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.8 Amyloid deposits in ophthalmic structures can occur as a primary or secondary process.1,4 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.8

Amyloid deposits can also occur in association with rheumatologic diseases, although involvement with systemic lupus erythematosus is distinctly uncommon.9 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.10 Amyloid in association with systemic lupus erythematosus is quite rare and usually manifests as renal involvement. Fewer than 20 cases have been reported.9

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.

Devon H. Char, MD
J. Brooks Crawford, MD
Ed Howes, MD
James A. Carolan, MD

Correspondence: Dr Char, 45 Castro St, Suite 309, San Francisco, CA 94114 (devron@tumori.org).

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Pathological Findings. In the lungs and enlarged paratracheal hilar lymph nodes, noncaseating granulomas were observed that contained a large number of Langhans or foreign-body–type multinucleated giant cells without asteroid or Schaumann bodies. Ziehl-Nielsen stain and a combination of rhodamine and auramine stain for mycobacteria were negative. Using polarized light no foreign-body material was found. Small noncaseating granulomas were also found in the heart, liver, and spleen. In the choroid there was a granulomatous inflammation with Langhans multinucleated giant cells and focal disruption of the RPE (Figure 3 A and B). In these specimens no signs of vasculitis were observed.

Both hemispheres showed diffuse swelling and infarcts, which were more pronounced on the left side. The sections through the origin of the left medial cerebral artery showed occlusion by a blood clot, eccentric hyperplasia of the intima, as well as a granulomatous inflammation of the vessel wall with degeneration (Figure 3 C and D), and the presence of Langhans-type multinucleated giant cells at the level of the lamina elastica interna, which also showed degeneration (Figure 3 E). Bi-refringent material inside the giant cells was not found. Parenchymatous granuloma was not found in the central nervous system.
Comment. There have been a number of reports of aseptic meningitis and cerebrovascular accidents in association with APMPPE.2-10 Cerebral angiographic findings in some of these cases were suggestive of vasculitic changes. In our case a focal granulomatous vasculitis affecting large cerebral arteries was demonstrated. The inflammation contained Langhans-type multinucleated giant cells at the level of the lamina elastica interna, which also showed fragmentation. This latter feature is frequently seen in giant cell (temporal) arteritis, and it is hypothesized to be the primary site to which the immunologic response is focused.11 These findings are in agreement with the only other documented histopathological study of cerebral vasculitis in association with APMPPE. Those authors reported a multifocal, granulomatous arteritis of medium arteries with fibrinoid necrosis, not dissimilar to the microscopic findings in temporal arteritis.7 As temporal arteritis is one of the large-vessel vasculitides that does not affect intradural vessels, and most of the patients are older than 50 years of age, it is unlikely that the findings in our and the mentioned case are due to “classic” temporal arteritis.

In his initial description, Gass12 proposed APMPPE to be primarily a disease of the RPE. Buskirk et al13 proposed a focal choroidal vasculopathy to explain the slow, irregular filling of the early hypofluorescent areas on the fluorescein angiogram. Various fluorescein angiographic studies confirmed malperfusion of the lamina choriocapillaris.4,8,14,15 However, ocular pathological characteristics of APMPPE have not been reported. In our case we found granu-
fluorescein angiogram shows a diphtheritic granulomatous vasculitis of large and medium vessels. The granulomas were situated near arterioles, capillaries, or venules. The chorio-capillaris itself did not show any sign of acute or chronic vasculitis. Furthermore, generalized granulomas were found in lung parenchyma, lymph nodes, heart, liver, and spleen. One could argue that the generalized granulomas with multinucleated giant cells are in line with advanced sarcoidosis. In favor of this idea are 2 case reports of APMPPE with probable sarcoidosis and posterior choroiditis. The granulomas in our patient contained no characteristic asteroid bodies or Schaumann bodies, which can be seen in sarcoidosis. Furthermore, the occurrence of stroke in sarcoidosis is extremely rare and the fluorescence angiogram shows a different pattern.

The absence of any signs of previous or present vasculitis in the chorio-capillaris does not support the hypothesis that APMPPE is caused by a choroidal vasculitis of the lamina chorio-capillaris. Instead, our findings indicate that APMPPE is caused by choroidal granulomas and can be part of a generalized granulomatous disease. The granulomas resemble those seen in sarcoidosis. However, its clinical presentation and the occurrence of a cerebral granulomatous vasculitis of large and medium arteries instead suggests that it may be a distinct multisystem granulomatous disease.

Recognition of this syndrome is important and our case illustrates that it can be rapidly fatal. Because cerebral vasculitis associated with APMPPE usually responds well to corticosteroid therapy, we propose that patients with APMPPE complicated by central nervous system manifestations should be treated immediately with intravenous corticosteroids.

Macular Hole in the Shaken Baby Syndrome

Retinal hemorrhages have been reported in 50% to 100% of infants diagnosed as having shaken baby syndrome (SBS). 1 Retinoschisis and circular perimacular retinal folds are associated with poor prognosis in SBS. 2 Although these ophthalmologic findings have been well documented in the literature, macular holes have not been described. We present 5 cases of children who developed macular holes as a sequela to SBS.

Five patients were diagnosed as having SBS based on systemic, intracranial, and ophthalmologic findings. The median age of trauma was 9 months (range, 6-10 months), and the median age of macular hole diagnosis was 10 months (range, 8-12 months). All macular holes were unilateral, despite severe bilateral retinal disease. Four patients had severe vitreous hemorrhage and intraretinal hemorrhage, and 1 patient had diffuse retinal hemorrhage affecting all retinal layers. The median size of the macular hole was 700 μm (range, 500-1500 μm). Three macular holes were centrally located; 2 macular holes were ectopically located (juxtapfoveal). The diagnosis of macular hole was made during initial funduscopic examination in 2 patients, during vitrectomy in 2 patients, and after clearing of the vitreous hemorrhage, initially obscuring the visual axis, in the final patient.

Surgical intervention was performed in 4 cases to clear the visual axis of vitreous and subhyaloidal hemorrhage. Surgery included vitrectomy, internal limiting membrane peel, and tamponade (no tamponade in patient 1, silicone oil in patient 2, perfluoropropane 12% tamponade in patient 3, and air in patient 4). The median age at the time of vitrectomy was 11.5 months. Three out of 4 eyes had successful hole closure following surgery (that in patient 1 remained open). The median follow-up period was 12 months (mean, 12.4 months; range, 6-24 months). In all 5 cases, blood was seen at the base of the hole, occasionally pluming outward.