Ciliary-Choroidal Mass in a 52 yo Female:

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Case presentation:

HPI: A 52 year old woman presented to her local ophthalmologist 6 mo prior to presentation at KEC with initial symptoms of blurred VA and partial scotoma OD. Patient denied pain or redness. Her outside ophthalmologist diagnosed iridocyclitis OD and initially treated with topical steroids without improvement in symptoms. Patient was subsequently treated with intravitreal triamcinolone OD which provided temporary improvement in VA, but VA gradually decreased again over last 4 months. Patient notes outside ophthalmologist performed "labs and chest x-ray" all of which were reported as normal. Patient referred to KEC for further evaluation and management.

Past medical history: Psoriasis – affecting ear canals **Review of Systems**: Negative

Family history - Uterine and breast cancer (paternal grandmother), lung cancer (paternal grandfather), GI cancer (maternal grandfather)

Past ocular history: Cataracts OU, Radial keratotomy OU, PVD OD, ERM OD **Ocular medications:** None

Ocular Exam:

VA OD: 20/40 OS:20/25 **IOP**: OD: 17 OS:14

Slit lamp: Conjunctiva: w/o injection OU, Cornea: RK scars, no KP, clear OU, Anterior chamber: Deep, very rare cell OD, deep/quiet OS, Iris: nl OU, Lens: faint NS OU, Anterior vitreous: rare cell OD, clear OS.

Fundus exam: Vitreous: Central posterior vitreous cells/haze OD, clear OS. Disc: C:D 0.25 OU, without edema or pallor. Macula: OD hazy view, mild ERM, possible submacular fluid, OS normal, Vessels: normal OU, Periphery: OD Lattice degeneration superiorly, inferior chorioretinal scarring, OS normal.

OCT: OD: CMT 698, ERM with surface wrinkling of macula, Submacular fluid present extending both inferiorly and temporary. Nodularity of choroid with replacement of normal choroidal vasculature, OS: CMT 323, Normal study.

FA: OD: Normal transit phase, with normal posterior retinal and choroidal vasculature early, with late fluorescein leakage noted in inferior periphery. OS: Normal study

Ultrasonography: OD: Moderately dense vitreous opacities, total vitreous detachment with moderately dense subhyaloid opacities, Low reflective choroidal thickening extending from 6 to 11:00, from posterior pole into mid-periphery, no Tenon's infiltration or orbital lesion detected. OS: Mild vitreous opacities, posterior vitreous detachment.

Laboratory studies obtained: Quantiferon TB Gold negative, FTA-Abs negative, ACE normal, serum lysozyme normal, cANCA negative, CBC normal

The ocular oncology service was consulted and had concerns of possible lymphoproliferative disorder. FNAB of the choroidal mass was recommended and the patient agreed to schedule. In the interim, CT of chest, abdomen and pelvis were ordered and all returned normal. Laboratories ordered included with

the following results: Quantiferon TB Gold negative, FTA-Abs negative, ACE normal, serum lysozyme normal, cANCA negative, CBC normal.

Patient returned unexpectedly in 3 weeks with complaints of worsening VA OD. Exam was unchanged OS but VA OD had declined to CF at 6 in, with KP on lower 30% of cornea, increased AC cell, and markedly increased vitreous cell, as well as a bullous exudate RD present. Ultrasonography OD demonstrated increased vitreous opacities, new ciliary body thickening in all quadrants, retinal detachment from 2-8:00 extending from the mid-periphery to periphery. OS remained normal. Recommended that patient have choroidal biopsy rather than FNAB.

A ciliary-choroidal biopsy was performed without complication. Partial thickness scleral flap created that extended 5 mm circumferentially from the limbus from 7-9:00. Vannas scissors were used to incise scleral bed to expose ciliary body, and choroid. A 0.6 x 0.1 x 0.1 ciliary-choroidal specimen was excised and sent to pathology. The scleral flap was closed with 8-0 Vicryl.

The ciliary-choroidal specimen contained extensive infiltration of chronic inflammatory cells visible at low power on the H&E slides. Localized areas had cells with abundant pink staining cytoplasm, possibly histiocytes. At higher power clusters of pink histiocytes could be seen within discrete granulomas and in confluent sheets. Immunohistochemical staining for CD68 and CD163 confirmed the identity of the histiocytic nature of the pink cells within these areas. CD3 and CD20 immunohistochemical staining also verified the presence of T-lymphocytes and B-lymphocytes, with T-lymphocytes predominating. The presence of non-caseating granulomas was consistent with a diagnosis of sarcoidosis.

Patient sent for rheumatology evaluation for extraocular sarcoidosis, which was negative. Prednisone 60 mg daily started and tapered off after Cellcept 1000mg po BID established. Subretinal fluid, ciliary/choroidal thickening, and vitreous debris rapidly improved in response to oral steroid therapy. Four months post-biopsy the VA improved to 20/30 OD.

Discussion:

Sarcoidosis is a chronic, granulomatous, autoimmune disease that may involve multiple organ systems and involves the eye in 25-60% of cases of known systemic sarcoidosis, most commonly presenting as uveitis or a conjunctival nodule.[1] The chest radiograph or chest CT are the best screening tests for sarcoid since 90% of sarcoid patients have pulmonary involvement (hilar lymphadenopathy or infiltrates). [2] Elevated serum levels of ACE and/or serum lysozyme may be supportive of a diagnosis of sarcoidosis, although these tests are only 73% and 60% sensitive, respectively. [3] However, a definitive diagnosis of sarcoidosis requires a tissue biopsy. The typical histopathologic findings in sarcoid are non-caseating granulomas in the involved organs. [4] The granulomas consist of epithelioid and multinucleated giant cells, with CD4 positive T cells, as well as CD8 positive T cells and B cells typically located in the periphery of the granuloma.[5]

Ocular involvement is recognized as an initial manifestation of sarcoidosis in up to 21% of patients, with the presenting ocular manifestations most often anterior, intermediate or posterior uveitis.[6,7] Our patient did have evidence of anterior and intermediate uveitis present, however, the presence of choroidal and ciliary body mass, is an atypical early presentation of sarcoid. A prior report of choroidal sarcoid nodule as an early presenting symptom, made the diagnosis of sarcoidosis, in the face of negative serology, via biopsy of hilar lymph nodes, which were evident on chest CT but not chest x-ray.[8]

In our patient, there was no evidence of extraocular sarcoid based on extensive radiographic screening, clinical examination, and serologic testing. Our case reaffirms that ocular sarcoid may be present in the absence of any evident systemic sarcoid. If significant ciliary or choroidal thickening is present, it may be possible to safely biopsy these sites, to not only rule out lymphoproliferative or infectious disease, but that these sites may be the only option to obtain pathologic confirmation of sarcoidosis.

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