SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES 2023; 40 (4): e2023039 DOI: 10.36141/svdld.v40i4.15122

Choroidal nodules in ocular sarcoidosis

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Abstract. *Background and aim:* Ocular sarcoidosis is present in 30-60% of all sarcoidosis patients. Our purpose is to increase awareness of the various presentations of ocular sarcoidosis. *Methods:* Short image-based clinical case report. *Results:* We report on a case of ocular sarcoidosis presenting with unilateral choroidal nodules in a middle-aged man. Sarcoid uveitis is generally bilateral and rather symmetrical. However, choroidal nodules are an exception to this rule, as they generally arise unilaterally. Choroidal nodules are highly responsive to oral corticosteroids. When left untreated, they may evolve to chorioretinal atrophy and secondary choroidal neovascularization. *Conclusions:* Knowledge of this presentation of ocular sarcoidosis can help clinicians optimize treatment outcomes for patients.

Key words: sarcoidosis, choroid, uveitis

We herein report the case of a 51-year-old man who presented to the ophthalmology clinic for routine follow-up. Five years prior, he had received a diagnosis of parotid gland sarcoidosis following recurrent bouts of bilateral parotid gland enlargement. Histopathological examination of a minor salivary gland biopsy had shown non-caseating granuloma. He had not received any treatment initially as the attacks were self-resolving and only hindered the patient very little.

The patient had no systemic or ocular complaints on questioning. On ophthalmological examination, best-corrected visual acuity was 20/20 in both eyes. Slit-lamp examination and intraocular pressure were both normal. Fundus examination of the right eye was unremarkable. Fundus examination of the left eye revealed scattered, round, yellowish spots at the level of the choroid, mostly visible in the nasal periphery (Figure 1A). Fluorescein (FA) and indocyanine green angiography (ICGA) of the left eye showed papillitis, rare hyperfluorescent spots on FA corresponding to certain choroidal nodules, and intermediate-phase ICGA showed many scattered hypofluorescent spots, including some in the macular area which were not visible on the fundus examination or FA (Figure 1 B-C). FA and ICGA were normal in the right eye and thus confirmed unilateral disease (not shown).

Considering the history of parotid gland sarcoidosis, choroidal nodules in the setting of ocular sarcoidosis were strongly suspected. Work-up for other causes of granulomatous choroiditis (syphilis, tuberculosis, Lyme disease, Birdshot retinochoroiditis) was negative. Laboratory studies showed an angiotensin converting enzyme (ACE) level of 225 units per liter (reference range, 20 to 70) and hypergammaglobulinemia. Computed tomography (CT) scan of the chest, abdomen and pelvis revealed numerous infracentimetric mediastinal, periaortic and splenic hilar lymphadenopathy. There were elevated cutaneous lesions on the right forearm. A diagnosis of sarcoidosis recurrence with lymph node, cutaneous and ocular involvement was made.

Sarcoid uveitis is generally bilateral (75-90%) and tends to be symmetrical (1). However, choroidal

Received: 20 July 2023
Accepted: 20 August 2023
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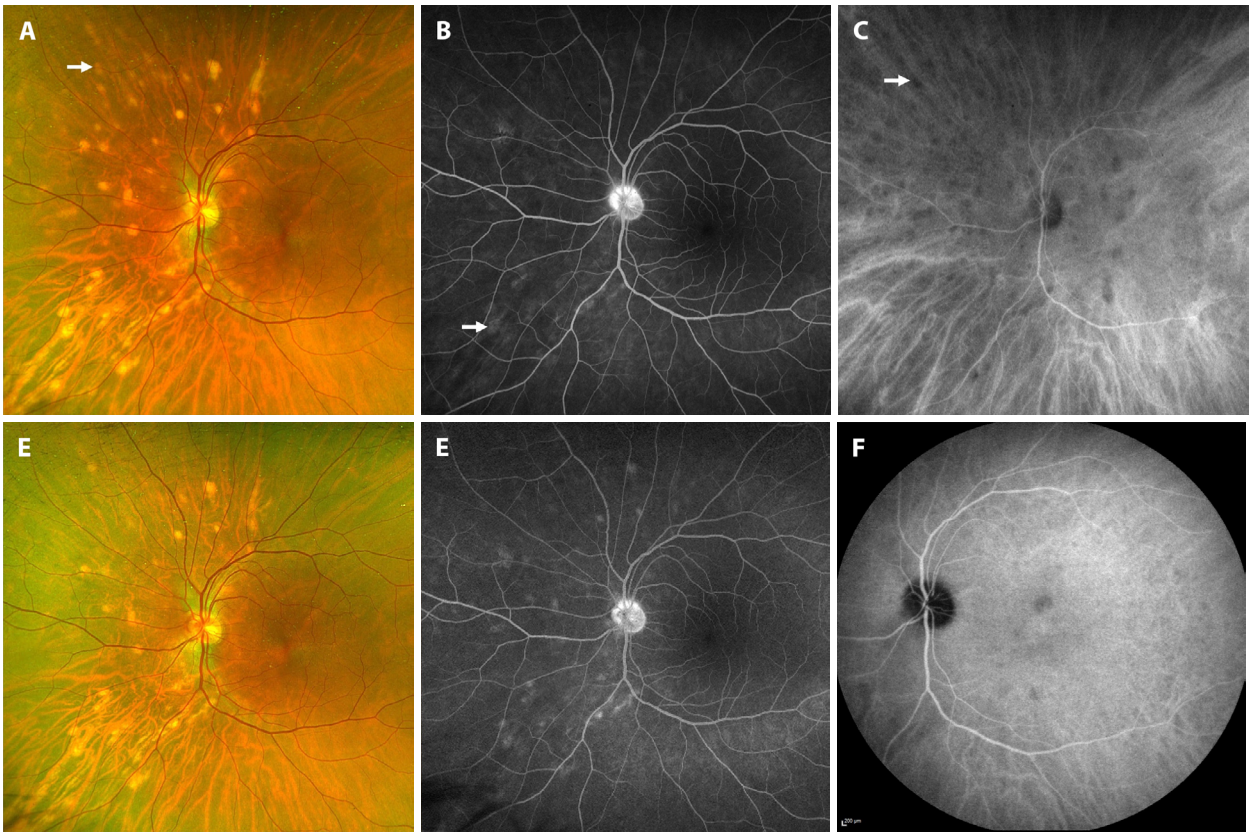

Figure 1. Multimodal imaging of choroidal nodules in the left eye, before and after steroid treatment. A-C: Imaging before treatment. A) Fundus photograph showing scattered, round, yellowish spots at the level of the choroid, mostly visible in the nasal periphery (arrow). B) Ultra-wide field (UWF) fluorescein angiography (FA) showing papillitis and some faint hyperfluorescent spots in the nasal periphery (arrow). There is no retinal vasculitis. C) Intermediate-phase UWF indocyanine green angiography (ICGA) showing multiple hypofluorescent spots in the nasal periphery (arrow), and also some spots in the macular region (which were not visible on fundus examination). D-F: Imaging 6 months after treatment initiation. D) Fundus photograph showing reduction in choroidal spot size and number. Some hypopigmented choroidal spots can still be seen however. E) UWF FA showing hyperfluorescence (window defects) at the level of the hypopigmented choroidal spots, indicating some degree of chorioretinal atrophy. Some mild papillitis can still be seen on the superior aspect of the disc. F) Intermediate-phase 55° ICGA showing reduction in the number of spots in the macular area. UWF ICGA showed reduction in spot size and number, with some hypofluorescent spots remaining corresponding to choroidal scars (unfortunately quality too poor for publication).

sarcoid nodules are an exception to this rule, as they generally arise unilaterally (2). Choroidal nodules are a classical finding in ocular sarcoidosis, and they tend to be highly responsive to oral corticosteroids (1,2). Refractory cases can benefit from conventional immunosuppressants like methotrexate (3), and biologics like the TNFα antagonists infliximab or adalimumab (3, 4).

The patient received treatment with 1 mg/kg/day of oral prednisone (starting dose of 70 mg/day) with gradual tapering. At follow-up 6 months later, while the patient was under 7 mg of prednisone per day, there was no recurrence of parotid gland enlargement, complete regression of the cutaneous lesions, reduction in lymphadenopathy on chest and abdominal CT scan, reduction in papillitis on FA, and reduction of choroidal spot size and number on ICGA (Figure 1 D-F). Some hypopigmented choroidal spots were still present on fundus examination after treatment however. These were hyperfluorescent on FA (Figure 1E) and hypofluorescent on late-phase ICGA (not shown), corresponding to inactive choroidal scars from previous asymptomatic granulomatous flares having left way to chorioretinal atrophic spots (5).

This case shows how the spectrum of sarcoidosis may evolve over time, from a mild parotid gland sarcoidosis initially to a multi-systemic presentation (lymph node, cutaneous, ocular) many years later. Treatment with oral glucocorticoids was essentially guided by the presence of multifocal unilateral choroidal nodules which if left untreated may lead to chorioretinal atrophy and secondary choroidal neovascularization (2).

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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