Case Report

Coats disease – An atypical presentation

Ankita A Patil[1*], Anjali D Nicholson[2], Anamika H Agrawal[3], Vidhya D Khobragade[4]

[1,4]Speciality Medical Officer, Department of Ophthalmology, GSMC & KEM Hospital, Mumbai, Maharashtra, INDIA.
[2]Professor and HOD, [3]Additional Professor, Department of Ophthalmology, TNMC & Nair Hospital, Mumbai, Maharashtra, INDIA.
Email: ankeeta_p@yahoo.com

Abstract

We present a 17 year old female with gradual onset diminished vision in left eye since 1 year. On examination she revealed telangiectasia and exudation with exudative subtotal retinal detachment in left eye superotemporally. USG B-scan showed dense echoes, choroidal thickening and exudative retinal detachment temporally. There was no evidence of calcification or mass lesion. On retinal fluorescein angiography, areas of capillary non-perfusion and dilatation together with fusiform aneurysms were observed. Thus we highlight an atypical presentation of coats disease; the accurate diagnosis and treatment of which would enable timely management and favorable visual prognosis for the patient.

Keywords: Exudative retinal detachment, coats disease, female

[*]**Address for Correspondence:**
Dr. Ankita A Patil, Speciality Medical Officer, Department of Ophthalmology, GSMC & KEM Hospital, Mumbai, Maharashtra. INDIA.
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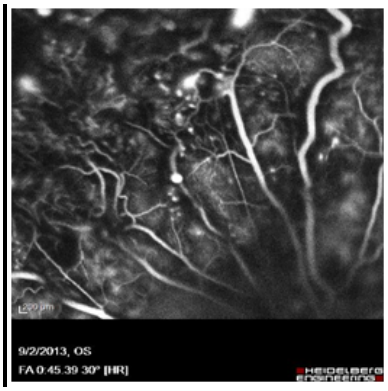
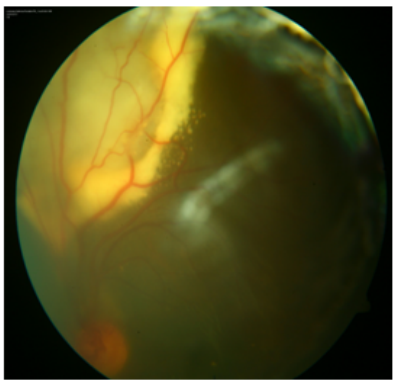
CASE REPORT

A 17 year old female came with the history of gradual onset diminished vision in her left eye since 1 year. No relevant significant family history or medical history was present. No history of trauma. Birth history was not significant. The bcva in right eye was 6/6 and left eye was 6/18.External examination was within normal limits. On fundus evaluation with Indirect Ophthalmoscope; right eye was within normal limits however the left eye showed telangiectasia and exudation with exudative subtotal retinal detachment in left eye superotemporally without any vitreous traction. Thus a clinical diagnosis of Atypical coats disease was made. The diagnosis was confirmed by USG B-Scan which showed dense echoes, choroidal thickening and exudative retinal detachment temporally: there was no evidence of any retinal mass lesion or calcification and retinal fluorescein angiography which showed areas of capillary non-perfusion and dilatation together with fusiform aneurysms. A surgery of Subretinal Fluid drainage with external band buckle with cryotherapy was planned for the patient.





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DISCUSSION

Coats disease is an idiopathic non hereditary condition characterised by retinal telengectasia with subretinal and intraretinal exudation and exudative retinal detachment without vitreous traction.[1] Coats disease usually occurs in boys with peak incidence between 6-8 years of age[2] The disease is usually progressive.[3] Primary abnormalities in Coats' disease are confined to retinal vessels and abnormalities can occur in all levels of the vasculature which include: micro- and macroaneurysms, capillary dilation (telangiectasis) and nonperfusion, and saccular outpouchings of retinal venules[4,5]

In mild cases, one or more localized foci of retinal telangiectasia are noted within the retinal capillary bed, typically in the temporal quadrants between the equator and oraserrata. As the disease progresses there is associated massive intraretinal and subretinal exudation leading to thickening of retina and exudative retinal detachment. The advanced stages of the disease include total retinal detachment, leucocoria, strabismus and painful glaucoma secondary to angle closure.[6] Birth history and family history help distinguishing coats disease from ROP, retinoblastoma and other causes of exudative retinal detachment. External examination is generally within normal limits. Fundus evaluation shows typical retinal telangiectasia and capillary aneurysms. On retinal fluorescein angiography, areas of capillary non-perfusion and dilatation together with fusiform aneurysms are observed[7].Ultrasonography and ct-scan are used to distinguish from retinoblastoma by ruling out the presence of any calcification or mass lesion.[8,9] To aid in the management Shields and co-authors classified Coats' disease as: stage 1, telangiectasia only; stage 2, telangiectasia and exudation (2A, extrafoveal; 2B foveal); stage 3, exudative retinal detachment (3A, subtotal; 3B total); stage 4, total detachment with secondary glaucoma; and stage 5, advanced end-stage disease. The management based on their staging scheme was as follows: Stage 1 disease, telangiectasia only, receives either periodic observation or laser photocoagulation. Stage 2, telangiectasia and exudation, is treated with cryopexy or laser photocoagulation. Stage 3A, subtotal detachment, requires cryopexy or laser photocoagulation; however, 3B, total detachment, receives cryopexy for shallow detachments and either scleral buckling or pars plana vitrectomy for more extensive involvement. Stage 4, total retinal detachment and glaucoma, usually requires enucleation to relieve severe pain. However, stage 5, advanced end-stage disease, requires no treatment because the eye is usually blind but comfortable.[10] Thus our case highlights an atypical presentation of coats disease in a young female supported by typical features of coats disease such as retinal telengectasia, exudation and exudative retinal detachment.

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