

Adult Coats' disease: a case report

Nor Sharina Yusof, Zunaina Embong, Mokhtar Ibrahim

Department of Ophthalmology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian 16150, Kelantan, Malaysia

Correspondence to: Zunaina Embong. Department of Ophthalmology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian 16150, Kelantan, Malaysia. zunaina@kb.usm.my

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Abstract

• **AIM:** To present a case of Coats' disease in adult and to illustrate the retinal findings.

• **METHODS:** A case report.

• **RESULTS:** A 63 years old Malay man, a known case of diabetes mellitus, presented with one year history of painless and progressive reduced vision in the right eye. Right eye visual acuity was counting finger 2 feet while left eye best corrected visual acuity was 6/12. Right fundus showed presence of extensive subretinal exudates at the temporal part of the posterior pole involving the macular area and nasal to the optic disc. It was associated with multiple areas of abnormal blood vessels temporal to the macula with attenuated vessels. Fundus fluorescein angiography (FFA) showed presence of telangiectatic vessels at the area of subretinal exudates and leakage of fluorescein from the telangiectatic vessels. The left eye was normal. Based on clinical and FFA findings, he was diagnosed to have adult Coats' disease. The right eye was treated with argon laser photocoagulation. Six months post laser, his visual acuity remained at 6/120 and there was minimal regression of the disease seen in the retina.

• **CONCLUSION:** The clinical and angiographic findings are important in diagnosing retinal telangiectasia. Treatment by laser photocoagulation in area of leakages may be beneficial in preventing visual loss.

• **KEYWORDS:** adult Coats' disease; telangiectasia; sub-retinal exudates

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INTRODUCTION

Retinal telangiectasia is a group of rare, idiopathic, congenital retinal vascular anomalies affecting the retinal capillaries. These conditions are characterized by dilatation and tortuosity of retinal blood vessels, formation of multiple aneurysms, leakage of vessels associated with exudation of lipid at the retina periphery and the macula^[1]. There are three types of retinal telangiectasia which have been

described; Idiopathic Juxtafoveal Telangiectasia, Leber miliary aneurysms and Coats' disease^[1]. Since this disease is rare and often difficult to diagnose, a detailed study of the retinal findings is of great importance. Herein, we report a case of adult Coats' disease to illustrate its presentation and the retinal findings.

CASE REPORT

A 63 years old Malay man, presented with a history of painless and progressive reduced vision in the right eye for one year. It was not associated with eye redness, floaters, flashes of light or metamorphosia. There was no history of ocular trauma. He is a known case of Diabetes Mellitus for one year and there was no other medical illness.

Ocular examination revealed visual acuity of the right eye was counting finger 2 feet. Visual acuity in the left eye was 6/15 and improved to 6/12 with pin hole. There was no relative afferent pupillary defect. Anterior segment examinations and intraocular pressure in both eyes were normal.

Fundus examination of the right eye showed presence of extensive subretinal exudates at the temporal part of the posterior pole involving the macular area (Figure 1). There was also presence of extensive subretinal exudates nasal to the optic disc (Figure 2). The retinal vessels appeared attenuated and presence of multiple areas of abnormal blood vessels temporal to the macula (Figure 1). The peripheral retina showed area of retinal scarring with fibrosis (Figure 3). The optic disc was pink with 0.3 cup disc ratio and presence of peripapillary atrophy temporally. There was no retinal hemorrhage or retinal detachment. Vitreous was normal with no sign of vitritis. The left fundus was normal with no features of diabetic retinopathy. Systemic examination was unremarkable.

Fundus fluorescein angiography (FFA) showed presence of telangiectatic vessels (capillary dilatation with localized hyperfluorescence) at the area of subretinal exudates (Figure 4, 5) during the early phase. Late phase of FFA showed leakage of fluorescein from telangiectatic vessels. Macular appeared normal and was not edematous. Infective screening and connective tissue disorder were investigated and showed normal results.

Based on clinical and fundus angiogram findings, he was diagnosed to have idiopathic adult retinal telangiectasia or adult Coats' disease. The right eye was treated with argon laser photocoagulation at the area of telangiectatic vessels. Six months post laser, his visual acuity remained at 6/120 and no improvement with pin hole. There was minimal regression of the subretinal exudates at the nasal area with no new telangiectatic vessels seen during the last follow-up (Figure 6).

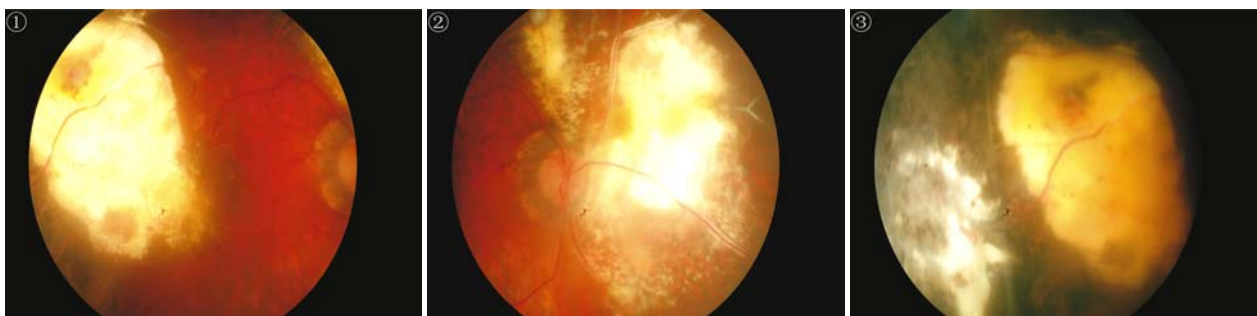


Figure 1 Right fundus photograph showing extensive subretinal exudates at temporal part of the posterior pole involving the macula with abnormal blood vessels.

Figure 2 Right fundus photograph showing extensive subretinal exudates nasal to the optic disc.

Figure 3 Right fundus photograph showing retinal fibrosis and scarring at peripheral retina temporally.

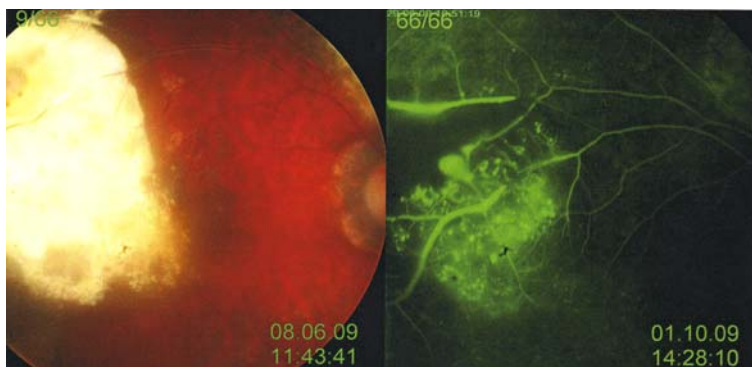


Figure 4 Fundus fluorescein angiogram of the right eye showed telangiectatic vessels at temporal to the fovea.

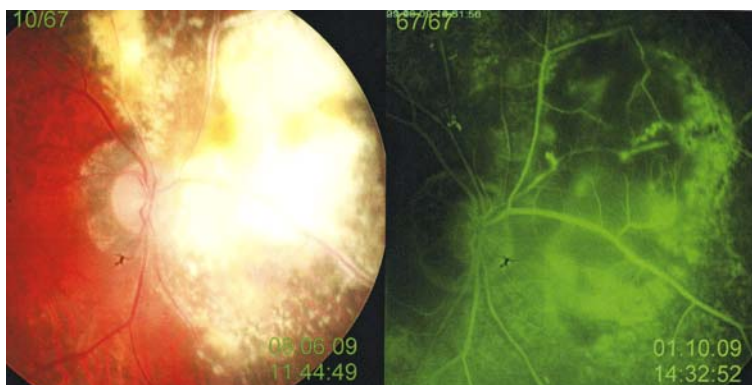


Figure 5 Fundus fluorescein angiogram of the right eye showed telangiectatic vessels nasal to the disc.

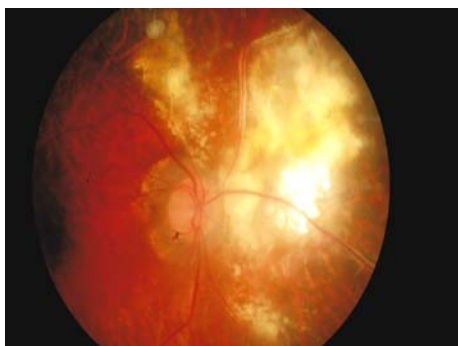


Figure 6 Six months post laser, right fundus showing minimal regression of subretinal exudates at the nasal area.

DISCUSSION

Retinal telangiectasia was a term proposed by Reese to describe retinopathies of dilated and incompetent vessels [1]. This type of vascular alterations differs from telangiectasia secondary to venous thrombosis, vasculitis, diabetes, carotid

occlusive disease and radiation retinopathy. It affects healthy adults whose visual acuity in one or both eyes decreases due to exudation, nutritional or hypoxic retinal changes [2].

The pathogenesis of retinal telangiectasia is unknown. Study done by Tripathy and Ashton suggested that initial defect that caused the disorder are due to functional or defect in the structural of blood retina barrier (vascular endothelial) with the consequent mural disorganization and formation of aneurysms or telangiectasis [3]. A defect at the vascular endothelial would be compensated for over a long period until the secondary anatomic alteration become sufficiently to be clinically manifest in adulthood [3].

Idiopathic adult retinal telangiectasia with exudation is a spectrum of disease which synonymous with Coats' disease. Coats' disease diagnosed in adulthood presents unilaterally, primarily in men with vascular telangiectasis, lipid exudation, microaneurysms and macroaneurysms, macular edema, areas

of capillary non-perfusion with adjacent webs of filigree-like capillaries and absence of retinal neovascularization^[4]. Adult Coats' disease patients often asymptomatic or present with good vision and did not have leukocoria, extensive areas of exudation or retinal detachment. In typical Coats' disease, vascular abnormalities are often in the far periphery, between the equator and the ora serrata^[5]. In this patient, he presented with extensive subretinal exudation at the temporal part of the posterior pole and the area nasal to the disc.

Fluorescein angiography can help in define the structural and permeability alteration in the affected vessels and demonstrate the extent of the extravascular leakage of serous exudates into and beneath the retina^[6]. In Coats' disease, the fundus angiography showed characteristic premature hyperfluorescence of telangiectasias and presence of microaneurysms with leakage at late phase^[7]. In previous angiographic studies of less advanced Coats' disease demonstrated that the arterial system seemed to be more damaged than the venous side. Most of the affected arteries ended in macroaneurysm-like dilatations surrounded by avascular areas or complete vascular closure in more advanced cases^[8]. In this patient, early phase of angiography showed dilatation of capillaries with hyperfluorescence localized at the exudates area. There was leakage of fluorescence from the telangiectatic vessels noted during the late phase. This findings demonstrate the typical retinal telangiectasia which correspond to the features of adult Coats' disease.

Laser photocoagulation may be taken into consideration at the beginning if telangiectasias are located in the periphery and presence of large exudation areas^[5]. Cryotherapy is generally reserved for more advanced cases and those patients with lesions confined to the far periphery^[9]. The purpose of these treatment modalities is to minimize vascular leakage and promote re-absorption of exudates and edema^[9]. If the exudation area is limited to a single quadrant or is located in the nasal sector, visual prognosis is more favourable^[7]. In this patient, he was treated with laser photocoagulation and after six months laser treatment showed some minimal regression of the subretinal exudates at the nasal area.

CONCLUSION

Adult Coats' disease is an uncommon cause of vision loss beginning in adulthood. It usually affects the visual acuity gradually and later may progress and become symptomatic later in life as a result of hemorrhage, edema or lipid exudation. The clinical and angiographic findings are important in diagnosing the retinal telangiectasias. Treatment

by laser photocoagulation into areas of leakage may be beneficial in preventing visual loss.

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成人 Coats 病 1 例

Nor-Sharina Yusof, Zunaina Embong, Mokhtar Ibrahim

(作者单位:马来西亚吉兰丹,马来西亚理科大学医学院眼科)

通讯作者:Zunaina Embong. zunaina@kb.usm.my

摘要

目的:报告成人 Coats 病 1 例,并探讨其引起的视网膜病变。

方法:病例报告。

结果:马来西亚 63 岁糖尿病患者 1 例,男,右眼无痛性视力下降 1a,视力指数/2 英尺,而左眼最佳矫正视力为 6/12。右眼眼底表明颞侧后极部出现广泛的视网膜渗出物,可累及黄斑区和鼻侧视盘。异常分布的多区域血管位于黄斑颞侧且呈衰减趋势。眼底荧光血管造影显示,毛细血管扩张在视网膜渗出区域且有荧光素渗漏现象存在。左眼表现正常。根据临床和 FFA 结果,患者被诊断为成人 Coats 病,右眼行氩激光光凝治疗。激光后 6mo,患者的视力保持在 6/120 并出现视网膜最小限度的回退。

结论:临床和血管造影结果是视网膜毛细血管扩张症的重要诊断依据。渗漏区域激光光凝治疗有利于防止视力丧失。

关键词:成人 Coats 病;毛细血管扩张;视网膜下渗出