Amyloid Mass of the Ciliary Body

Making the correct diagnosis of an iris–ciliary body tumor can be challenging. A 35-year-old woman sought care because of a growing amelanotic lesion that destroyed the iris root and produced a large ciliary body mass. She had a history of systemic lupus erythematosus. The tumor was completely removed with an iridocyclectomy, which revealed that the plasma cells had been producing a large amount of amyloid. To our knowledge, this is the first description of an amyloid lesion simulating a solid intraocular tumor.

Amyloid involvement of ophthalmic structures is uncommon and rarely simulates a neoplasm. We and others have reported involvement of the vitreous in familial and nonfamilial primary amyloidosis; this process could be confused with an intraocular lymphoma. Similarly, there are several reported cases of conjunctival, eyelid, or orbital amyloid tumors. Rarely, amyloid can also be found infiltrating the anterior chamber angle or sclera, but no amyloid lesions simulating a solid intraocular tumor have been reported.

The differential diagnosis of focal iris–ciliary body tumors is relatively straightforward. The majority of cases are uveal melanomas; less commonly, benign cystic lesions, melanocytomas, smooth muscle tumors, Fuchs adenoma, primary carcinomas, medulloepitheliomas, lymphoid masses, neurofibromas, and metastases have been reported. We describe a unique case of a growing iris–ciliary body tumor that was shown, after excision with an iridocyclectomy, to be a plasma cell proliferation with a secondary amyloid deposit.

Report of a Case. This 35-year-old woman had a 2-year history of a right iris–ciliary body growth and a 2-month history of decreased right eye vision. Her medical history was significant for systemic lupus erythematosus vasculitis, muscle pain, and both joint and facial swelling. She managed her symptoms with holistic medicines and was in remission with a negative anti-nuclear antibody. Aside from easy bruisability, she stated that she was currently in good health. None of the holistic medicines, based on a computer search, were associated with amyloid deposition.

On examination, the visual acuity was 20/50 OD with current correction and 20/30 OS. The left anterior segment was unremarkable and the intraocular pressures were 18 OU. From the 3- to 5-o’clock position in the right eye, there was an amelanotic iris–ciliary body mass with minimal posterior pigment epithelial proliferation at the edges (Figure 1). The bilobed amelanotic mass predominantly involved the ciliary body with erosion through the iris root. The pupillary margin was not distorted. There was no altered iris vasculature or lens opacity. The angle had no neovascularization. The fundi were unremarkable.

An iris angiogram did not show a marked leakage or any intrinsic tumor vasculature. A large iris–ciliary body mass (Figure 2) was visible on high-frequency ultrasound. An iridocyclectomy was performed with complete removal of this mass.

On gross examination, the mass appeared to be a ciliary body tumor with iris involvement. It was 6 × 5 × 3 mm. There were no ciliary process cysts. Beneath the ciliary epithelium, a tumor was composed of atypical plasma cells (Figure 3A), scattered lymphocytes, and a large amount of homogeneous eosinophilic material that was stained with crystal violet and Congo red (Figure 3B) and showed slight apple-green birefringence. It was strongly positive for κ light chains (Figure 4A) and negative for...
λ chains (Figure 4B). The pleomorphic plasma cells were stained with CD 138 (Figure 4C) and CD 79a (Figure 4D). The results of an amyloid A stain and CD20 stain were negative.

These histologic features were consistent with a diagnosis of plasmacytoma of the ciliary body with early amyloid deposition and κ light chain deposition. The pleomorphic plasma cells were negative for a general B-cell antigen, which is usually absent on plasma cells, but were positive for a more differentiated B-cell antigen (CD 79a) and a plasma cell antigen (CD138). The plasma cells and the homogeneous protein deposit expressed a monoclonal κ light chain antigen.

Test results for a complete blood cell count, antinuclear antibody, immunoelectrophoresis, protein electrophoresis, and 24-hour urine collection for Bence-Jones proteins were all negative. Based on these negative results and the advice of 2 medical oncologists, no bone imaging was performed.

Comment. The differential diagnosis of iris–ciliary body tumors is usually straightforward but has a higher diagnostic error rate than posterior uveal masses. In our case, the history, physical findings, and ancillary testing suggested that more common entities such as melanomas, melanocytomas, metastases, and cystic lesions were not the cause of this lesion. The 2-year history of growth and slow visual loss made a metastatic lesion unlikely. The lesion was completely amelanotic, unlike most melanomas or melanocytomas; fluorescein angiography revealed no intrinsic tumor circulation, which is often noted with melanomas of this size. There was no cystic component to the lesion visible.
on high-frequency ultrasound. Preoperatively, we suspected that this most likely represented a benign smooth muscle tumor that had eroded through the iris root. Amyloid can occur as a localized ocular or widespread systemic process. Approximately 4% of amyloid deposits in the head and neck region involve the orbit. Amyloid deposits in ophthalmic structures can occur as a primary or secondary process. Primary deposits (which can be familial or sporadic) occur in the absence of an associated disease. Secondary deposits have been noted after a myriad of processes, including trauma, infection, myeloproliferative disorders, and immune-mediated diseases. Most of the reported ophthalmic cases have been in association with familial amyloidosis with systemic involvement. Some cases have been noted to have only ophthalmic deposition of amyloid without evidence of systemic disease. Amyloid deposits in association with myeloproliferative entities such as lymphomas or plasma cell proliferations can produce paraproteinemia and involve the eye. In our case, the negative study results make this entity unlikely at present. In patients with extrasosseous plasmacytoma, a myeloma develops within 10 years in 10% to 30% compared with 55% in patients with osseous plasmacytomas. Amyloid deposits can also occur in association with rheumatologic diseases, although involvement with systemic lupus erythematosus is distinctly uncommon. Our patient has either a primary amyloid deposit from an extrasosseous plasmacytoma or a focal iris–ciliary amyloid deposit in association with systemic lupus erythematosus. It is conceivable that benign but aberrantly localized plasma cells are part of the systemic lupus erythematosus process in this patient. Alternatively, there is an increase in lymphomas in patients with systemic lupus erythematosus, even when they are not treated with immunosuppression. Amyloid in association with systemic lupus erythematosus is quite rare and usually manifests as renal involvement. Fewer than 20 cases have been reported. This case illustrates the problem, despite newer diagnostic techniques, of potential diagnostic errors in anterior uveal tumor diagnosis. Our patient is probably at risk for local and systemic recurrence and is being closely observed.

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Acute Posterior Multifocal Placoid Pigment Epitheliopathy With Cerebral Vasculitis: A Multisystem Granulomatous Disease

Typically, acute posterior multifocal placoid pigment epitheliopathy (APMPPE) is seen along with acute binocular visual disturbance (ie, visual blurring, metamorphopsia, or scotomas) in young adults of whom approximately one third experience a flulike illness at onset. The clinical course is usually self-limited, with remarkable visual recovery. A few cases with cerebral vasculitis or meningoencephalitis have been reported. Although circumstantial evidence for a choroidal vasculitis is gathering, the exact pathological origin of APMPPE remains unknown. Herein we report a case of APMPP-associated cerebral vasculitis with angiographic, radiologic, and, to our knowledge, for the first time in the literature autopsy findings that include both ocular and cerebral histopathological descriptions.

Report of a Case. Clinical History. A 23-year-old white man developed acute loss of vision in the right eye. There was no history of a flulike illness and his medical history only denoted resection of nasal polyps. On examination visual acuity was confined to perception of a waving arm at a distance of 1 m OD and 20/40 OS. Fundoscopic examination showed multiple creamy white lesions just below the retinal pigment epithelium (RPE) in both eyes. Besides a slightly raised C-reactive protein level, findings from other routine blood tests (including angiotensin-converting enzyme, anti-nuclear antibody, and antineutrophil cytoplasmic antibody screening) were normal. A fluorescein angiogram (Figure 1) showed early-stage hypofluorescence and late-stage hyperfluorescence consistent with APMPPE in both eyes. Next he developed bilateral anterior uveitis, and 3 days later, he had acute pain behind the right eye and severe headache. Results of neurological examination showed a left-sided hemiparesis and hypesthesia. He developed a tonic-clonic status epilepticus that did not respond to intravenous treatment with clonazepam and phenytoin sodium. A magnetic resonance imaging study of the brain and magnetic resonance angiography (Figure 2) revealed occlusion of the left medial cerebral artery and narrowing of the right posterior cerebral artery with infarctions in these vascular territories. Massive swelling of the left hemisphere with midline shift and temporal herniation resulted in death.