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Essential thrombocythemia manifesting as ophthalmic artery occlusion

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ABSTRACT

Purpose: To report the first case of a patient with essential thrombocythemia (ET) who presented with unilaterally extensive choroidal ischemia coinciding with central retinal artery occlusion (CRAO).

Observations: A 64-year-old man presented with a classic sign of short posterior ciliary artery (SPCA) occlusion, Amalric triangular choroidal infarction, coinciding with CRAO in the left eye. He was later diagnosed as having ET due to a massive platelet count of 1,100,000 cells/mm^3 and confirmed genetic testing. The magnetic resonance angiogram subsequently revealed severe occlusions of neuro-ophthalmic circulation. Interestingly, occult choriocapillaris occlusions were detected in the right eye despite being visually asymptomatic.

Conclusion and importance: Patients with ET can present with concurrent CRAO and SPCA occlusion. Identifying the causes of such events is crucial as these can precede systemic thrombocytosis or hemorrhagic complications.

1. Introduction

Essential thrombocythemia (ET) is one of the myeloproliferative neoplasms where excessive numbers of platelets result in bleeding diathesis and vascular thrombosis.[1] Although most occlusive episodes present with neurologic manifestation,[2] central retinal artery occlusion (CRAO) has been described as a rare ophthalmic complication.[3,4] We herein report the first case that presented with unilaterally extensive choroidal ischemia coinciding with CRAO.

2. Case report

A 64-year-old man was referred to our clinic because of a 4-week history of profound visual loss in the left eye (OS). Two weeks prior to the referral, he was admitted for gastrointestinal hemorrhage, and vague cherry-red spot with marked thrombocytosis (platelet count = 1,100,000 cells/mm^3) were noted.

On presentation, visual acuity was 20/63 in the right eye (OD), and hand motion OS. Both eyes (OU) showed moderate cataract, with afferent pupillary defect OS. Funduscopy revealed optic atrophy and attenuation of retinal arterioles OS. Strikingly, retinal pigment epithelium (RPE) in the territory supplied by the distal short posterior ciliary artery (SPCA) was markedly altered (Amalric triangular choroidal infarctions) (Fig. 1A). Fundus fluorescence angiography (FFA) showed enlarged foveal avascular zone and mottling hyperfluorescence, corresponding to the areas of RPE alteration OS (Fig. 1B). Simultaneous indocyanine green angiography (ICGA) revealed large wedge-shaped areas of choroidal nonperfusion (Fig. 1C). Further, macular optical coherent tomography (OCT) showed hyperreflectivity of the inner retina that, when combined with the FFA features, were compatible with recent history of CRAO (Fig. 1D). The FFA OD disclosed multiple microaneurysms in the superotemporal macula (Fig. 2A), and the ICGA showed multiple hypocyanescent spots representing macular choriocapillaris occlusions (Fig. 2B). The funduscopy and OCT images were unremarkable OD.

The platelet count reduced slightly to 896,000 cells/mm^3. Blood test for JAK2 V617F mutation, a genetic marker for myeloproliferative disorders, was positive. Tests for clotting factor deficiency and chronic myeloid leukemia were negative. He had normal blood pressure and lipid profile, and stated no history of smoking.

Considering the combined features of CRAO and SPCA occlusion, magnetic resonance angiogram (MRA) of the intracranial and carotid vessels was performed to pinpoint the occlusion site, and later identified complete occlusion of the left internal carotid artery (ICA) (Fig. 2C). Nonetheless, this patient has never developed clinical hemiplegia because of a vascular reconstitution to middle cerebral artery. A carotid endarterectomy was omitted due to the completely blocked ICA. He received oral aspirin and hydroxyurea for the treatment of ET. At 10 months, the platelet count was 193,000 cells/mm^3. Visual acuity remained unchanged with no neovascularization-related complications.

3. Discussion

Our patient experienced both forms of ET complications; complete

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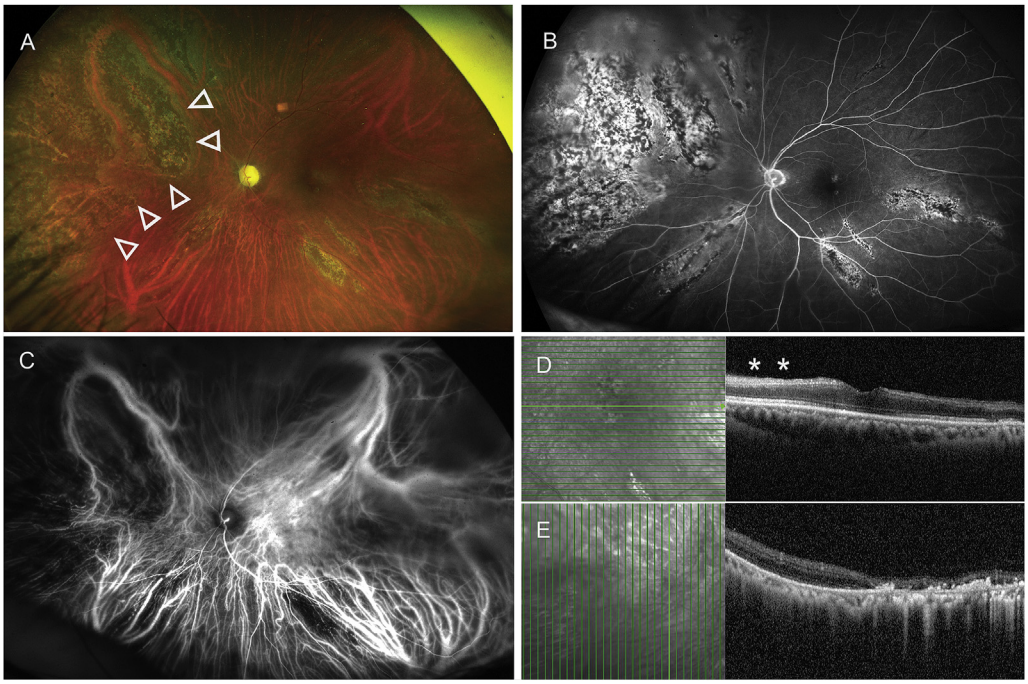

Fig. 1. Multimodal imaging of the left eye.
A: Fundus photograph showing optic atrophy, constricted retinal arteries, and prominent retinal pigment epithelium (RPE) alteration in the region supplied by distal short posterior ciliary artery (SPCA) (hollow arrowheads). These changes involve the nasal more extensively than the temporal fundus.
B: Mid-phase fluorescein angiogram revealing widening of the foveal avascular zone and mottling hyperfluorescence due to RPE window defect.
C: Simultaneous indocyanine green angiogram demonstrating large areas of wedge-shaped choroidal nonperfusion, consistent with a diagnosis of SPCA occlusion.
D: Macular optical coherence tomography (OCT) showing hyperreflective and disorganized innermost two thirds of the retina (asterisks), which reflects a consequence of central retinal artery thrombosis.
E: The vertical OCT image of the nasal side of RPE atrophy illustrating subretinal deposit of hyperreflective material and absence of RPE-photoreceptor complex. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

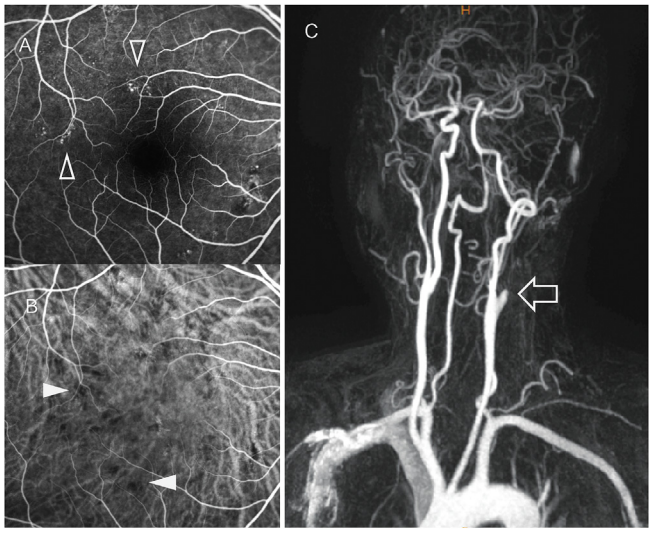

Fig. 2. Fundus angiography of the right eye, and magnetic resonance angiogram (MRA) imaging.
A: Early-phase fluorescein angiogram detecting clusters of microaneurysms in the perifoveal area (hollow arrowheads).
B: Simultaneous indocyanine green angiogram revealing multiple spots of macular choriocapillaris dropout (solid arrowheads).
C: MRA of the neck and brain showing total occlusion of the left internal carotid artery (ICA) (hollow arrow) extending out to the carotid bifurcation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

ICA obstruction brought a devastating chorioretinal ischemia, whereas the gastrointestinal bleeding was associated with defective von Willebrand factors occurring when the platelet count exceeds 1,000,000 cells/mm^3.[2] Notably, ET therapy was delayed for nearly a month after the onset of visual loss. The ICGA disclosed a classic sign of acute SPCA occlusion, the extensive triangular syndromes, while the OCT findings support the history of CRAO by showing hyperreflectivity in the innermost two thirds of the macula. Both findings are strong indicators of more severe systemic vascular insufficiency, and should prompt clinicians to explore underlying etiologies.

From a diagnostic perspective, the three main etiologies responsible for such triangular syndromes include vasospasm (hypertensive crisis), occlusion (atherosclerosis or myeloproliferative neoplasm) and inflammation (giant cell arteritis or polyarteritis nodosa).[5] Amongst the spectrum of myeloproliferative neoplasm, only ET and polycythemia vera were reportedly associated with acute ophthalmic artery occlusion.[1]

Regarding the ET treatment algorithm, the patient is classified as high risk for future thrombotic events, due to three factors: age > 60 years, history of thrombosis and positive JAK2 mutation.[6] Therefore, he requires a combination of cytoreductive and antiplatelet agents.

Interestingly, the novel finding of choriocapillaris occlusions was detected in the asymptomatic eye despite a lack of other cardiovascular risk factors. Hence, such angiographic changes may represent an early thrombocytosis of neuro-ophthalmic circulation. Identifying these microvascular disturbances could change the disease risk category and subsequent treatment.[6]

4. Conclusions

Patients with ET can present with concurrent CRAO and SPCA occlusion. Early ophthalmic evaluation may be beneficial for ET patients to prevent its devastating complications.

Patient consent

No identifying information is included, and the patient consented to publication of the case orally.

Disclosures

This work was carried out in Vajira Hospital and has not been presented at any prior meetings.

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Authorship

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

Declaration of competing interest

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