Finally, patients with chronic unilateral conjunctivitis refractory to routine therapy, a diagnosis of neoplasia and floppy eyelid syndrome needs to be considered. Thorough clinical evaluation and histologic features of the excised tissue (granulomatous lesion, presence or absence of necrosis, associated inflammatory cell response) and identification of the possible infectious agents by special stains and molecular methods help differentiate these lesions.

Conjunctival scrapings and biopsy specimens may have a low sensitivity for the detection of acid-fast organisms, requiring approximately 1000 to 10000 organisms per milliliter of sample for morphologic detection and 100 to 1000 viable organisms per milliliter for a positive culture. This could explain the absence of organisms in the initial conjunctival and corneal scrapings. Polymerase chain reaction, based on the principle of DNA amplification, is highly sensitive and can detect as few as 10 microorganisms. This is a useful modality when special stains do not reveal organisms in the presence of caseating granulomas.

In summary, patients with chronic unilateral conjunctivitis need a detailed evaluation, including a conjunctival biopsy, and if possible, molecular diagnostic techniques for rapid diagnosis and institution of appropriate treatment. A high index of suspicion for tuberculosis is warranted before administering intraslesional depot corticosteroid injections, as this may result in a flare-up of the underlying infection and lead to tarsal necrosis.

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Reports from 1964 and 1972 described smooth muscle tumors in the iris. Foss et al., using immunohistochemical analysis, have reclassified previous reported cases as melanocytic lesions. As a result, stricter pathologic criteria for diagnosing intraocular neoplasms of smooth muscle were established in 1994. Since then, there has been only one well-documented example of iris leiomyoma reported. We report an HIV-infected girl with an iris neoplasm, histologically and immunohistochemically proven to be leiomyosarcoma.

Report of a Case. A 4-year-old Thai girl with vertically acquired HIV infection was born at our hospital in 1996. She was asymptomatic until age 2 years, when she developed dermatis, dilated cardiomyopathy, hepatosplenomegaly, lymphadenopathy, and parotitis. The CD4 count was 1%. At 3 years of age, treatment with zidovudine and didcoxycyctidine was started, but the adherence to antiretroviral agents was poor. Her subsequent course was complicated by pneumonia, leading to several hospital admissions.

At the age of 4 years 10 months, she experienced pain in her right eye. Ophthalmologic examination revealed a whitish mass located on the lower half of the iris, 4 mm in diameter. There was vascular dilatation on the anterior stromal surface. The lower part of the anterior chamber was shallow, with peripheral anterior synechiae. There were no signs of inflammation seen in the anterior chamber. The lens was normal. Computed tomography (CT) did not demonstrate any other intraocular lesions, but CT of the brain showed a 3-cm cystic mass in the parenchyma of the left frontal lobe. Aspiration of the cyst revealed numerous red cells, some mononuclear cells, and no malignant cells.

Inferior sector iridectomy was carried out, and the iris mass was surgically removed by an inferior limbal incision. The entire tissue was fixed in 10% formalin, routinely processed, and embedded in paraffin wax. In addition to routine staining, immunohistochemical analysis was performed with antibodies against the following proteins: S100, HMB45, muscle-specific actin, smooth-muscle actin, desmin, and EBV latent membrane protein. In situ hybridization for EBV was also performed, using EBV-encoded RNA oligonucleotide probe. Both the immunohistochemical analysis and in situ hybridization were done in the presence of adequate controls.

Microscopically (Figures 1 and 2), the lesion consisted of plump spindle-shaped tumor cells forming interlacing fascicles. The neoplastic nuclei were hyperchromatic, and some of them possessed distinct small nucleoli. There are scattered mitotic figures, approximately 5/10 per high-power field. Tumor cells strongly expressed muscle-specific actin and smooth-muscle actin, supporting the smooth muscle origin of the lesion. Desmin, S100 protein, and HMB45 were nonreactive. The above findings are characteristics of leiomyosarcoma. Evidence of EBV infection was revealed by in situ hybridization but not by immunohistochemical analysis.

At the age of 5 years 3 months, she was admitted to the hospital with persistent and severe abdominal pain. Duodenal tissue biopsy showed acid fast bacilli in the mucosa, and Mycobacterium gordonae were later isolated. She died at the age of 5 years 6 months.

Comment. Leiomyosarcoma has been well-documented to be associated with HIV infection, and usually occurs in the visceral organs. The tumor rarely arises in the uveal tract. A literature search, using the keywords “iris,” “uvea,” “uveal tract,” and “leiomyosarcoma” from major electronic databases (MEDLINE, EMBASE, BIOSIS, and CINAHL), did not reveal any previous cases of histologically and immunohistochemically proven iris leiomyosarcoma.

Before the advent of immunohistochemical analysis, intraocular leiomyomas and their malignant counterparts, leiomyosarcomas, have been a frequent subject of debate. Spindle cell melanoma is the main differential diagnosis. Immunohistochemical study is therefore particularly important. Foss et al. proposed that immunohistochemical and/or electron microscopic investigation are crucial in establishing the correct diagnosis of ocular smooth muscle tumors. In our case, the positive staining for smooth muscle markers (smooth-muscle actin and muscle-specific actin) and the absence of immunolabeling for melanoma markers (S100 protein and HMB45) strongly support the diagnosis of smooth muscle tumor and rule out the possibility of melanocytic lesion. Although histologic criteria for the distinction of benign and malignant smooth muscle neoplasms outside the uterus are not well established, the high mitotic in-

Figure 1. Hematoxylin–eosin-stained section demonstrates interlacing fascicles of plump spindle cells (A). Mitotic figures are frequently observed (arrows) (B).
dex favors the diagnosis of leiomyosarcoma.

The presence of the cystic lesion in the left frontal lobe raises questions concerning the possible relationship between the cyst and the iris tumor and whether both represent leiomyosarcoma. If this was the case, it would be difficult to determine the primary site of the tumor. However, most reported HIV-associated intracranial leiomyosarcomas are dura-based, solid (nonglystic) masses.10-12 We consider both of these lesions to be unrelated. Unfortunately, no tissue was obtained from the cyst for definite histologic evaluation.

Regarding the histogenesis, smooth muscle tumors of the iris are thought to originate from its sphincter or dilator muscle, but the concept that neoplasms can affect the iris is dubious.5-11 Our case has confirmed that a cellular progenitor of smooth muscle tumors does exist in this location. The association between EBV and smooth muscle tumors in HIV-infected subjects has been well documented,3 as evident in our patient. In addition to the reduction in tumor surveillance cells and the inability to remove EBV in immunocompromised subjects, it has been shown that EBV receptors are unregulated and thus may enhance viral entrance into and transformation of the human cells.5-12 The discrepancy between EBV-encoded RNA in situ hybridization and immunohistochemical characteristics of EBV latent membrane protein is the same as for previously reported EBV-associated smooth muscle tumors.10,13 Failure to detect EBV latent membrane protein may be due to the absence or subthreshold level of the antigen in this tumor cell type.13

This case is the first immunohistochemically verified leiomyosarcoma of the iris that has occurred in an HIV-infected child, to our knowledge. Although the association between the tumor and EBV in immunosuppressive patients has been previously determined, this report reiterates that HIV infection facilitates the development of this tumor, even in an unusual location such as the iris.

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hemorrhage and chorioretinal atrophic lesions nasal to the optic disc.

Figure 1. Only a few well-documented cases of malignant neoplasms of the retinal pigment epithelium (RPE) are rare. Some of these tumors developed in association with choroidal neovascularization, 

Adenocarcinoma of the Retinal Pigment Epithelium

Malignant neoplasms of the retinal pigment epithelium (RPE) are rare. Only a few well-documented cases have been described in the last decades. Some of these tumors developed in association with choroidal neovascularization, 

Report of a Case. A 37-year-old man was referred to our clinic in January 1980 for a progressive loss of vision in his right eye during the preceding months. His visual acuity was counting fingers OD and 20/20 OS. Funduscopy showed a subfoveal hemorrhagic lesion in the right eye, several atrophic chorioretinal scars nasal to the optic disc, and peripapillary atrophy (Figure 1A). Fluorescein angiography revealed a subfoveal neovascular membrane (Figure 1B).

Figure 1. Color fundus photography (A) and fluorescein angiography (B) of the right eye in 1980, showing a subfoveal neovascular membrane with subretinal hemorrhage and chorioretinal atrophic lesions nasal to the optic disc.

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