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Multimodal imaging of unusual macular macroaneurysm rupture after navigated retinal laser in a patient with adult onset Coats disease

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ABSTRACT

Purpose: To report the uncommon rupture of a macular macroaneurysm (MAR) during navigated retinal laser (Navilas®) focal treatment in a patient with adult onset Coats disease.

Observation: A 30-year-old man consulted for progressive decrease of vision in his right eye from one week. Fundoscopy examination showed macular hard exudates, aneurysms, vascular telangiectasias in the temporal inferior quadrant consistent with an adult onset Coats disease (CD). Spectral domain optical coherence tomography (SD-OCT) and fluorescein angiography (FA) revealed macular edema, vessels abnormalities associate to non-perfused areas. Ultra-widefield optical coherence tomography angiography (UWF-OCTA) clearly showed the blood flow abnormalities in both superficial and deep capillary plexus. Focal laser photocoagulation of abnormal vessels by navigated retinal laser and intravitreal injections (IVT) of aflibercept, successfully resolved macular edema. During supplemental navigated focal laser treatment, a macular macroaneurysm rupture occurred, causing intravitreal hemorrhage with a self-limiting resolution in three months. Indeed, visual acuity progressively improved during follow-up and absence of macular edema was observed at 18 months.

Conclusion: Adult onset CD is a rare condition. Our patient presented an unusual intravitreal hemorrhagic complication due to a MAR rupture after focal navigated laser treatment. Despite this complication, early laser photocoagulation and IVT injections of anti-VEGF, successfully resolved macular edema. UWF-OCTA follow-up clearly showed abnormal vessels in both superficial capillary plexus (SCP) and deep capillary plexus (DCP) and successfully guided additional navigated focal laser treatment.

1. Introduction

In 1908, George Coats first described CD as characterized by idiopathic congenital retinal vessels' abnormalities (telangiectasia and aneurysms), subretinal and intraretinal lipid exudates, without retinal or vitreous tractions.[1] CD mainly occurs in childhood under 10 years old, and can cause severe visual loss.[2] It is frequently unilateral (95%) and predominantly affects males (75%).[2] Vessels' abnormalities are usually localized in the temporal inferior quadrant of the retina and involve the posterior pole in less than 5% of cases.[2] Adult-onset CD is less common and presents less extended vascular abnormalities, indeed it is characterized by slower and more benign natural course compared to the childhood-onset one.[3] Many therapies have been used to treat vascular abnormalities and complications of CD; laser photocoagulation, intravitreal vascular endothelial growth factor (VEGF) inhibitor, cryotherapy, diathermy, vitreous surgery of retinal detachment.[3] Recently a novel laser device has been introduced: the NAVILAS® (OD-OS GmbH, Teltow, Germany). It consists of a retinal navigation system and laser device incorporating a digital fundus imaging system that allows live red-free, infrared, and FA imaging useful for treatment planning.[4] In this paper we report the multimodal imaging of an unusual rupture of MAR during automatized Navilas® laser focal treatment in a patient with adult onset CD.

2. Case report

A 30 years old male presented to the ophthalmology emergency department for progressive decrease of vision in his right eye (RE) from one week. At baseline, best corrected visual acuity (BCVA) was 20/200 in the RE and 20/20 in the left eye (LE). Slit lamp and fundus examination of the LE were unremarkable. Conversely, fundus examination of the RE showed extensive macular hard exudates, retinal hemorrhages, microaneurysms, vascular telangiectasias in the inferior temporal area of the posterior pole and of the middle retinal periphery (Fig. 1A et 1B).

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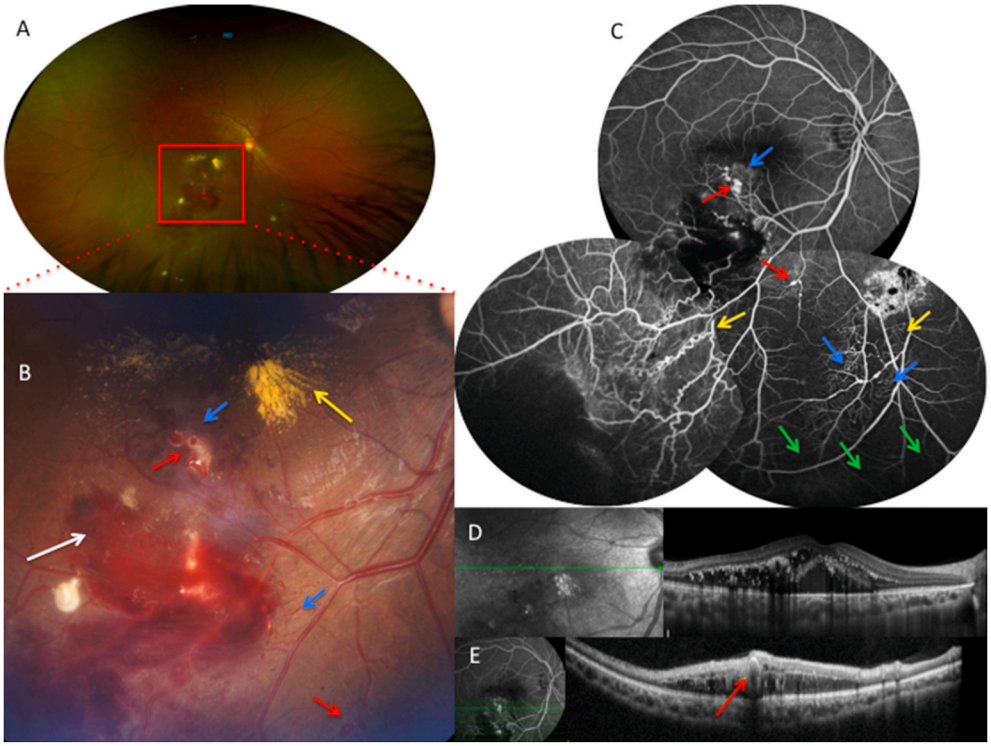

Fig. 1. Multimodal imaging of the RE of a patient with adult onset CD. Ultra Widefield color photograph (Optos PLC, Dunfermline, Scotland, UK) (A) and magnification of the macular region (B) showing central hard exudates (yellow arrow), retinal hemorrhages (white arrow), aneurysms (red arrow), telangiectasias (blue arrow) in the temporal inferior macular region (B). FA montage showing in intermediate phases (3min): pooling of telangiectasias (blue arrow) and aneurysms (red arrow), peripheral capillary dropout (green arrow) and vascular loops (yellow arrow) (C). SD-OCT showing increased macular thickness, intra retinal fluid, exudative retinal detachment (D) and aneurysms in the macular area (E). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

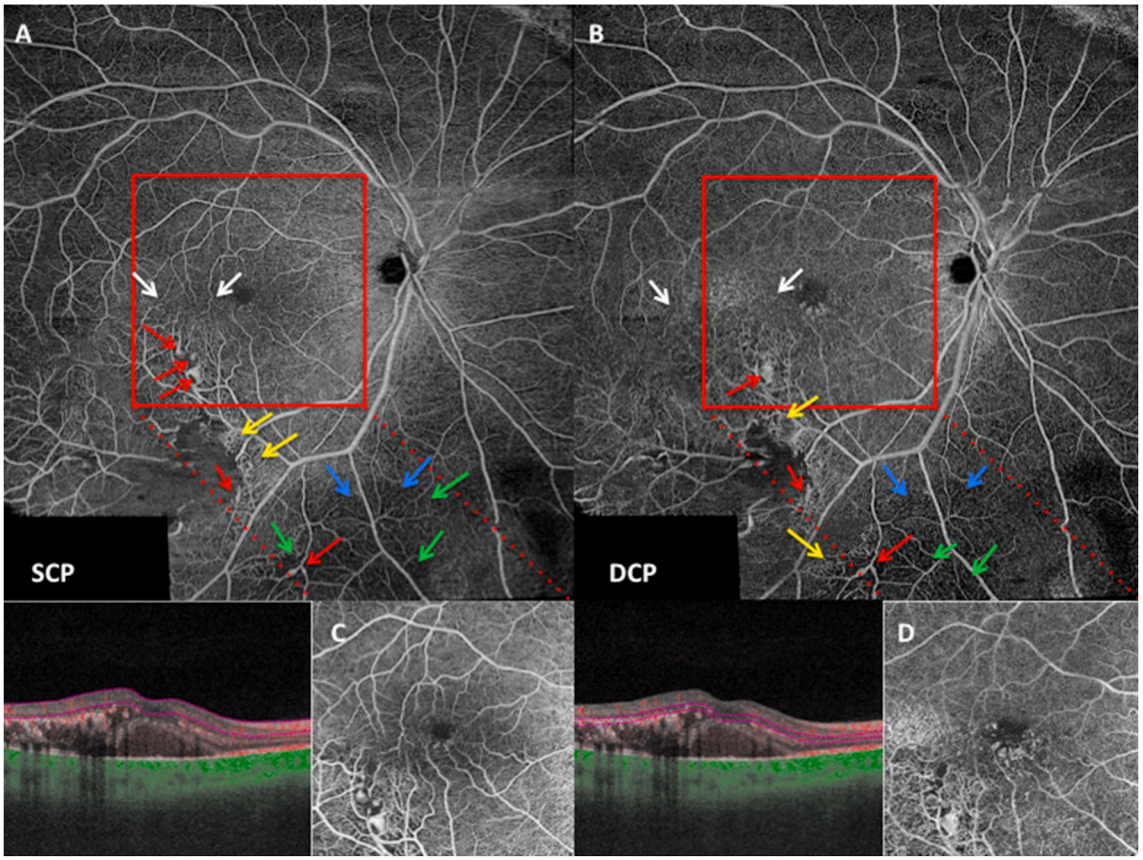

Fig. 2. UWF-OCTA (12mm × 12mm) montage (A, B) and 6mmx6mm (C, D) OCTA images centered on the macula with the corresponding B-scan passing trough the fovea (below) showing dilated capillary network (blue arrow), outpouchings of aneurismes and vascular beading (red arrow), capillary loops (yellow arrow), connections between superiors and inferiors vascular branches (white arrow), faint flow, dilated intercapillary space and non perfused areas (green arrow) in the SCP (A,C) and DCP (B, D). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

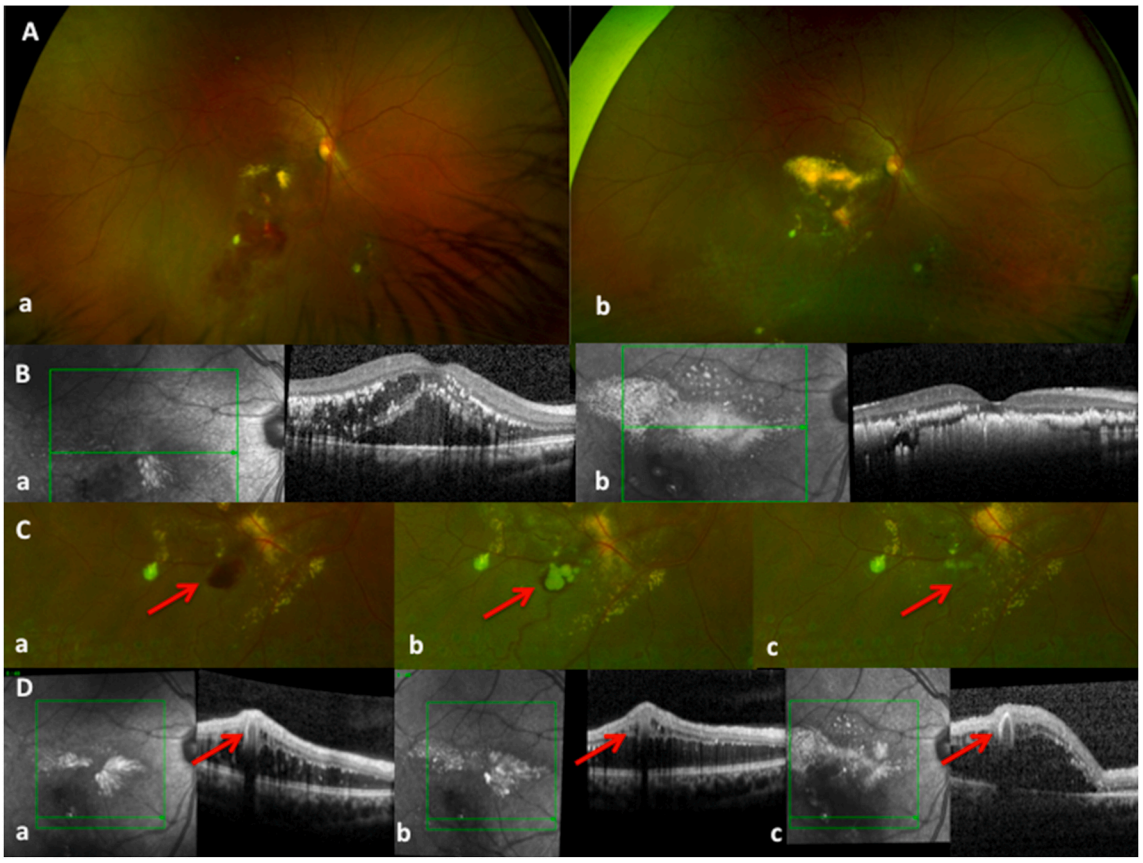

Fig. 3. Wiedfield color photos (Optos PLC, Dunfermline, Scotland, UK) (A) of the RE at baseline (a) and at 12 months of follow-up (b), demonstrating augmentation of lipid hard exudates in macular area and yellow laser photocoagulation marks in the temporal inferior area. SD-OCT (B) showing increased hard exsudate, fibrosis and atrophy of outer retina in macular area after laser photocoagulation and aflibercept IVT injections from baseline (a) to month 12 (b) of follow up. Color photos (Optos PLC, Dunfermline, Scotland, UK) (C) showing the macular macroaneurysm before the first navigated laser treatment (a) one month before navigated laser treatment (b) and the reperfusion of the macroaneurysm at 9 months follow up (c). SD-OCT (D) centered on the macroaneursym before the first navigated laser treatment (a) one month before navigated laser treatment (b) and at 9 months follow up showing the reperfusion of the MAR and increased macular edema (c). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

FA (Spectralis® HRA-OCT Heidelberg Engineering, Heidelberg, Germany) revealed leakage in the macular area, vascular loops, microaneurysms, telangiectasis vessels and non-perfused areas in the temporal inferior quadrant (Fig. 1C). SD-OCT, (Spectralis® HRA-OCT Heidelberg Engineering, Heidelberg, Germany) revealed macular edema with intra retinal fluid, exudative serous retinal detachment and hyperreflective intraretinal lesions corresponding to macroaneurysms and lipidic exudates (Fig. 1D et 1E). UWF-OCTA (Plex Elite 9000; Carl Zeiss Meditec) images obtained by montage of five 12mmx12mm squares, showed both in the SCP and DCP, dilated capillary networks, outpouchings of aneurisms, vascular beading, capillary loops, connections between superior and inferior vascular branches, faint flow, dilated intercapillary spaces

Fig. 4. UWF-OCTA (15mmx9mm) in the SCP showing vascular anomalies after hemorrhage resorption (a) baseline (b) at 7 months follow up.

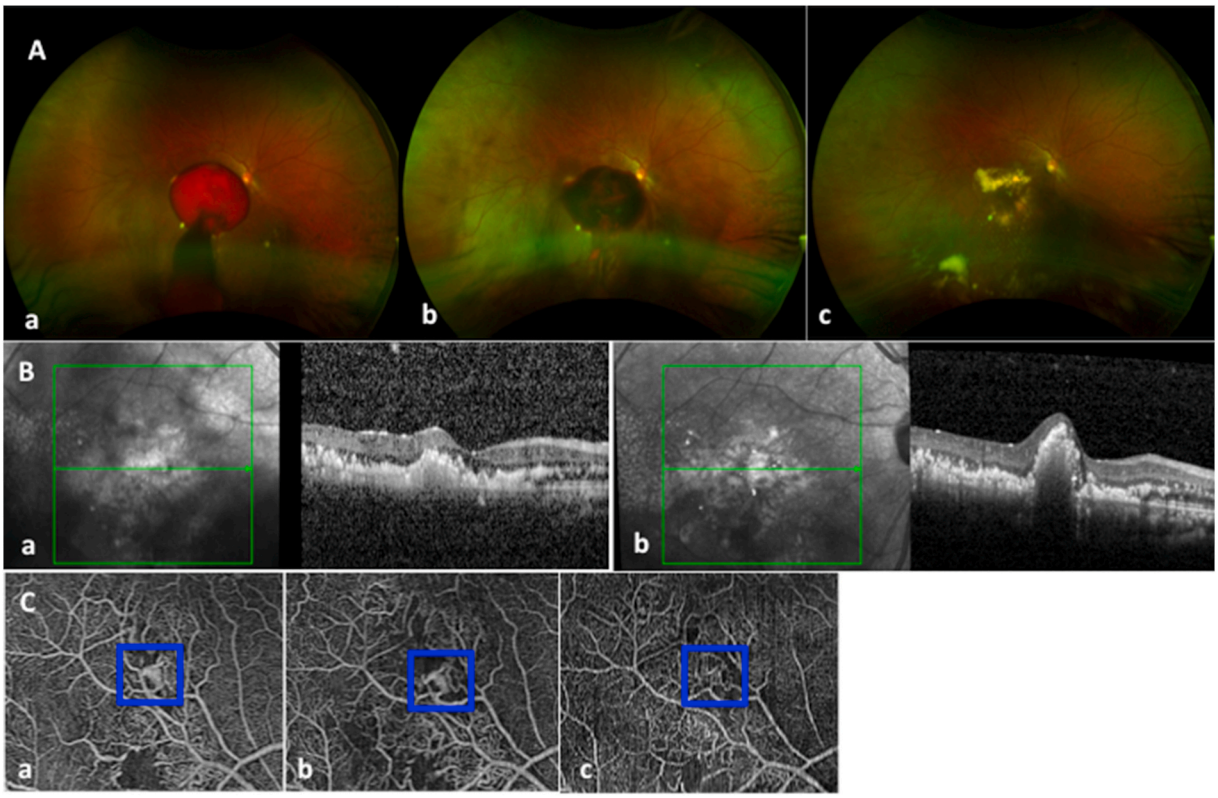

Fig. 5. Widefield color photos (Optos PLC, Dunfermline, Scotland, UK) (A) of the RE immediately after focal navigated laser photocoagulation (a) one month later (b) and after 3 months. SD OCT (B) revealed persistence of macular edema (a) and increased hard lipid exudates 4 months after last focal laser treatment (b). OCTA (C) centered in the MAR showing his regression from baseline (a) one month after first focal yellow laser photocoagulation (b) and 4 months after last treatment. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

and non-perfused areas (Fig. 2). Detailed blood analysis was performed to detect possible infective causes like syphilis, toxoplasmosis, toxocariasis, cytomegalovirus, sarcoidosis or systemic disease like diabetes, hemoglobinopathy. All the tests resulted negative. Pulmonary and cerebral magnetic resonance imaging resulted also normal. Therefore, the diagnosis of a stage 3 A 2 shields classification of CD was made.[5] Navigated Laser photocoagulation (Navilas®) of peripheral non perfused areas of the retina was performed as well as focal navigated photocoagulation of extrafovel vessels abnormalities (Fig. 3A). In addition, four monthly Aflibercept (Eylea 2 mg/0,05 ml) IVT injection were administered, contributing to successfully resolve macular edema (Fig. 3B). At five months, hard macular lipid exudates showed to be increased resulting in outer retina alterations and macular fibrosis (Fig. 3B). BCVA was 20/160 at the 7 months. Montaged 15 × 9 mm UWF-OCTA follow up showed clearly vascular anomalies after hemorrhage resorption in the SCP which guided additional laser photocoagulation (Fig. 4). Unfortunately, two months later, vision in the RE decreased due to reperfusion of a MAR and the consequent focal macular edema (Fig. 3C and D). Then additional focal navigated retinal laser was necessary. During this procedure performed with the following parameters: laser power 150 mw, pulse duration 200 ms, size of the spot 270 μm, after only three impacts the MAR rupture occurred. It was immediatly complicated by retrohyaloidal hemorrhage which was spontaneously resorbed only 3 months later (Fig. 5A). At 18 months follow up, no outpouchings of MAR were seen at the OCTA (Fig. 5C). SD-OCT showed increased foveolar lipid exudates and complete resorption of the macular edema (Fig. 5B). Final BCVA was 20/64.

3. Discussion

Presentation of CD in adult patients is rare.[6] The mean age at the time of diagnosis in adults is approximately 47 years, with a prevalence of males. It is often associated with systemic hypertension (22% of cases).[3] In contrast with the childhood onset form, the adults' one has insidious symptoms and presents a good visual acuity. Moreover, the clinical course is often benign because of the slow progression rate of the disease and the more favorable treatment outcome.[3] Rishi et al. reported decreased vision as the predominant symptom revealing CD in adults (83%).[3] Indeed our patient consulted for a significant vision impairment, consequent to diffuse macular edema. The combined approach based on IVT injections of Aflibercept and navigated focal laser treatment of abnormal dilated vassels guaranteed the resorption of the exudation at five months. However, when, because of MAR reperfusion, supplemental navigated laser treatment was required, the arterial MAR rupture occurred during the procedure.

Navigated laser photocoagulator (Navilas®) is a new laser technology that superimposing fluorescein or indocyanine green angiography, color fundus, red-free and infrared images with a computer manageable therapeutic 532 nm laser, allows controlled and targeted retinal treatment.[7] Several recent studies reported no complications associated with Navilas® laser for the treatment of diabetic macular edema correlated to microaneurysm leakage.[8–10] Navigated focal laser treatment was able to close retinal capillaries macroaneurysms with significative improvement in BCVA.[11] All these reports confirmed that focal navigated photocoagulation was safe, well tolerated with efficacy comparable to conventional laser photocoagulation.[8–11]

Even if in our patient laser treatment was performed using low power, long duration, and large spots to slowly occlude the aneurysm, may be the rupture occurred because of the great fragility characterizing these abnormal dilated vessels.[1] It cannot be concluded that the navigated focal laser directly caused the breakdown of the MAR. The fragility that characterizes the abnormal dilated vessels has already been

described. It is probably the combination of the fragility of the wall with the focal laser that caused the MAR rupture.

However, despite this important hemorrhagic complication, the navigated laser treatment contributed to resolution of macular edema and MAR occlusion as documented also by UWF-OCTA 15 × 9 mm (Fig. 5). Thanks of the UWF-OCTA it was possible to evaluate the occlusion of the MAR in a noninvasive approach.

Furthermore OCTA has been largely utilized to assess macular abnormalities in pediatric patients with CD.[12,13] To the best of our knowledge, the present report is the first to describe adulthood onset CD by UWF-OCTA. UWF-OCTA clearly showed the dilated vessels, outpouchings of aneurysms, dilated capillary network, vascular beading, capillary loops, connections between superiors and inferiors vascular arcades branches, dilated intercapillary space, faint flow and non perfused areas. Moreover, UWF-OCTA successfully oriented laser photocoagulation in our patient and was useful in the follow up. Indeed, it revealed after initial treatment and hemorrhages resorption, additional vascular abnormalities and therefore indicating complementary laser photocoagulation. It also confirmed the complete regression and occlusion of the MAR 4 months after iatrogenic rupture. Indeed it provides detailed microvasculature of both capillary plexuses while UWF-FA does not allow the evaluation of DCP.[4] Even if it does not show directly vascular wall and visualize capillary leakage, it provides, combinated to B scan OCT, indirect signs of the capillary wall permeability.[2] Because of the short duration of the exam, the reproducibility and the noninvasiveness, we believe the UWF-OCTA to be more useful than FA for the CD follow up.

4. Conclusion

UWF-OCTA is useful to visualize CD vascular abnormalities. Combined use of B scan OCT and UWF-OCTA can facilitate the diagnosis and treatment of CD. Exceptional hemorrhagic complication during navigated laser photocoagulation of CD MAR may occur with favorable outcomes.

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Conflicts of interest

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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Patient consent

The patient consented to publication of the case.

Declaration of competing interest

The authors report no competing interests pertaining to this study.

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