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Sir,

Response to Grzybowski and Justynska

We thank Dr Grzybowski and Justynska[1] for their interest in our article.

In response to their comments on our report[2] we acknowledge the inadvertent omission of articles that emphasize the occurrence of inflammatory-marker-negative disease seen in giant cell arteritis (GCA). Unfortunately some papers referred to were not published at the time of writing.[3,4] The table provided by Dr Grzybowski and Justynska highlights some important articles, some of which were referenced in our original report[5] and others which were summarized by key articles referenced.[6,7,8]

Dr Grzybowski and Justynska remark that typical features commonly associated with GCA were not presented in our case. However, the patient we described was indeed unique in that the patient did not display symptoms usually found in GCA other than AION-induced loss of vision and corresponding RAPD in a patient with known polymyalgia rheumatica. These features were described in our report. We fully agree that scrutinizing the clinical picture is critical in the diagnosis of GCA but would like to emphasize that this is exactly what we did. We specifically looked at the clinical presentation including increased pre-test probability due to ethnicity in order to come to the conclusion that the negative CRP needed to be ignored!

We are very grateful for the additional case reports and have integrated them into our original table, thereby giving a more detailed understanding of inflammatory-marker-negative disease (Table 1).

Table 1 Summary of current literature—revised and updated according to comments by Grzybowski and Justynska

	ESR-negative disease	*CRP-negative disease*	*ESR- and CRP-negative disease*
Parikh *et al*[5]	14.3%	1.7%	0.8%
Ellis and Ralston[9]	22.5% at initial presentation		
Weintraub[10]	1 case report		
Levy *et al*[2]		1 case report	
Laria *et al*[4]			1 case report
Kermani *et al*[3]			4%
Yoeruek *et al*[11]			1 case report
Raja *et al*[12]			1 case report
Man and Dayan[13]			1 case report
Poole *et al*[14]			1 case report

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Is the mechanism of 'poppers maculopathy' photic injury?

I read with interest the excellent series by Davies *et al*,[1] describing maculopathy in patients using 'poppers.' Together with two recent series from France,[2,3] their report provides important evidence for an association between abuse of alkyl nitrite compounds and specific, sub-foveal changes. Whether this association is causal remains to be determined, and Davies *et al* suggest causality is likely.

What I find most striking about the cases attributed to 'poppers maculopathy' in the Davies series (and which is consistent with the two series from France) is the SD-OCT imaging—which has an uncanny resemblance to photic maculopathy (Figure 1). In both 'poppers maculopathy' and photic maculopathy, there is focal disruption of the IS-OS junction centred at the fovea.[1–4] Moreover, the size, shape, echogenicity, and temporal evolution of the SD-OCT lesions appear indistinguishable in the two conditions. Patients also present with the same symptoms (scotoma, reduced vision, and phosphenes) and have the same slit-lamp findings (a pale yellow foveal lesion).[1–4] Indeed, it appears that in people using poppers, the two conditions can only be reliably distinguished by eliciting a history of excess light exposure and not by clinical features.

Unfortunately, Davies *et al*[1] did not report to what extent excessive light exposure was specifically queried in their patients. In the two French series,[2,3] all patients 'denied staring at bright lights'—yet how reliable is their history? Poppers are frequently combined with psychotropic drugs and alcohol, which can alter consciousness and memory, potentially making history unreliable.[5] Poppers themselves can cause transient visual hallucinations and heighten sensory perception[5]—effects that are known to increase light-gazing behaviour in other recreational drugs such as LSD.[6] Poppers are frequently used in raves, where bright strobe lights and lasers are common.

Given the points discussed, it should be crucial when considering the diagnosis of 'poppers maculopathy' to document a thorough history of the patient's drug behaviour and light exposure. Do they take multiple drugs? Do they hallucinate or experience altered consciousness? Are they ever entranced by bright lights, candles, or the sun?

The endemic use of poppers[4] and the mere handful of reports of maculopathy suggests that compounding factors or susceptibilities may be involved. It is not inconceivable that 'poppers maculopathy' represents a sub-group of patients who have unrecognised photic injury. If poppers maculopathy is indeed a distinct entity, then the remarkable ultrastructural similarity with photic injury suggests that the two conditions share a common

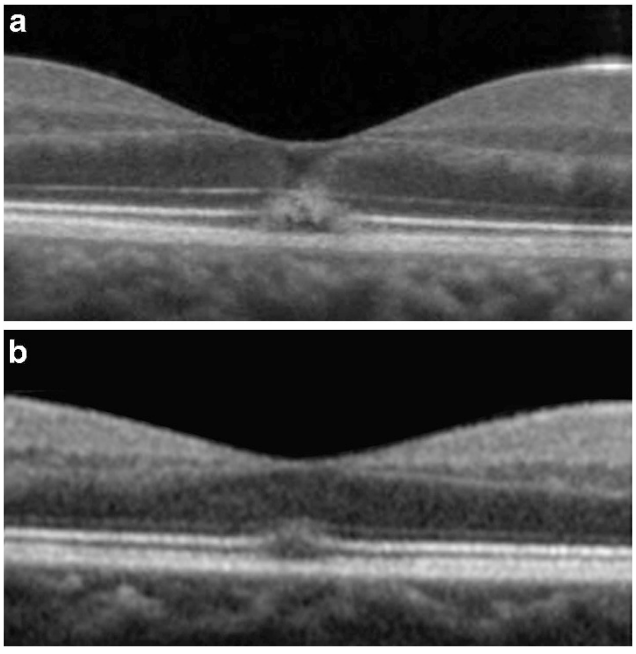

Figure 1 A comparison of SD-OCT images in (a) 'poppers maculopathy' as presented in Case 2 of Davies *et al*[1] with (b) photic retinopathy in a 30-year-old male who presented to my clinic 2 weeks after sun-gazing. Notice the similarity in location, size, shape, and echogenecity of the respective lesions in the IS-OS junction.