

Case report

An overlap of Coats and Eales diseases or a Coats variant?

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Abstract

Objective: To report an association between Coats and Eales diseases, an uncommon presentation. **Case:** A 21-year-old male presented with gradual visual impairment of two years duration in his left eye. The slit-lamp examination of the affected eye revealed +2 vitreous cells. The other findings were peri-papillary fluid accumulation and extensive macular lipid exudate deposition. Small white vessels were coursing over the macula. The major veins were dilated and tortuous and massive sheathing of both arteries and veins was forming a common sheath. In the mid-periphery and periphery of the retina, discrete hard exudates, tiny superficial retinal hemorrhages and massive vascular sheathing were present. In the inferotemporal region, two intra-retinal macrocysts were located distal to the retinal vasculature. Fluorescein angiography (FA) of the left eye highlighted numerous aneurysmal dilatations throughout the posterior pole. Fluorescein angiography also showed para-foveal telangiectasia and tiny telangiectatic vessels on the optic disk that led to late staining of the macula and optic disk. Hyperfluorescent patches of deep choroiditis were present in the early phases. There was segmental but no diffuse staining of the retinal veins which showed massive sheathing on funduscopy. In the periphery, segmental venous staining and choroidal leakage to a lesser extent were observed. In the infero-temporal quadrant, a clear-cut zone of non-perfusion and vascular abnormalities (micro-macro aneurysms, veno-venous shunts, venous beading) at the junction between the perfused and non-perfused zones were present. The findings were reminiscent of both Coats and Eales diseases. **Conclusion:** Though known as two distinct entities, both retinal pathologies may present in a single form.

Key words: Coats disease, Eales disease

Introduction

Coats and Eales diseases are remembered with the names of their denominators who described the characteristics of these entities a long time ago (Coats, 1908 and Eales, 1882). They are listed in the same differential diagnosis list as both commonly affect young males and show

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similar fundus changes. But specific hallmarks help us to differentiate between these two entities. At all ages, Coats disease is usually unilateral, the vast majority is male, and no racial or ethnic predilection has been shown. Fundamental pathology is massive exudation secondary to telangiectatic vessels which usually starts in the equatorial and peripheral retina, with the temporal quadrants most commonly involved. On the other hand Eales disease affects

young males with bilateral involvement, which may show subtle changes on one eye at the earlier stages. Basically, what makes it different from Coats disease is the presence of inflammation and the related hallmark findings of phlebitis, peripheral non-perfusion and retinal neovascularization. As the disease progresses, both may show similar vascular changes (micro or macro-aneurysms, new vessels on the disk or elsewhere on the retina, vascular sheathing), fibrotic tissue proliferation and similar further stage complications (retinal detachment, glaucoma, cataract, uveitis). Though attempts to classify Coats have been made by some authors, there is no universally accepted standard classification system for Coats disease at present. Cahill et al have recently offered a new staging system in which stage 4 is designed for the presence of focal telangiectatic vessels associated with clinical evidence of other eye disease or features suggesting a systemic disease (Cahill et al, 2001). Soon after Henry Eales coined the term, some atypical Eales cases were reported (Elliot, 1954). Eliot AJ reported two atypical cases in which there was no retinal or vitreous hemorrhage, but exudation and venous abnormalities were prominent. We herein report such an atypical case that presented with clinical features reminiscent of both Coats and Eales diseases.

Case report

A 21-year-old Caucasian male presented with decreased vision and a history of floaters in the left eye. His complaints were present for two years and deteriorated gradually over time. His medical history was unrevealing for any systemic or ocular disease. On examination, visual acuities were 20/20 and 20/200 in the right and left eyes respectively. The anterior and posterior segment evaluation of the right eye was normal. The slit-lamp examination of the left eye revealed +2 vitreous cells. On funduscopy of the left eye, peri-papillary fluid accumulation and extensive macular lipid exudate deposition were observed. Small white vessels were

coursing over the macula. The major veins were dilated and tortuous, massive sheathing of both arteries and veins was forming a common sheath. In the mid-periphery and periphery of the retina, discrete hard exudates, tiny superficial retinal hemorrhages and massive vascular sheathing were present. In the infero-temporal region two intra-retinal macrocysts were located distal to the retinal vasculature. Fluorescein angiography (FA) of the left eye highlighted numerous aneurysmal dilatations throughout the posterior pole. Fluorescein angiography also showed parafoveal telangiectasia and tiny telangiectatic vessels on the optic disk that led to late staining of the macula and optic disk. Hyperfluorescent patches of deep choroiditis were present in the early phases. There was segmental but no diffuse staining of retinal veins which showed massive sheathing on funduscopy. In the periphery, segmental venous staining and choroidal leakage to a lesser extent were observed. In the infero-temporal quadrant, a clear-cut zone of non-perfusion and vascular abnormalities (micro-macro aneurysms, veno-venous shunts, venous beading) at the junction between the perfused and non-perfused zones were present. Optical coherence tomography (OCT) assessment of the left eye revealed increased central macular thickness (588 μm) with sub-foveal and intra-retinal cystic fluid accumulation. Flash electroretinography (f-ERG) showed a reduction in the amplitudes in the affected eye. Multi-focal ERG (mf-ERG) responses were both suppressed and impaired in the affected eye relative to the healthy eye (Figure 1). Systemic work-up was done to rule out other possible etiologies, such as diabetes and hypertension (normal glucose level and normal diurnal blood pressure), hematologic malignancies such as leukemia or lymphoma (normal complete blood cell and platelet counts), proliferative hemoglobinopathy (normal electrophoresis findings), inflammatory/uveitic conditions including sarcoidosis (normal erythrocyte sedimentation rate, normal angiotensin converting enzyme level, normal chest radiography results, and normal C-reactive

protein level; undetectable levels of cytoplasmic anti-neutrophil, anti-cardiolipin, and antinuclear antibodies), infections such as tuberculosis and syphilis (normal tuberculin skin test result, normal chest radiography, and non-reactive TPHA test), and some causes of hypercoagulable states (normal prothrombin and partial thromboplastin times). Except for the finding of hypochromic anemia [Hb = 8.3 g/dl (normal: 13.6 - 17.2), Htc = 27.8 % (normal: 39.5 - 50.3), MCV = 63.4 fl (normal: 80.7 - 95.5)], the results of all routine laboratory tests were normal. Investigation to identify the etiology of the anemia was unrevealing.

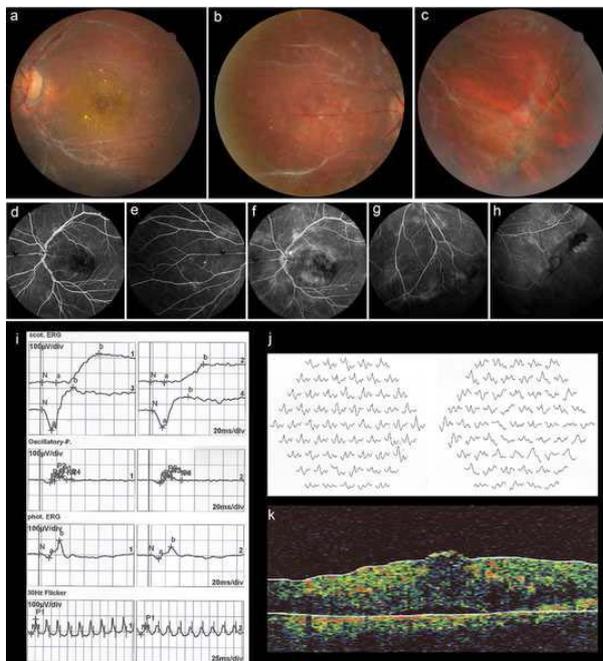


Figure 1: Color fundus photographs showing prominent sheathing of major vasculature, massive exudation in the macula, micro and macroaneurysms in the posterior pole (a), discrete hard exudates and massive white vascular sheathing of affected vessels in the midperiphery (b), retinal macrocysts located in the periphery (c); Fluorescein angiography images depicting multiple micro/macroaneurysms, early leakage from optic nerve head and choroid in the posterior pole (d), segmental vascular leakage in the mid-periphery (e), extensive leakage involving macula, optic

disk and deep choroid in the late phase (f), ongoing choroidal leakage and microaneurysms (g), vascular abnormalities and non-perfused retina in the periphery (h); Flash electroretinography amplitudes were depressed in the affected eye (i); Multifocal electroretinography showed diffuse retinal involvement in the affected eye (j); Optical coherence tomography showed sub-retinal fluid and intra-retinal cystoid spaces.

Discussion

In 1908, George Coats described the typical manifestation of the disease - a juvenile male approximately 16 years of age presents with massive exudation especially in the temporal quadrant in one eye and without associated systemic illness (Coats, 1908). But later some authors reported cases with retinal telangiectasia and exudation of varying severity in adults and children with or without associated disease (Khan et al, 1988 and Tolmie et al, 1988). This led to a terminologic braching, and the classical disease was called "Coats disease" and the atypical forms were called "Coats reaction", "Coats syndrome" or "Coats-type disease". But long before, in 1912, Coats had already presented atypical cases and recently Cahill et al proposed a staging system that encompasses all the forms of the disease (Cahill et al, 2001). Some previously reported associations are those with retinitis pigmentosa, central nervous system disorders and dyskeratosis (Khan et al, 1988 and Tolmie et al, 1988).

Though in both Coats and Eales diseases the most common presenting complaint is reduced vision, the reduced vision is mainly due to macular involvement (exudation or cystoid edema) in Coats disease while it is recurrent vitreous hemorrhage in Eales disease (Lee et al, 1991 and Das et al, 2010). The history of floaters in our case raised our suspicion about recurrent vitreous hemorrhage but we did not detect any clear sign of this. Still this observation does not readily exclude Eales since the decline in visual acuity may be due to retinal vasculitis but not as a result of vitreous hemorrhage. On the other

hand, as seen on FA, our case lacked neo-vascularization, which could be the reason for the absence of vitreous hemorrhage. Thus, decreased vision in the current case was probably due to massive exudation and cystoid edema of the macula secondary to telangiectasia. Though some fundoscopic findings such as macular telangiectasia and massive exudation were consistent with Coats disease, the massive vascular sheathing, the deep choroiditis and the clear-cut zones of non-perfusion were not. Cases with Coats disease may show vascular sheathing by yellow cholesterol deposits, but this kind of white glial sheathing with no leakage on FA indicates Eales (Das et al, 2010). Also, there were some findings that both entities have in common such as irregularities in vessel caliber, focal telangiectasia and aneurismal dilatations. However, vascular changes adjacent to the perfused-nonperfused retina border was typical of Eales disease. Retinal macrocysts associated with Coats disease are mostly seen in the exudative detachment phase, but we observed smaller ones on the edge of the non-perfused retina (Chang et al 1984). Depressed rod and cone responses on the f-ERG were probably the result of impaired choroidal circulation due to deep choroidal inflammation, and impaired mf-ERG was the reflection of the outer retinal involvement.

Frosted branch angiitis is the other rare entity that must be excluded. Though it has a racial predilection for the Japanese, some cases were also reported from outside Japan (Atmaca et al, 1993). It also predominantly affects the young and fit. The reported cases were mostly female and bilaterality is almost always constant. Widespread retinal vasculitis with typical florid translucent perivascular exudate may resemble that in Eales, but diffuse late leakage from the affected large vessels differentiates it from Eales.

Conclusion

Coats disease may show different associations with other eye or systemic diseases. This case

report presents a new association between Coats and Eales diseases that has never been reported before.

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