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Case Report

The "Pearl Necklace Sign" In Adult-onset Coats Disease

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Abstract

We present a case of adult-onset Coats disease to highlight the presence of the OCT "pearl necklace sign" in this rare condition. This case underscores the potential role of this specific OCT findings as negative prognostic factor, indicating persistent retinal damage and limited response to conventional therapies. Recognizing this OCT sign may help in prediction of disease progression and optimize treatment strategies.

Keywords: Adult-onset Coats disease; Pearl Necklace Sign; OCT; Macular Edema; Lipid Deposition
Abbreviation & Acronyms: OPL: Outer Plexiform Layer; CME: Chronic Cystoid Macular Edema; BCVA: Best-Corrected Visual Acuity (BCVA); OCT: Spectral-domain Optical Coherence Tomography

Introduction

The "pearl necklace sign" was firstly described by Gelman et al in 2014 [1]. It was defined as a contiguous ring of small hyperreflective dots, observed with SD-OCT, that accumulate around the inner wall of cystoid spaces placed in the Outer Plexiform Layer (OPL) and can suggest lipid-laden macrophages. It has been described in patients with exudative maculopathy and chronic Cystoid Macular Edema (CME) caused by neovascular AMD, diabetic retinopathy [2], retinal venous occlusive disease, retinal artery macroaneurysm, and Coats' disease.

In this case, we described this sign in a rare condition such as Coats' disease with adult onset, which differs from classic Coats disease because of vascular anomalies in the equatorial and peripheral areas, as well as in the juxtamacular region in the great majority. Lipid deposition, which is often vast and widespread in young people, is localised and confined in adult's form [3].

Case Report

We present the case of a 62-year-old female with adult-onset Coats disease in her right eye. Her medical history includes systemic arterial hypertension and mild type 2 diabetes, though without signs of diabetic retinopathy. Baseline Best-Corrected Visual Acuity (BCVA) in the right eye was limited to hand motion. Anterior segment biomicroscopy revealed a mild cataract, and intraocular pressure was 15 mmHg. Dilated fundus examination showed both macular and peripheral exudation, with notable vascular abnormalities, including "light bulb" aneurysms. No signs of diabetic retinopathy were detected. Wide-field color fundus photography highlighted vascular telangiectasia, retinal aneurysms, lipid exudation, macular edema, and fluorescein angiography showed regions of capillary non-perfusion without retinal neovascularization **(Figure 1A-B)**.

Spectral-domain Optical Coherence Tomography (OCT) (Spectral is HRA, Heidelberg Engineering; Heidelberg, Germany) demonstrated small hyperreflective dots arranged in a continuous ring along the inner wall of cystoid spaces in the Outer Plexiform Layer (OPL), accompanied by exudates and macular edema **(Figure 1C)**. This OCT feature, referred to the "pearl necklace sign", suggests the presence of lipid-laden macrophages. Peripheral ischemic regions were treated with argon laser photocoagulation, and the macular edema was managed with an intravitreal injection of dexamethasone (Ozurdex). At the 3-month follow-up, visual acuity remained unchanged at hand motion, though OCT showed a reduction in macular edema. However, there was an increase in lipid deposition within the intraretinal space **(Figure 1D)**. This case report was performed according to the Declaration of Helsinki and patient informed consent has been acquired.

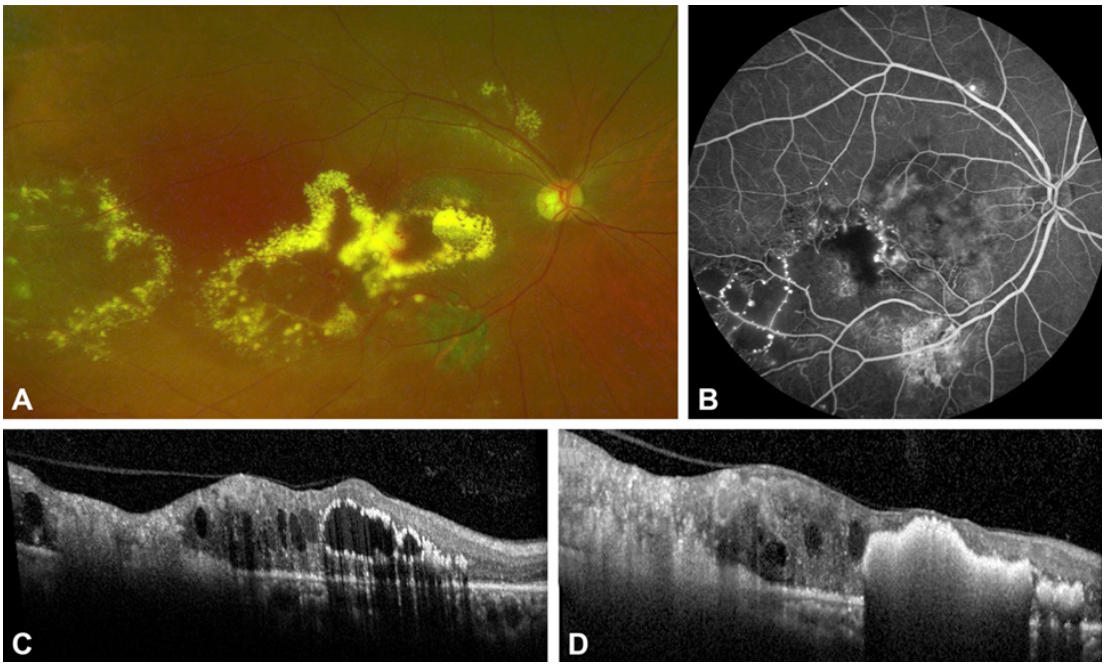

Figure 1: (A) Widefield fundus color photography at baseline shows vascular telangiectasia, lipid exudation, micro- and macroaneurysms, and macular edema. (B) Fluorescein angiography reveals capillary nonperfusion areas without retinal neovascularization, light bulb aneurysms, and retinal capillary atresia. (C) OCT illustrates the "pearl necklace sign," a contiguous ring of small hyperreflective dots surrounding the inner wall of cystoid spaces in the OPL, suggestive of lipid-laden macrophages. (D) OCT at 3 months post-intravitreal dexamethasone implant shows reduction of macular edema but increased lipid deposition in the intraretinal space.

Discussion

This case represents a particular presentation of adult-onset Coats' disease, characterized by the "pearl necklace sign," a diagnostic feature first identified by Gelman et al. in 2014 [1]. This sign, consisting of a contiguous ring of hyperreflective dots on SD-OCT surrounding cystoid spaces in the Outer Plexiform Layer (OPL), is thought to represent lipid-laden macrophages[4]. In our case, intravitreal steroid treatment led to a reduction of macular edema; however, persistent blood-retinal barrier disruption and ongoing lipid leakage from abnormal retinal vessels resulted in increased lipid deposition. Despite anatomical improvement, visual acuity remained poor, likely reflecting the chronicity of the disease and irreversible retinal damage. This underscores the continued pathological leakage and exudation, even after macular edema resolution. In conclusion he presence of the "pearl necklace sign" could be considered as a negative prognostic marker, indicating advanced retinal changes and a more refractory disease course. Patients with this sign often show reduced response to conventional treatments and experience persistent or recurrent exudation and retinal damage.

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