Vascular Tumors of the Iris in 45 Patients

The 2009 Helen Keller Lecture

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Objective: To report on a series of vascular tumors of the iris.

Design: Noncomparative case series. A retrospective medical record review of all patients with an iris vascular tumor was performed to identify the clinical features and develop a simple classification of these lesions. Included were demographics, clinical features, systemic associations, complications, management, and histopathology.

Results: There were 54 eyes in 45 patients with an iris vascular tumor. These were categorized as racemose hemangioma (41 eyes: 29 simple and 12 complex), cavernous hemangioma (3 eyes: 2 localized and 1 systemic), capillary hemangioma (1 eye, localized), varix (3 eyes, localized), and microhemangiomatosis (6 eyes, localized). The hemangiomas occurred in adults at a median age of 55 years, whereas capillary hemangioma occurred in infancy and cavernous hemangioma with systemic involvement occurred in a child. Of the 41 eyes with iris racemose hemangioma, none showed systemic involvement. Of all 54 eyes, transient hyphema was the main complication, found at some point in 30% or more of each affected eye except for iris capillary and racemose hemangioma. Surgical resection was necessary in 1 cavernous hemangioma and 1 varix. The remainder were managed with observation.

Conclusions: There are now well-documented examples of iris racemose hemangioma, cavernous hemangioma, capillary hemangioma, varix, and microhemangiomatosis. Transient hyphema is the main complication. Observation is usually advised. Most are solitary lesions confined to the iris and some (cavernous hemangioma and microhemangiomatosis) can have important systemic associations.

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THE EXISTENCE OF TRUE IRIS vascular tumors was once questioned. In 1972, Ferry[1] reviewed reported cases of histopathologically proven iris and ciliary body hemangiomas on file at the Armed Forces Institute of Pathology and other sources. He came to the surprising conclusion that all cases had been misdiagnosed histopathologically and they actually represented vascular melanoma, juvenile xanthogranuloma, florid iris neovascularization, or other lesions.[1] He raised doubts about the existence of iris hemangioma and proposed that most reported cases in humans were not hemangiomas. Since that time, however, there have been several reports of iris hemangiomas.[2-34]

Since the inception of our oncology service in 1974, we have coded and filed all cases of iris vascular tumors and pseudotumors examined by our team. In our literature review and personal experience, there have been genuine cases of iris vascular tumors and there are some that remain controversial.[2-52] In a recent case report of an iris varix, Broaddus and associates[3] provided a review on iris vascular tumors. The purpose of this contribution is to describe a series of iris vascular tumors seen on an ocular oncology service and to review the English language literature of the subject.

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METHODS

This study was approved by the institutional review board of the Wills Eye Institute of Thomas Jefferson University. A classification was designed as listed below. Although not all of these conditions represent true tumors in the classic sense of a distinct mass, they were included in the classification of vascular tumors in this communication, in keeping with classic usage in the literature.

Table 1. Patients With Vascular Tumors of the Iris

Tumor Type	No.		
	Patients	Eyes	Bilateral
Racemose	35	41	6
Cavernous	3	3	0
Capillary	1	1	0
Varix	3	3	0
Microhemangiomatosis	3	6	3
Total	**45**	**54**	**9**

Table 2. Vascular Tumors of the Iris by Category

Tumor Type	No. of Eyes
Racemose	41
Simple	29
Complex	12
Cavernous	3
Localized	2
Systemic	1
Capillary	1
Localized	1
Systemic	0
Varix	3
Localized	3
Systemic	0
Microhemangiomatosis	6
Localized	6
Systemic	0
Total	**54**

Based on our prior observations and literature review, we classified iris vascular tumors and pseudotumors into the following categories:

1. Racemose hemangioma
   Simple
   Complex
2. Cavernous hemangioma
   Localized
   Systemic
3. Capillary hemangioma
   Localized
   Systemic
4. Varix
   Localized
   Systemic
5. Microhemangiomatosis
   Localized
   Systemic

Using this classification, a retrospective medical record review was conducted on all patients coded with 1 of these diagnoses, and the patient, ocular, and tumor features were evaluated. The patient features that were recorded included age at diagnosis, race, sex, symptoms, and presence of related systemic hemangiomas or syndromes. The ocular features recorded included iris color, visual acuity, and intraocular pressure. The tumor features that were recorded included referring diagnosis, tumor quadrant location, size (base in millimeters and clock-hours), blood vessel feeding from the sclera or iris, and intrinsic blood vessels. Related ocular findings such as glaucoma, cataract, hyphema, and corneal changes were listed. Medical and surgical intervention were reviewed and the ultimate outcome was listed.

Table 3. Referring Diagnosis in 54 Eyes of 45 Patients With Iris Hemangioma

Referring Diagnosis	No. (%)
Iris	
Hemangioma	8 (15)
Melanoma	2 (4)
Nevus	5 (9)
Cyst	1 (2)
Prominent vessels nonspecific	6 (11)
Ciliary body melanoma	2 (4)
Choroidal melanoma	2 (4)
No diagnosis submitted	27 (50)

RESULTS

There were 54 eyes in 45 patients with an iris vascular tumor (**Table 1**). The tumors were classified as racemose hemangioma in 41 eyes of 35 patients (29 simple and 12 complex), cavernous hemangioma in 3 eyes of 3 patients (2 localized and 1 systemic), capillary hemangioma in 1 eye of 1 patient, localized varix in 3 eyes of 3 patients, and microhemangiomatosis in 6 eyes of 3 patients (**Table 2**). Histopathologic confirmation of the diagnosis was available in a single case of solitary cavernous hemangioma and a single case of varix, both following removal of the lesion by iridectomy. The referral diagnoses are shown in **Table 3**. The diagnosis of iris hemangioma was suspected in 8 of the 54 eyes and no diagnosis was submitted in 27 (50%) cases. Uveal melanoma (iris, ciliary body, and choroid) was the referral diagnosis in 6 cases.

The demographic data are shown in **Table 4**. The median patient age was 55 years at original examination in all cases with the exception of iris capillary hemangioma (0.1 year) and cavernous hemangioma with systemic involvement (7 years). All but 1 tumor was found in white patients. The iris color was blue or green in 48 cases (89%). The intraocular pressure was under 22 mm Hg except for 1 patient with open-angle glaucoma and a pressure of 37 mm Hg. Ectropion was absent in all cases. Corectopia was found in 1 case of iris varix and 1 case of solitary cavernous hemangioma. Individual tumor features are shown in Table 4 and **Table 5**, and specific information per tumor classification will be discussed.

Racemose hemangioma (arteriovenous communication) was the most common lesion in our series, accounting for 35 of the 45 patients. In 29 patients, it was unilateral and in 6 patients bilateral. It appeared as a solitary large tortuous iris blood vessel, approximately twice the normal caliber and located within the iris stroma. The racemose hemangioma intertwined in a sinuous fashion within the iris stroma from iris root to the pupillary border and then back to the root.[8-14] In 29 patients, the racemose hemangioma was classified as simple, characterized by a single tortuous vessel (**Figure 1** A and B). Six of the simple racemose hemangiomas were bilateral. In 12 cases, it was classified as complex, characterized by multiple ramifying, intertwining blood vessels forming a sessile vascular mass (Figure 1C and D).[13] The complex cases were all unilateral. Racemose hemangioma was

Table 4. Iris Hemangioma in 54 Eyes of 45 Patients

Characteristic	No. (%) by Hemangioma Type						
	Racemose		Cavernous				
	Simple (n=29)	Complex (n=12)	Solitary (n=2)	Systemic (n=1)	Capillary, Localized (n=1)	Varix (n=3)	Microhemangiomatosis, Localized (n=6)
Age, median (mean, range), y[a]	56 (61, 30-82)	55 (52, 28-82)	56 (56, 55-56)	7 (7, 7-7)	0.1 (0.1, 0.1-0.1)	59 (57, 39-78)	70 (70, 66-73)
Race[a]							
White	23 (51)	12 (27)	2 (4)	1 (2)	1 (2)	2 (4)	3 (7)
African American	0	0	0	0	0	0	0
Hispanic	0	0	0	0	0	1 (2)	0
Asian	0	0	0	0	0	0	0
Sex[a]							
F	14 (31)	5 (11)	1 (2)	1 (2)	1 (2)	0	3 (7)
M	9 (20)	7 (16)	1 (2)	0	0	3 (7)	0
Iris color[b]							
Blue	24 (44)	10 (19)	2 (4)	1 (2)	1 (2)	1 (2)	6 (11)
Green	2 (4)	1 (2)	0	0	0	0	0
Brown	2 (4)	0	0	0	0	2 (4)	0
Hazel	1 (2)	1 (2)	0	0	0	0	0
Visual acuity[b]							
20/20-20/40	25 (46)	12 (22)	2 (4)	1 (2)	0	3 (6)	6 (11)
20/50-20/100	4 (7)	0	0	0	0	0	0
20/200-HM	0	0	0	0	1 (2)	0	0
Intraocular pressure, median (mean, range), mm Hg[b]	24 (16, 10-37)	15 (15, 10-19)	18 (18, 12-23)	FTN[c]	FTN[d]	17 (17, 15-19)	15 (14, 12-17)

Abbreviations: FTN, finger tension normal; HM, hand motions.
[a] Forty-five patients.
[b] Fifty-four eyes.
[c] Child.
[d] Infant.

better visualized in a blue iris than a brown one. Fluorescein angiography clearly depicted the abnormal blood vessels with sparse vascularity in the adjacent iris and no leakage (Figure 1B and **Figure 2**B). A dilated, slightly tortuous episcleral blood vessel, termed *sentinel vessel*, was visible in the limbal sclera at the entry point of the hemangioma in the iris root in 22 of the 35 affected eyes.[13] All racemose hemangiomas remained stable on follow-up examinations. Only 1 racemose hemangioma produced hyphema. This occurred twice, spontaneously and transiently, leaving no complications in a 40-year-old healthy woman.

There were 3 patients (3 eyes) coded as iris cavernous hemangioma, 2 of which were solitary lesions localized to the iris in adults and 1 in a child who had additional brain, kidney, and skin hemangiomatosis.[19] Two of the cavernous hemangiomas produced multiple recurrent hyphemas. In each case, it appeared as a multilobular blue-red iris stromal mass (Figure 1E and F), with features identical to the well-known orbital cavernous hemangioma. One localized lesion was removed by iridectomy and confirmed histopathologically after more than 30 hyphemas during 18 years (Figure 1G and H).[17]

There was 1 patient coded as having an iris capillary hemangioma in our series. An infant girl was referred for a red iris lesion in her right eye that was first noted by her parents at age 4 weeks. Our examination results were entirely normal except for a 3-mm diameter red lesion inferiorly in the iris stroma of her right eye. There was no distinct periocular capillary hemangioma, but a few prominent dilated blood vessels were present on the right upper eyelid. The iris lesion was purely vascular and did not resemble a juvenile xanthogranuloma. The iris and skin lesions were subsequently documented by the referring pediatric ophthalmologist to have disappeared within 6 weeks. Although photographs were not possible in our case, iris capillary hemangioma is well documented in the literature (Figure 2A and B).

Three of our cases, all of which were in adults, were classified as iris varix, each of which was considered to be a thrombosed varix. The varix appeared in the iris stroma as a dark brown mass that resembled a blood clot and showed hypofluorescence on fluorescein angiography (Figure 2C and D). Two of the patients had transient hyphema preceding our examination, one of which achieved intraocular pressure of 52 mm Hg. In 1 case, varix removal by iridectomy confirmed the diagnosis (Figure 2E and F).[30]

The series included 3 patients with microhemangiomatosis. It was bilateral and characterized by multiple dilated vascular channels arranged circumferentially around the pupillary margin. The microhemangiomatoses were hyperfluorescent with angiography and showed leakage of fluorescein into the aqueous humor (Figure 2G and H).

COMMENT

Ferry's publication[1] in 1972 temporarily raised doubts as to the existence of iris hemangiomas. In fact, he titled

Table 5. Tumor Features in 54 Eyes of 45 Patients With Iris Hemangioma

Characteristic	No. (%) by Type of Hemangioma						
	Racemose		Cavernous		Capillary, Localized (n=1)	Varix (n=3)	Microhemangiomatosis, Localized (n=6)
	Simple (n=29)	Complex (n=12)	Solitary (n=2)	Systemic (n=1)			
Symptoms[a]							
Decreased vision	5 (9)	1 (2)	0	1 (2)	0	1 (2)	1 (2)
Flashes/floaters	0	1 (2)	0	0	0	0	0
Red eye	0	1 (2)	1 (2)	0	1 (2)	0	0
No symptom	24 (44)	9 (17)	1 (2)	0	0	2 (4)	5 (9)
Main iris quadrant[a]							
Superior	1 (2)	0	0	1 (2)	0	0	2 (4)
Inferior	9 (17)	3 (6)	1 (2)	0	1 (2)	0	2 (4)
Lateral	16 (30)	8 (15)	1 (2)	0	0	1 (2)	0
Medial	3 (6)	1 (2)	0	0	0	2 (4)	0
Diffuse	0	0	0	0	0	0	2 (4)
Blood vessels[a]							
Sclera feeder vessel	16 (30)	6 (11)	0	0	0	1 (2)	0
Iris feeder vessel	1 (2)	3 (6)	0	0	0	1 (2)	0
Tumor intrinsic vessels	29 (54)	10 (19)	1 (2)	1 (2)	1 (2)	2 (4)	6 (11)
Tumor size, median (mean, range), mm[a]							
Basal dimension largest	1.7 (1.7, 1-3)	3 (3, 2-4)	2.8 (2.8, 1.5-4)	1.5 (1.5, 1.5-1.5)	NA	3 (3.3, 2.8-4)	NA
Thickness	0.2 (0.2, 0.2-0.2)	1.8 (1.8, 1.5-2)	1.7 (1.7, 1.4-2)	1.5 (1.5, 1.5-1.5)	NA	2 (2.3, 2-3)	NA
Tumor size, median (mean, range), clock-hours involved[a]	2 (1.9, 1-6)	2 (3, 1-9)	1 (1, 1-1)	1 (1, 1-1)	1 (1, 1-1)	2 (1.7, 1-2)	3 (5, 1-12)
No. of episodes of hyphemas[a]							
1	0	0	0	0	0	2 (4)	1 (2)
2-5	1 (2)	0	1 (2)	1 (2)	0	0	1 (2)
6-10	0	0	0	0	0	0	0
>10	0	0	0	0	0	0	0
Associated eye findings[a]							
Glaucoma	0	1 (2)	2 (4)	0	0	1 (2)	0
Cataract	6 (11)	1 (2)	1 (2)	0	0	0	4 (7)
Corneal edema	0	0	0	0	0	0	0

Abbreviation: NA, not available.
[a] Fifty-four eyes.

his article "Hemangiomas of the Iris and Ciliary Body: Do They Exist? A Search for a Histologically Proved Case." In his meticulous histopathologic study of reported cases, he either disproved or rejected virtually every reported and unreported case on file at the Armed Forces Institute of Pathology.

The high rate of misdiagnosis in Ferry's series is understandable when one considers the histopathology of the most frequently misdiagnosed conditions in his review, namely iris melanoma in adults and iris juvenile xanthogranuloma in children. Iris melanoma often clinically and histopathologically contains large cavernous blood vessels that may be so prominent that the pathologist can easily overlook more subtle spindle melanoma cells and make the erroneous diagnosis of hemangioma. Similarly, juvenile xanthogranuloma is often highly vascular such that the pathologist may overlook the granulomatous inflammation and make the erroneous diagnosis of hemangioma.

It is clearly established that vascular tumors occur in other ocular structures, including the eyelid, conjunctiva, orbit, choroid, and retina, and hence similar lesions would be expected to occur in the iris. Judged from subsequent personal experience and scrutiny of the literature, it is now evident that vascular tumors of the iris also exist.

There are shortcomings in determining the types and frequency of the vascular tumors of the iris. First, reports in the literature of iris hemangiomas are often confusing and not well documented. Some authors have reported cases of iris "angiomas" or "hemangiomas" but have not specified the type of vascular tumor.[5,8,10-12] Second, most reported cases have not had histopathologic confirmation of the diagnosis. Third, even when histopathologic material is available, it may be extremely difficult in some cases to differentiate cavernous hemangioma from capillary hemangioma from varix and other lesions, because there may be histopathologic overlap of these lesions.[1,4-7] Fourth, some skeptics might challenge the diagnosis of hemangioma to define lesions like racemose hemangioma and microhemangiomatosis in which there is not a distinct mass. However, such lesions have traditionally been defined as hemangiomas in other parts of the eye and elsewhere in the body and it seems reasonable, for purposes of the communication, to do the same for iris vascular tumors. Hence, we will discuss the

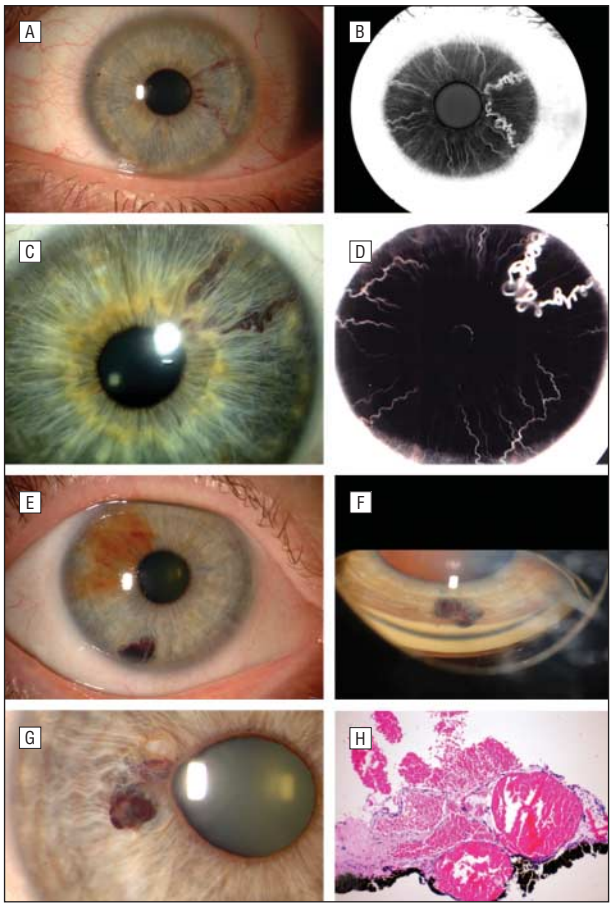

Figure 1. Clinical and angiographic features of vascular tumors of the iris. A, Clinical appearance of simple racemose hemangioma of iris. B, Fluorescein angiogram of lesion shown in A, better depicting the dilated, tortuous blood vessels. There is a sparsity of blood vessels in the area between the 2 components of the lesion. C, Clinical appearance of complex racemose hemangioma of iris. D, Fluorescein angiogram of lesion shown in C, depicting a more complex arrangement, tortuous blood vessels, forming a vascular mass. Again, there is sparsity of blood vessels in the area between the 2 components of the lesion. E, Cavernous hemangioma of the iris inferiorly in the right eye. The temporal iris nevus is coincidental and unrelated. The vascular lesion was relatively hypofluorescent throughout the angiogram, suggesting minimal blood flow through the tumor. F, Goniophotograph of lesion shown in E, showing the typical blood-filled cavernous vessels. G, Iris cavernous hemangioma adjacent to the pupil, causing slight peaking of the pupil. The lesion was removed by sector iridectomy. H, Photomicrograph of lesion shown in G. There are large blood-filled cavernous channels lined by endothelial cells.

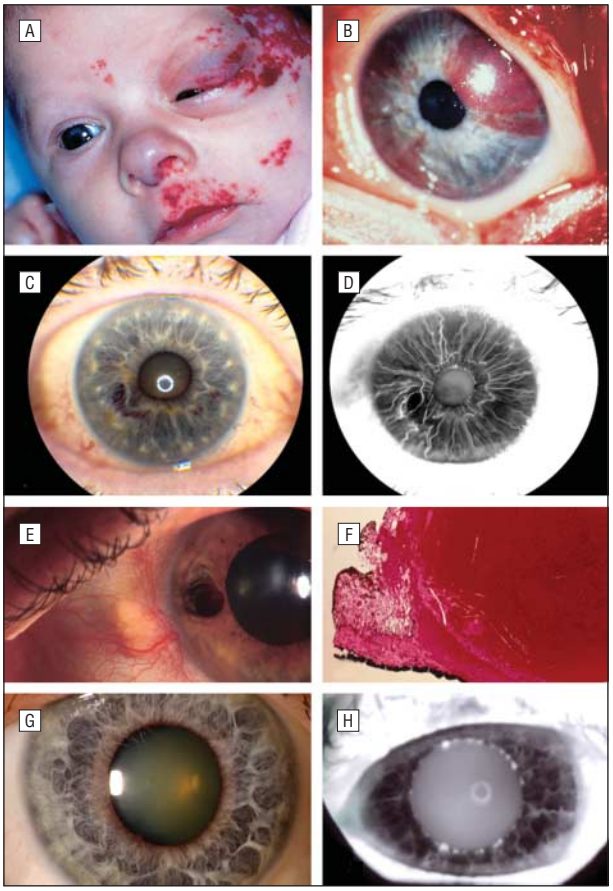

Figure 2. Further clinical and angiographic features of vascular tumors of the iris. A, Capillary hemangioma in an infant, showing involvement of the face and eyelid. Reprinted with permission from Slack Inc.[24] B, Anterior segment of child seen in A, showing capillary hemangioma involving superotemporal quadrant of the iris. The skin and iris lesion both subsequent spontaneous regression. Reprinted with permission from Slack Inc.[24] C, Iris varix located in iris stroma adjacent to the pupil. D, Fluorescein angiogram of lesion shown in C, revealing that the varix is hypofluorescent with slight leakage of dye from the adjacent iris vessels. E, Thrombosed iris varix. The brown homogeneous lesion temporal to the pupil is in the iris stroma, some overlying rarification of the more superficial stroma. The lesion was removed by sector iridectomy. F, Histopathology of lesion shown in E, demonstrating a large blood clot. It was lined by thin endothelial cells. G, Microhemangiomatosis. There are numerous small vascular tufts at the pupillary margin that are difficult to visualize and photograph. They are barely visible at 7- and 11-o'clock. H, Fluorescein angiogram of lesion shown in G, revealing numerous hyperfluorescent lesions at the pupillary border.

features of iris vascular lesion using our classification. A similar variation of this classification is used in a recent textbook[2] and a recent literature review.[3]

By far, the most common vascular lesion in our series was racemose hemangioma (arteriovenous communication). The clinical features of this iris lesion are described in the "Results" section and in the recent literature.[8-14] Until recently, there were only a few reported cases. In a report of 14 cases in 2006, the authors emphasized that iris racemose hemangioma may be more common than previously believed, but it may be subtle and difficult to recognize clinically, particularly in a brown iris.[13] Since that publication, we have seen more referrals for this lesion, perhaps owing to increased clinical awareness. Fluorescein angiography can dramatically depict the full extent of the lesion. It tends to occur in asymptomatic adults, some of whom have diabetes or hypertension, though no predisposing factors have been identified. None of the affected patients in our series have experienced hyphema or other complications. In 1 recorded case of bleeding from an angle vessel, the lesion did not appear to meet our anatomic criteria for a racemose hemangioma.[9] Iris racemose hemangioma has no recognized association with the retinal and brain racemose hemangioma seen with the Wyburn-Mason syndrome.[13] An interesting association with iris racemose hemangioma is a contiguous dilated episcleral blood vessel seen in 22 of the 41 eyes in our series. This raises concerns for an underlying ciliary body melanoma, which was not present in any of our cases.[13]

Cavernous hemangioma is rare, accounting for only 3 eyes in 3 patients in our series. A review of the literature reveals few well-documented cases.[15-19] Two of our 3 cases were localized to the iris and 1 was part of a sys-

temic cavernous hemangiomatosis. Concerning the localized type, it appears as a fairly well-circumscribed grapelike mass composed of large blue-red cavernous blood vessels that are remarkably similar grossly and histopathologically to the better known cavernous hemangioma of the orbit (Figure 1E and G). Our review of the case reported by Prost[15] suggests that the lesion may be microhemangiomatosis (discussed subsequently) and not a true cavernous hemangioma, based on clinical images and fluorescein angiograms. A case reported by Lam[16] may be a cavernous hemangioma, but the images also suggest a possible thrombosed varix. There may be overlap between cavernous hemangioma and varix, and it is possible that they represent parts of a spectrum, with varix being a thrombosed variant of cavernous hemangioma. Despite some doubt about its existence, there is a recently reported clinicopathologic correlation of a classic cavernous hemangioma of the iris in a man who had experienced more than 30 hyphemas over about 20 years. There is no doubt about the diagnosis in that case.[17] Our second patient had a localized cavernous hemangioma that we considered to be a classic case (Figure 1E), though histopathology is not available. The patient has been followed up and the lesion has remained stable for 4 years. These 2 cases are excellent examples of cavernous hemangiomas localized to the iris.

Importantly, there is another variant of iris cavernous hemangioma that is not confined only to the iris but is associated with multiple systemic cavernous hemangiomas.[18-21] There are several vascular lesions that have a systemic association and they appear to represent different syndromes. Larson and Oetting[18] described a 48-year-old woman with a cavernous hemangioma of the iris and multiple central nervous system hemangiomas. One of our cases was a child who developed hydrocephalus at age 3 months and was found to have hemangiomas of the skin, brain, and kidney, requiring multiple treatments. We saw the child at age 7 years, and 1 year later the iris hemangioma had spontaneously deflated, leaving a fibrous scar.[19] Iris cavernous hemangiomas can also be a component of diffuse congenital hemangiomatosis, which is often fatal in infants.[20,21] An iris cavernous hemangioma has been reported following evisceration for a perforated globe in a patient with leprosy, but the diagnosis is questionable and the association may be coincidental.[22]

True capillary hemangioma of the iris is extremely rare. There was 1 case in our series that possibly represented a capillary hemangioma based on its clinical appearance and a very subtle ipsilateral eyelid vascular lesion. It was not possible to obtain adequate photographs of the iris lesion in that infant. Both lesions showed spontaneous regression. Iris capillary hemangioma has been seen mainly in association with periocular cutaneous capillary hemangioma of infancy. A well-documented case was reported by Ruttum and associates.[24] Other possible cases are less well documented.[23,25] The classic case appears as a periocular cutaneous hemangioma with an ipsilateral iris component (Figure 2A and B). Like the cutaneous counterpart, the iris component shows similar spontaneous regression.[24] A case reported by Cashell[23] as an angioma of the iris in a 2-year-old child was likely a capillary hemangioma based on the published histopathology. Based on our review, we believe that the case reported by Woo and associates[25] as a cavernous hemangioma in an infant was possibly a capillary hemangioma.

There were 3 cases of iris varix in our series (Table 1). These are discussed briefly in the "Results" section. As with other iris vascular lesions, their diagnosis is often difficult to confirm clinically and histopathologically.[30-34] Two recently reported cases, however, had good histopathologic confirmation of the diagnosis.[3,30] Unlike some iris vascular lesions, iris varix apparently has no known systemic associations.[3]

There were 3 patients with iris microhemangiomatosis, also called iris vascular tufts, in our series. A literature review suggests that it is by far the most common iris vascular tumor, with hundreds of cases having been reported.[35-50] It is most often detected by ophthalmologists following a spontaneous hyphema sometimes with secondary elevated intraocular pressures. Hence, it is important to differentiate microhemangiomatosis from iris neovascularization due to diabetic retinopathy or retinal vein obstruction. The pattern of iris neovascularization is different in that the distribution of the blood vessels in iris neovascularization is more widespread on the iris and in the anterior chamber angle. The hyphema often occurs after cataract surgery and does not create enough suspicion for a true neoplasm to warrant referral to an oncology center, explaining the low number of cases in our series. However, the hyphema and vascular lesion sometimes raise suspicion for an iris melanoma.

Microhemangiomatosis is occasionally asymptomatic and is found on routine slitlamp biomicroscopy. Cobb[49] looked carefully at the pupillary margin of a series of patients and identified 44 patients, none of whom had any symptoms related to the iris lesions. It generally appears as multiple very subtle dilated vascular channels that arise from the pupillary vascular arcade. Fluorescein angiography reveals that they are more numerous than suspected clinically, become rapidly fluorescent, and often leak dye into the aqueous.[35] Microhemangiomatosis that causes recurrent hyphema has been treated successfully with laser photocoagulation.[39,43,44] There is little histopathology available for microhemangiomatosis, but a report of 1 lesion removed at cataract surgery described it as a hamartoma of capillary hemangioma type with features specific for iris blood vessels.[42]

Microhemangiomatosis is 1 of the iris vascular lesions that has been found to have intriguing systemic associations, mainly with myotonic dystrophy and possibly with diabetes mellitus.[3,49,50] Cobb and associates[50] found iris microhemangiomatosis in 5 of 8 men in a series of 10 patients with myotonic dystrophy. The reason for this odd association is not known.

In summary, although the existence of iris vascular tumors was once challenged, there are examples of racemose hemangioma, cavernous hemangioma, capillary hemangioma, varix, and microhemangiomatosis in our series and in the literature. Each type has characteristic clinical features and different ocular complications, mainly spontaneous hyphema. This study has reviewed a series of 45 patients with iris vascular tumor and related conditions seen on an ocular oncology service. A simple classification is proposed and the demo-

graphics, clinical features, complications, management, and histopathology are discussed. Although most are solitary lesions, some have serious systemic implications.

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