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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

A case of multifocal presumed solitary circumscribed retinal astrocytic proliferation lesions in the same eye

Presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP) is a recently described retinal tumour that is a distinct entity from other white lesions of the retina such as astrocytic hamartoma and acquired astrocytoma.[1] The exact origin of these lesions is of some debate within the literature as no histological diagnosis exists to date.[2,3] These lesions are not associated with tuberous sclerosis complex or related syndromes and can be distinguished from astrocytic hamartomas with imaging.[4] They have been reported in middle-aged patients and do not appear to be congenital in nature and are not associated with features of any systemic disease.[1,2] In the majority of the reported cases, visual acuity is maintained at better than 20/50; however, there is one report of a PSCRAP lesion involving the fovea.[3] With foveal involvement, the patient reported a gradual change in vision in adulthood, supporting the hypothesis that these lesions are acquired rather than congenital. This unique case reports a patient with multifocal PSCRAP lesions in 1 eye, whereas PSCRAP lesions have been previously reported only as solitary.

A 33-year-old, asymptomatic man with no personal or family history of tuberous sclerosis or neurofibromatosis was referred for evaluation of 2 pearly-white concretions in the retina of the left eye with no associated subretinal fluid, hemorrhage, or traction. His medical history was significant for asthma and type 2 diabetes mellitus. Visual acuity at presentation was 20/25 OD and 20/30 OS. Anterior segment was unremarkable. Dilated fundus examination (Fig. 1A, B) revealed a normal-appearing optic disc with 2 peripapillary pearl white retinal lesions, one temporal to the optic disc measuring approximately 1.0−1.2 mm in size and another small, white lesion superotemporal to the optic disc measuring approximately 0.5 mm in size (Fig. 1A). There was retinal pigment epithelial hyperplasia adjacent to or surrounding the lesions (Fig. 1B). Fundus autofluorescence demonstrated moderate hyperautofluorescence of both lesions (Fig. 1C). Optical coherence tomography−angiography (OCTA; Zeiss Cirrus 5000, Carl Zeiss, North York, Ont.) demonstrated the presence of an outer intraretinal mass deep to the overlying retinal vasculature (Fig. 1D). The superficial retinal vascular plexus appeared intact with no intrinsic vascularity. The vasculature, similar to the neuroretinal layers overlying the lesion, appeared compressed forward. Spectral domain optical coherence tomography (OCT) (SD-OCT; Zeiss Cirrus 5000) demonstrated a hyper-reflective, intraretinal mass with an abrupt elevation, optical shadowing, and a smooth surface with draping of the overlying retinal tissue (Fig. 1E, F). The lesions appeared to be originating from the outer retina or retinal pigment epithelium (RPE) with no underlying subretinal fluid. This was more apparent in Figure 1F as the lesion was much smaller, clearly demonstrating the preserved inner retinal layers. B-scan ultrasonography demonstrated absence of calcium in both lesions. The lesions remained stable with no changes after 6 months of follow-up.

PSCRAP was first described by Shields et al. in 2011.[1] These pearl-white lesions are rare benign retinal tumours that are discrete entities from other white retinal lesions and have unique and consistent characteristics on multimodal imaging.[1,2,3] All lesions reported tend to remain stable over long-term follow-up aside from 2 cases that spontaneously resolved.[1,5] Table 1 summarizes all published cases of PSCRAP and the corresponding characteristics of these lesions. PSCRAP can be distinguished from similar-appearing lesions such as astrocytic hamartomas by the appearance on OCT.[2,4,6] SD-OCT analysis in several recently published cases demonstrates the discrete separation of PSCRAP lesions from the overlying retinal nerve fibre layer (RNFL).[2−4] This is in contrast to OCT analysis of astrocytic hamartomas, which show an elevated or dome-shaped appearance confined to the RNFL, which is hyperreflective, with subtle posterior shadowing of the deeper retinal structures and a normal RPE.[6] Furthermore, PSCRAP is not associated with extension or adhesions into the vitreous like those reported with astrocytic hamartomas.[6] In addition, OCTA in the present case demonstrated a normal retinal vasculature of the surrounding superficial plexus as well as no intrinsic vascularity within the PSCRAP lesions. This is consistent with intravenous fluorescein angiography and OCTA in previous reports.[1,3] In contrast, astrocytic hamartomas demonstrate an intrinsic blood supply.[5] Therefore, OCTA analysis is a noninvasive method to further delineate PSCRAP from other white retinal lesions.

These pearl-white retinal lesions first coined as PSCRAP lesions originally were thought to derive from astrocytes of the RNFL.[1] With the aid of extended depth SD-OCT more recently, it was postulated that PSCRAP lesions may originate from fibrous metaplasia of the RPE or, alternatively, may be of retinal glial origin rather than derived from retinal astrocytes as first suggested.[2,3] However, on fundus autofluorescence imaging, RPE metaplasia should appear hypoautofluorescent because there is no intrinsic or extrinsic lipofuscin production. Both lesions in our case produced

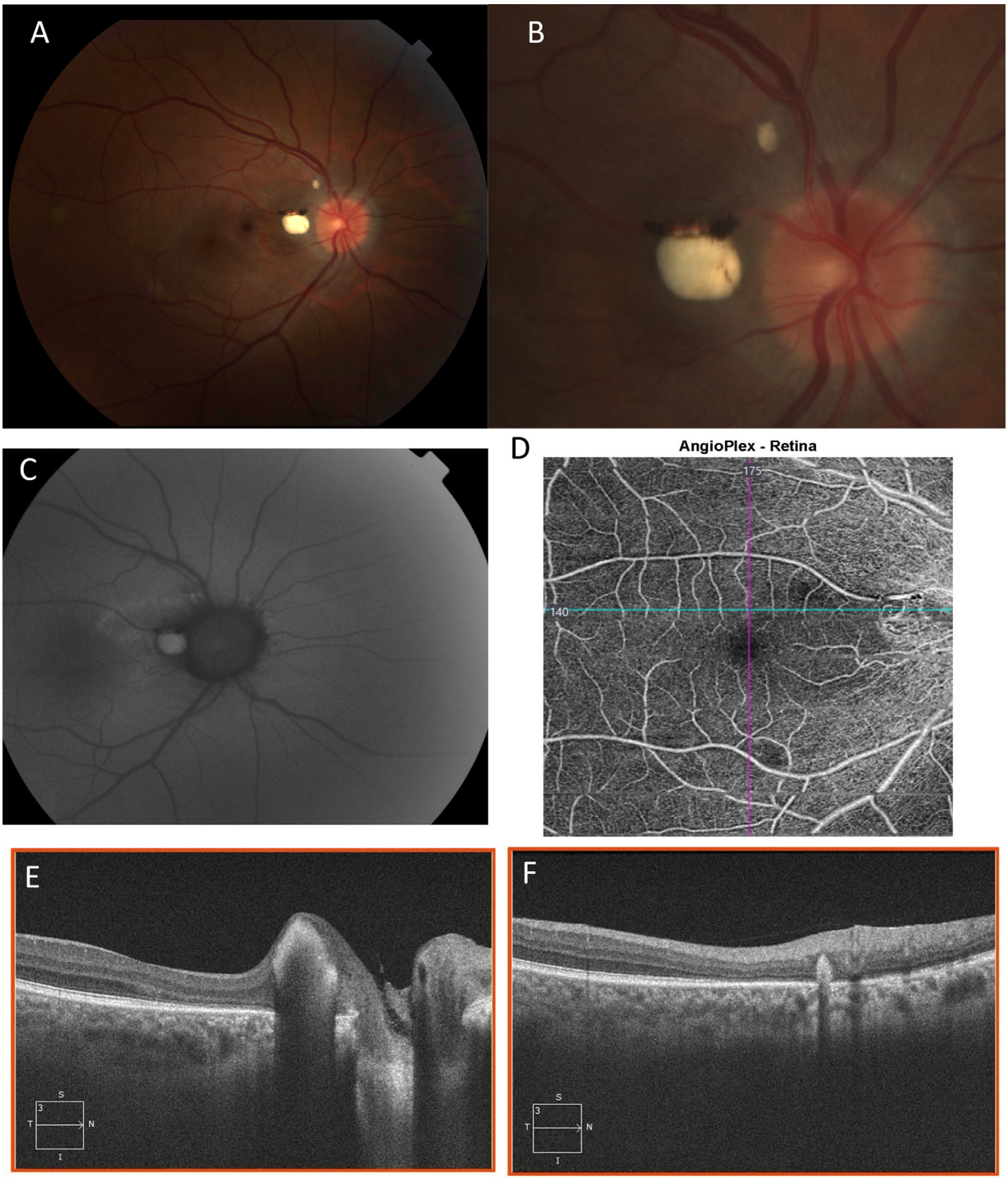

Fig. 1—(A) Colour fundus photograph of the right eye, demonstrating the position of the 2 white retinal lesions. (B) Higher magnification of the posterior pole of the right eye, highlighting the 2 distinct presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP) lesions. (C) Fundus autofluorescence of the right eye; note the low-moderate hyperautofluorescence of the larger lesion and isoautofluorescence of the smaller lesion in the right eye. (D) Optical coherence tomography–angiography (OCTA) analysis of the superficial retinal vasculature of the right eye. Spectral domain OCT analysis of the right eye demonstrating the larger lesion (E) and the smaller lesion (F).

slightly increased autofluorescence. Lipofuscin and calcium may appear hyperautofluorescent, but this was not present on clinical examination or B-scan ultrasonography.

To our knowledge, this is the first case of multifocal PSCRAP lesions in the same eye confirmed with multimodal imaging. All previous reports of PSCRAP described these lesions as unifocal and solitary in nature (see Table 1).[1–5,7] Furthermore, multimodal imaging suggests that these lesions do not appear to be astrocytic in etiology, but rather may originate from RPE or glial cells in the outer retina.[2,3] Therefore, as recently proposed, the term "PSCRAP" appears to be a misnomer in addressing these retinal pearl-like lesions.[2,3] With the

Table 1—Summary of all published reports of presumed solitary circumscribed retinal astrocytic proliferation lesions, including the present multifocal case

Age (y), Sex	Visual Acuity	Presenting Complaint	Anatomical Location	Stability	Reference
33, M	20/25	Nil	Multifocal: juxtapapillary and superotemporal to disc	Stable	Present study
46, M	20/20	Nil	Temporal equator	Stable	Shields et al. (2011)[1]
53, M	20/30	Decrease vision/cataract	Inferotemporal, postequatorial	Stable	Shields et al. (2011)[1]
37, M	20/20	Nil	Juxtapapillary	Stable	Shields et al. (2011)[1]
43, M	20/20	Nil	Superior equator	Stable	Shields et al. (2011)[1]
78, M	20/30	Decrease vision/cataract	Nasal equator	Stable	Shields et al. (2011)[1]
85, F	20/20	Nil	Superior to disc	Stable	Shields et al. (2011)[1]
76, F	20/25	Nil	Inferior to disc	Regression	Shields et al. (2011)[1]
57, F	20/20	Nil	Juxtapapillary	Not reported	Schwartz and Harbour (2015)[4]
61, M	20/50	Nil	Macula	Not reported	Shields et al. (2017)[2]
75, M	20/25	Nil	Nasal equator	Not reported	Shields et al. (2017)[2]
75, F	20/40	Floaters	Superior equator	Not reported	Shields et al. (2017)[2]
46, F	20/20	Nil	Temporal equator	Not reported	Shields et al. (2017)[2]
58, M	20/20	Nil	Inferonasal to disc	Stable	Asensio-Sánchez and Díaz-Cabanas (2017)[7]
74, F	20/250	Metamorphopsia	Macular	Not reported	Goldberg and Raja (2018)[3]
56, F	20/20	Nil	Juxtapapillary	Regression	Asensio-Sánchez (2019)[5]

emerging use of swept-source OCT and extended depth imaging it may be possible to further elucidate the origin of these "retinal pearls" in the near future.

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Solitary reticulohistiocytoma: a rare ocular surface mass

Reticulohistiocytoma, also known as solitary epithelioid histiocytoma, is a rare but well-documented histiocytic proliferation of the skin and soft tissues.[1] It is a non-Langerhans cell histiocytic lesion that typically affects adults. According to the revised classification of histiocytic disorders, solitary reticulohistiocytoma (SRH) is listed under the C group (xanthogranuloma family).[2] Another form, which is also listed under the C group, is multicentric reticulohistiocytosis, in which patients present with extensive cutaneous and joints involvements.[2]

As the name implies, SRH presents as an isolated lesion without systemic involvement.[3] The age of onset of cutaneous SRHs ranges from 2.5 to 74 years.[1] To the best of our knowledge, there are only 2 cases of isolated, histopathologically confirmed SRH of the ocular surface reported by Allaire et al.[3] Both cases were aged 21 years, females, and medically free. The first patient presented with an ovoid, salmon-colored, inferonasal limbal mass with minimal encroachment on the peripheral cornea. The lesion started 2 months before the patient's presentation. The clinical appearance suggested a lymphoid lesion.

The second patient had a history of a slowly growing mass covering the left cornea for which she underwent a keratoplasty, but the excised lesion was not available for microscopic examination. However, clinical examination of the same eye demonstrated a "large, polypoid, fatty-like growth covering the lateral aspect of the cornea with encroachment