Multimodal Imaging in a Case of Presumed Solitary Circumscribed Retinal Astrocytic Proliferation

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Keywords
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Abstract
Provisionally referred to as presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP), the lesion is a rare, benign retinal tumour that typically presents as white-yellow, opaque, and well circumscribed. Typically, the lesion is stable or may regress spontaneously. In light of the adjacent pigmentation of the tumour and from our retinal imaging, we suggest that the lesion originates from the deep neurosensory retina or the retinal pigment epithelium. Herein, we present a case of this entity in a 36-year-old man with a roundish, parapapillary tumour in his right eye and share its characteristics in the different diagnostic imaging modalities.

Clinical Case
A 36-year-old male patient was referred to us for a roundish parapapillary tumour in his right eye, which was noticed during a routine examination. He had no personal or family history of tuberous sclerosis complex. The refraction of his right eye was $+2.25 \times -1.75/54^\circ$,
and his best-corrected visual acuity was 20/16 (Snellen), while best-corrected visual acuity in the left eye was 20/12.5. Intraocular pressure in his right and left eye was 13.0 and 10.0 mm Hg, respectively. He reported no symptoms. Slit-lamp examination revealed no significant findings in the anterior segment. His pupils were round, medium in size, and isocor, and there were no signs of an afferent pupillary defect. Fundus examination of his left eye was normal. In his right eye, ophthalmoscopy revealed a circumscribed, roundish, yellow opaque lesion just superior to the disc in between the temporal vein and artery blood vessels with a diameter of roughly 1.1 mm × 0.8 mm (shown in Fig. 1a). The temporal edge showed slight hyperpigmentation. No feeder, draining, or network vessels were observed. Optical coherence spectral tomography (OCT) using the SPECTRALIS® (Heidelberg Engineering, Heidelberg, Germany) showed a hyperreflective 0.7-mm dome-shaped prominent mass in the inner retina with an overlying intact appearing retinal nerve fibre layer and a slight optical acoustic shadow in the outer retinal layers (shown in Fig. 1b). The border was well defined. In the AngioPlex® OCT Angiography (CIRRUS™ HD-OCT Model 5000; Carl Zeiss Meditec, Jena, Germany), no intrinsic vasculature of the lesion was observed, although there were significant artefacts from overlying retinal vasculature (shown in Fig. 1b, c). Fluorescein angiography showed mild hyperautofluorescence (shown in Fig. 1d), a regular arteriovenous transit time, and a slight hyperfluorescence (shown in Fig. 1e) of the lesion with an increase to the late phase without any signs of network vessels in the retinal mass. B scan ultrasonography demonstrated an elevated retinal mass without signs of calcification (shown in Fig. 1f). None of the above examinations showed associated subretinal fluid, haemorrhage, or retinal traction.

Discussion
In 2011, Shields et al. [1] reported 7 patients with an asymptomatic, solitary, opaque, or yellow retinal mass and named this entity presumed solitary circumscribed retinal astrocytic proliferation (PSCRAP). The exact pathogenesis and pathological features of PSCRAP remain unknown, and only few case reports have been published so far. PSCRAP seems to occur more frequently in middle-aged males. Typically, these benign lesions are solitary and unilateral, opaque, develop abruptly, and are not associated with retinal exudation, feeding vessels, intrinsic vascular flow, vitreoretinal traction, or tuberous sclerosis complex and other systemic diseases. A more recent report detailed the first known case of multiple lesions in one eye [2]. To date, there is still no histological confirmation of the features observed during in vivo imaging.

On OCT, the lesions present with several interesting characteristics [3, 4]. In line with previous reports, we found severe disruption of the outer retinal layers with displacement of retinal tissues by a well-defined, hyperreflective mass. The nerve fibre layer seems to be unaffected apart from the mass displacement. Angiography confirmed the absence of any intrinsic vasculature. Similarly, OCT angiography did not show measurable blood flow within the lesion. However, there were significant artefacts due to large retinal vasculature with blood flow in the overlying layers, interfering with accurate interpretation. Also, segmentation of the retinal layers proved challenging as the retinal structure was severely disrupted by the tumour. In accordance with Goldberg and Raja, we suggest that the lesion is more fibrous and not derived from the retinal astrocytes [5]. We therefore also agree that the term astrocytic proliferation represents a misnomer, given the growing evidence for its deeper retinal origin. As in our case, some PSCRAP lesions are associated with adjacent pigmentation. We interpret this finding in the context of a reactive process of the retinal pigment epithelium (RPE), suggesting that the RPE may play a role or at least be affected
Fig. 1. a Fundus photo shows a circumscribed, roundish, yellowish lesion just superior to the disc. b OCT scan revealed a dome-shaped prominent mass in the inner retina. OCT angiography shows no intrinsic vasculature but significant artefacts from overlying layers. c C-scan of the deep vascular plexus from OCT angiography. d In the fundus autofluorescence, the lesion appears as mild hyperautofluorescent. e Fluorescein angiography shows slight hyperfluorescence in the late phase. f B scan ultrasonography demonstrates no signs of calcification.
by the growth of the lesion. Whether this would indicate a potential weakness of the RPE with the risk of choroidal neovascular membrane formation remains open for discussion. To our knowledge though, no secondary choroidal neovascular membrane lesions have been reported so far.

Depending on the localization of the tumour, it may lead to visual field loss or even visual deterioration. In some reported cases, the benign tumour appeared parapapillary, which presented as an enlarged blind spot on perimetry. These negative effects are caused by loss of retinal structural integrity at the site of the lesion and do not extend elsewhere. However, if the lesion is located in the fovea, it may also lead to significant visual deterioration. So far, no treatment approach seems to be feasible, in part due to the often central location and small size of PSCRAP lesions. Interestingly, spontaneous regression within 1 year after initial diagnosis of the lesion has been described [6]. Due to our shorter follow-up, we cannot yet give a definite answer regarding the natural course in our case. Fortunately, the patient remains asymptomatic at the time of writing, and treatment is not indicated.

It is important to differentiate PSCRAP from other similar entities with serious ocular or systemic implications. Our initial differential diagnosis included inflammatory granuloma, but the choroid did not show any affection on subsequent imaging, and there was no indication of past or present ocular inflammation. Retinal astrocytoma was another early consideration, but OCT allowed us to quickly rule out a retinal astrocytic lesion as the inner retina was not affected. Importantly, no imaging modality was suggestive of an invasive growth. Given the small size of the lesion and its asymptomatic nature, we felt comfortable recommending observation and have not been informed of any deterioration so far.

**Conclusion**

We confirm that OCT represents a very useful, non-invasive tool in the workup for rare cases of PSCRAP. Moreover, the nomenclature should be revised to better represent the more likely glial origin of this entity while still awaiting histologic confirmation.

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**Statement of Ethics**

To publish this case report and any accompanying images, we received the written informed consent from the patient. This study was conducted in accordance with the Declaration of Helsinki. No vote of the Ethics Committee is required for this Case Report.

**Conflict of Interest Statement**

No potential competing interest was reported by the authors: K.P.K. (none); S.K. (none); M.D.B. (Roche, C); and F.M.H. (Roche, C)
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Author Contributions

Klemens Paul Kaiser co-wrote the main text and designed the case report. Stephan Kinzl performed the clinical examination and workup and reviewed the manuscript. Matthias Dieter Becker reviewed the manuscript and provided critical feedback. Florian Moritz Heussen co-wrote the main text, helped with the design, and provided critical feedback.

Data Availability Statement

All data generated or analysed during this study are included in this article. Further enquiries can be directed to the corresponding author.

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