Case report

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

Murat Kucukevcilioglu,¹ Gungor Sobaci,¹ Ali Hakan Durukan,¹ Fazil Cuneyt Erdurman², Osman Melih Ceylan³
¹Gulhane Military Medical School, Department of Ophthalmology, Ankara, Turkey
²Canakkale Military Hospital, Canakkale, Turkey
³Ardahan Military Hospital, Ardahan, Turkey

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 fundoscopy. 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 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 fundoscopy of the left eye, peri-papillary fluid accumulation and extensive macular lipid exude 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 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 retinal veins which showed massive sheathing on fundoscopy. 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 hyper-coagulable 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.

**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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