



Symptomatic idiopathic bilateral multifocal retinal pigment epithelial detachments

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ABSTRACT

Purpose: To report a rare case of idiopathic bilateral serous retinal pigment epithelial detachments.

Observations: We present a case of a 43-year-old female patient with metamorphopsia with the presence of innumerable bilateral serous retinal pigment epithelial detachments. Detailed ocular and systemic clinical work-up revealed no underlying disease.

Conclusions: /**Importance:** Idiopathic multiple serous retinal pigment epithelial detachments are an extremely rare condition, that has been observed as an incidental clinical finding in asymptomatic patients. Multimodal imaging and detailed systemic work up are essential to rule out an underlying disease. Initial symptoms, as in our case, are uncommon and usually occur only in patients with complications such as choroidal neovascularization or hemorrhages. Therefore, regular follow ups are recommended to early detect and treat possible complications.

1. Introduction

A retinal pigment epithelial detachment (PED) results from the disruption of the junction between the basement membrane of the retinal pigment epithelium (RPE) and the inner collagenous layer of Bruch's membrane.¹ Single or multiple serous retinal PEDs are common in a variety of eye diseases such as age-related macular degeneration, central serous chorioretinopathy (CSCR), polypoidal choroidal vasculopathy, angioid streaks and Vogt-Koyanagi-Harada disease and hereditary chorioretinal degenerations.²⁻⁴ PEDs have also been described in patients with renal disorders, malignant hypertension, systemic hypercortisolism and in acute leukemias.⁵⁻⁷ Idiopathic PEDs, without an underlying disorder, are a very rare condition and only a few cases have been published so far.⁸⁻¹⁰ We present a case of multiple diffuse serous retinal PEDs in a 42-year old female complaining of metamorphopsia.

2. Case report

A 42-year-old women described metamorphopsia in both eyes since the last 2 weeks. Upon examination her best-corrected visual acuity was 20/15 in her right eye and 20/20 in her left eye. Pupillary reflexes were normal. Bilateral anterior segment examination (including intraocular pressures) was unremarkable. Fundus examination revealed multiple

well circumscribed yellowish elevations surrounded by a darker halo in the macula and periphery in both eyes, involving mostly the temporal fundus, but they were found circularly (Fig. 1). The PEDs located in the central macular area were larger and showed greater variation in their shape and size than the PEDs in the periphery. Spectral domain optical coherence tomography (SD-OCT) showed that these lesions were serous PEDs (Fig. 2 right). They correlated with hyporeflective areas on near-infrared imaging (Fig. 2 left). The subfoveal choroidal thickness was 330µm. Fundus autofluorescence showed multiple hyperautofluorescent spots corresponding to the PEDs.

Fluorescence angiography (FA) demonstrated well circumscribed hyperfluorescent lesions without any evidence of leakage in the early and late phases in both eyes (Fig. 3 a-b). Indocyanine green angiography (ICG) showed corresponding hypercyanescence lesions of the retina in the late time frames (Fig. 3 c-d). Thus, a choroidal tumor or an infectious origin of the lesions could be ruled out. Automated visual field testing was unremarkable.

Detailed ocular and systemic history were taken to rule out any ocular or systemic diseases.

The patient was not taking any systemic medication. No chronic diseases were diagnosed previously, in particular no arterial hypertension or kidney disease. The patient had never undergone treatment with steroids. The patient was not pregnant either.

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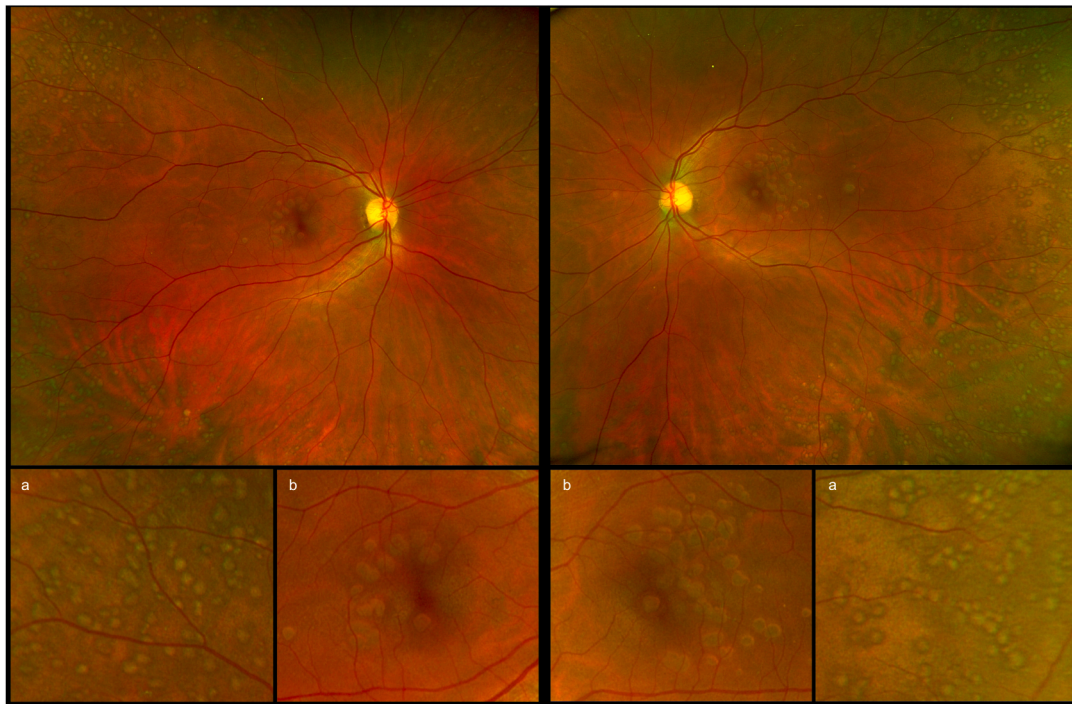


Fig. 1. Scanning laser ophthalmoscopy images demonstrating multiple well circumscribed serous retinal pigment epithelial detachments in the macula (a) and temporal retinal periphery (b) in both eyes.



Fig. 2. Near-infrared imaging of the macula (left) and SD-OCT of the macula (right) showing.

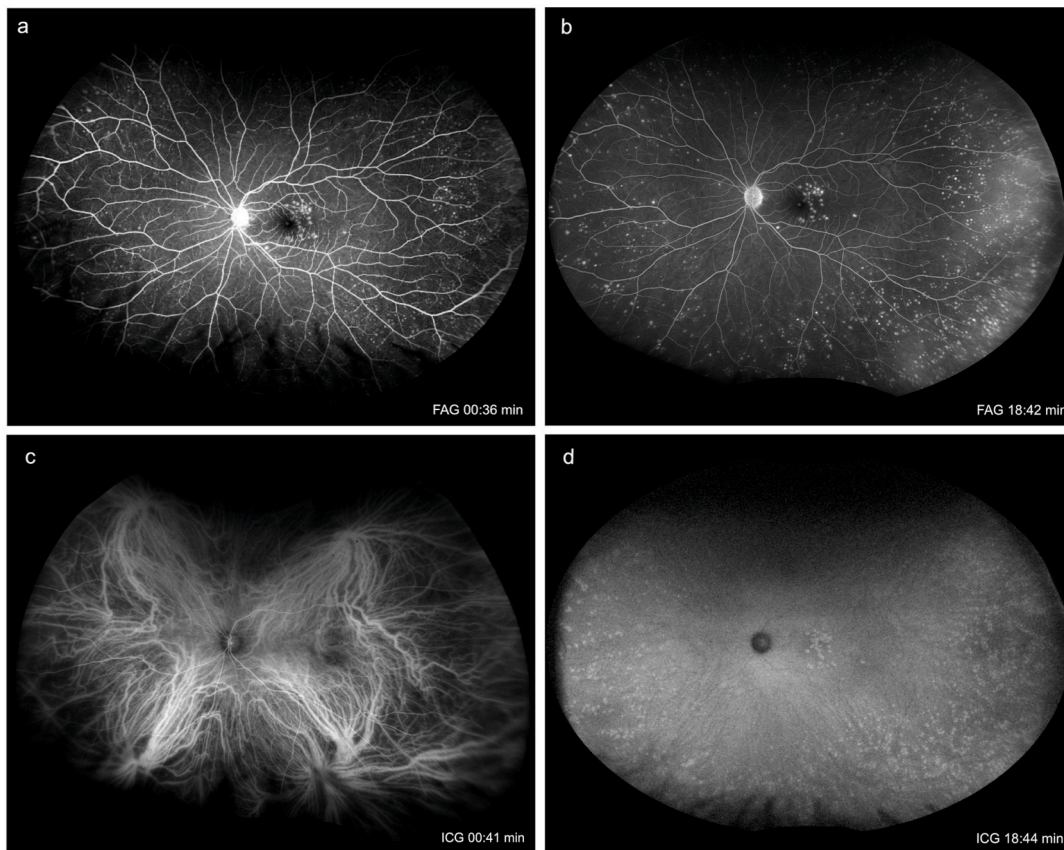


Fig. 3. Fluorescein angiography (a–b) shows hyperfluorescent well circumscribed lesions in the early phase and late phase. Fundus indocyanine green angiography (c–d) reveals hypercyanescence lesions in the late phase.

A complete blood count, C-reactive protein, erythrocyte sedimentation rate, angiotensin converting enzyme, serum lipid levels, coagulation status, hormone levels and thyroid gland parameters were all within normal limits.

Elispot testing to detect tuberculosis was negative and the serological screening for other infectious diseases was unremarkable.

Her human leukocyte antigens (HLA) are A12 and A68. Specific anti-nuclear antibodies (ANA) subtypes (dsDNA AB, Histon AB, DFS70) were detected, but an autoimmune disorder, especially systemic lupus erythematosus, was ruled out by the Department of Rheumatology. The patient denied arthralgia. We referred her to an additional examination to the Department of Dermatology, but no indication for collagenosis was found. For further clarification the patient underwent a gynecologic, endocrinologic and nuclear medical check-up. The findings did not reveal any abnormalities.

In synopsis with the results of the clinical and laboratory tests, there was no evidence of an underlying malignancy.

She was evaluated regularly until 12 months after initial presentation. She described unchanged visual symptoms, but visual acuity improved to 20/15 Snellen on both eyes under local therapy with dorzolamide. SD-OCT imaging remained unchanged.

Regular systemic and laboratory check-ups did not show any changes and there was still no indication given for a systemic autoimmune disorder.

3. Discussion

Isolated serous PEDs in the macula are a frequent finding in healthy, middle-age patients with no associated etiology.¹ However, idiopathic multiple serous retinal PEDs without significant visual impairment are an extremely rare condition. Gass et al.¹⁰ initially described three

middle-aged patients with innumerable retinal PEDs without underlying disease and only a few similar case reports have been published so far. In most of these cases the multiple serous PEDs have been observed as an incidental clinical finding in asymptomatic patients.^{8–11} Symptoms occurred solely in patients with complications such as hemorrhagic PEDs or subretinal hemorrhage.^{11,12} In our case the patient was initially complaining of metamorphopsia without any complications.

Multimodal imaging with SD-OCT, FA and ICG is essential to confirm the serous nature of the PEDs and to rule out presence of a pachychoroid disease. The pachychoroid disease spectrum, that includes CSCR and polypoidal choroidal vasculopathy, is a relatively novel concept suggesting that formerly independent disease entities represent a continuous disease process driven by choroidal dysfunction.¹³ Diffuse or focal increased choroidal thickness visualized on OCT, hyperpermeability on ICG angiography, the loss of the inner choroid and dilated outer choroidal vessels (pachyvessels) as well as the presence of pachydrusen (drusenoid deposits) characterize the disease entities.^{14,15} As in our case, ICG angiography showed hypercyanescence lesions, but no signs of hyperpermeability or dilated choroidal vessels and there was no signs of increased choroidal thickness on SD-OCT we ruled out an underlying pachychoroid disease.

On the other hand, it has been postulated that idiopathic multiple serous retinal PEDs might be a variant of CSCR as it is mostly seen in middle-aged patients and the lesions show a pooling phenomenon similar to CSCR lesions in FA.^{8,11}

Single or multiple PEDs can also occur in several different systemic disorders like Vogt-Koyanagi-Harada-disease, sarcoidosis, systemic hypercortisolism, renal disorders, malignant hypertension, acute leukaemia and infections such as Cytomegalovirus (CMV).^{1–3,5,6,10,11} Therefore, systemic exam should be carried out including a complete blood count, erythrocyte sedimentation rate, angiotensin converting

enzyme, autoimmune markers, elispot testing, serum lipid and cortisol levels and chest X-ray as well as syphilis, CMV, herpes simplex and HIV serologies. Moreover vasculitis, collagenosis, malignant hypertension, hyperviscosity syndromes and lymphoproliferative disorders must be excluded.⁸

Currently, there is no known treatment for idiopathic multiple serous retinal PEDs. Gass et al.¹⁰ reports, that in one case thermotherapy to the central macular was applied as a treatment option but proved to be ineffective. Therefore, he recommended conservative management for patients with good visual acuity and no evidence of choroidal neovascularization. As our patient expressed symptoms, we treated her with dorzolamide eye drops twice a day which improved visual acuity to 20/15.

Topical carbonic anhydrase inhibitors can have an effect on the distribution of retinal fluid - in patients with CSCR - and thus can lead to a faster reduction of central macular thickness by improving the pumping function of RPE.¹⁶

We refrained from alternative, more invasive treatment options such as PDT laser or intravitreal injections, because of the patient's good visual acuity and the fact, that no choroidal neovascularization was detected. Besides, these treatment options could possibly cause decreased vision due to side effects such as choroidal atrophy or endophthalmitis. Hence, we have decided for a non-invasive therapy with dorzolamide.

4. Conclusion

In conclusion, idiopathic bilateral, multiple serous PEDs in middle-age healthy patients are a rare condition that usually are found in asymptomatic patients. Multimodal imaging and detailed systemic work up are essential to rule out an underlying disease. Symptoms like metamorphopsia are rare and commonly occur with complications such as choroidal neovascularization or hemorrhages. Therefore, regular follow ups are recommended to early detect and treat possible complications.

Patient consent

We obtained written consent of the patient to use her data and images.

Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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multiple pigment epithelial detachments as dome shaped elevations and a normal choroidal thickness.

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