed a medium internal reflectivity in ciliary body, with iris and lens affected.

Figure 7

Hematoxylin-eosin-stained slide (A) with immunohistochemical examination of S-100 (B) and Vimentin (C) of Case 28.