



A case of Coats Disease presenting with Retinal Detachment in Bhutan

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ABSTRACT

Coats disease is a rare retinal disease. It is characterised by abnormal dilatation of retinal capillaries (telangiectasia) and increased vascular permeability (intraretinal and subretinal exudation). In advanced stage of disease the patient can have exudative retinal detachment where the retina detaches from its normal attachment. In some cases there will be secondary increase in intraocular pressure if not treated on time. It has a unilateral presentation in most of the cases. This article describes about a four year old boy who presented with retinal detachment due to coats disease. The patient was managed effectively with external subretinal fluid drainage along with laser and cryotherapy. It is essential to rule out aggressive eye cancer (retinoblastoma) as both can have similar presentation.

Keywords: Coats disease; Cryotherapy; Exudation; Laser; Telangiectasia.

INTRODUCTION

Coats disease is a rare condition. The incidence of coats disease is 0.09 per 100,000 population¹. The hallmark of coats disease is unilateral presentation (95%) in young males (76%)². Shields et al defined coats disease as idiopathic retinal vascular telangiectasia with intraretinal or subretinal exudation and without vitreoretinal traction³. Coats disease presents with decreased vision and strabismus in later stage of disease. The signs are leukocoria, telangiectatic vessels in retina, retinal hemorrhages, exudation and retinal detachment. If not treated on time it can lead to secondary glaucoma and blindness. The staging and stage wise management was first described in 2001 based on their experience with 150 cases³. Indirect ophthalmoscopy is the gold standard to diagnose coats disease. If not treated early it can progress to exudative retinal detachment and neovascular glaucoma with painful blind eye. Enucleation is the treatment of choice for painful blind eye^{3,4}. However, such destructive procedures can be prevented with different treatment armamentarium like laser, cryotherapy, intravitreal steroids and surgery³⁻⁶.

We present a coats disease in a young boy with a stage 3 disease. He was treated effectively with laser, cryotherapy and surgery. It is not reported in our country till now. It is essential to rule out retinoblastoma before initiating any treatment as both can have similar presentation.

CASE REPORT

A four year old boy presented with whitish mass in the black part of the right eye for one month duration. The history was given by his father during their visit to National Eye Center in the month of July, 2022. On examination, the visual acuity in right eye was hand movement and left eye was 6/6. The intraocular pressure was 8 and 12 mm of Hg in right eye and left eye respectively. The anterior segment examination was normal in both eyes. The fundus examination was normal in left eye.

The right eye had total retinal detachment and retina was almost at the level of posterior surface of lens. The vessels were tortuous and telangiectasia were noted. An ultrasound B scan was not done as fundus was clinically visible. A computed tomography scan of orbit was done to rule out retinoblastoma. However, the report was not consistent with retinoblastoma. So the patient was diagnosed as stage 3 coats disease. Stage 3 coats disease refers to telangiectasia with exudative total retinal detachment.

The patient's right eye (RE) underwent external subretinal fluid (SRF) drainage along with cryotherapy and laser therapy under general anaesthesia. He underwent trans scleral external SRF drainage with 26 G needle with infusion from anterior chamber maintainer. The retina settled immediately during intraoperative procedure. The SRF was sent for cytology to rule out any ocular malignancy. The cytology report did not show any evidence of malignancy.

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The patient was followed up at day one and day seven. During first followup the visual acuity in RE was 5/60. The cornea, anterior chamber depth and lens were unremarkable. Retina was attached though there was few pockets of subretinal fluid at posterior pole. On follow up one week, visual acuity in

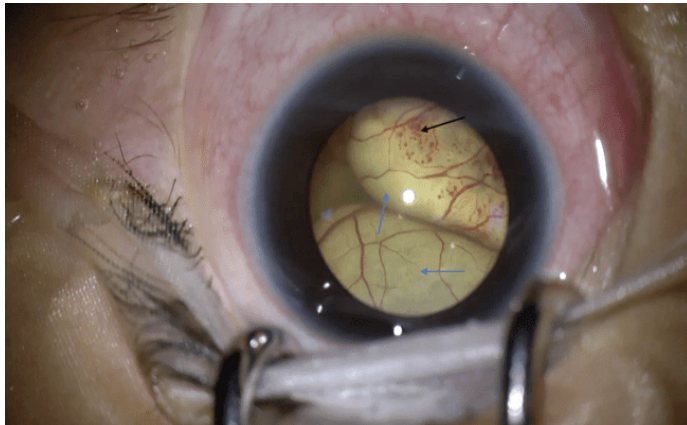


Figure 1. Fundus picture showing telangiectatic vessels (black arrow) and exudative retinal detachment (blue arrow)

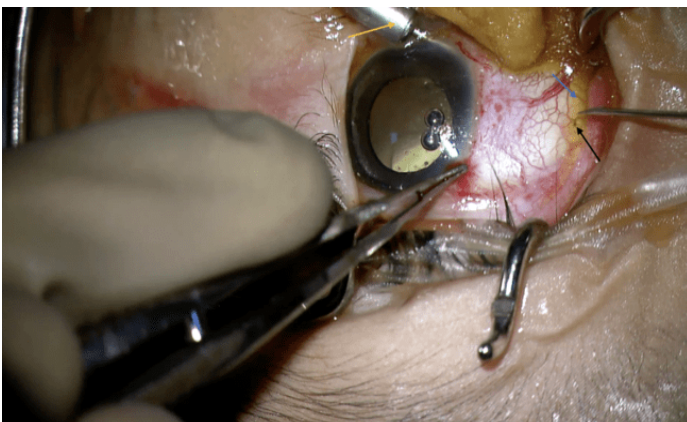


Figure 2. External trans scleral drainage of subretinal fluid. Infusion via anterior chamber maintainer (yellow arrow). Sclerotomy made with 26 G needle (black arrow). Drainage of yellowish subretinal fluid (blue arrow). (Surgery done by Dr.Phuntsho and Dr.Sushma)

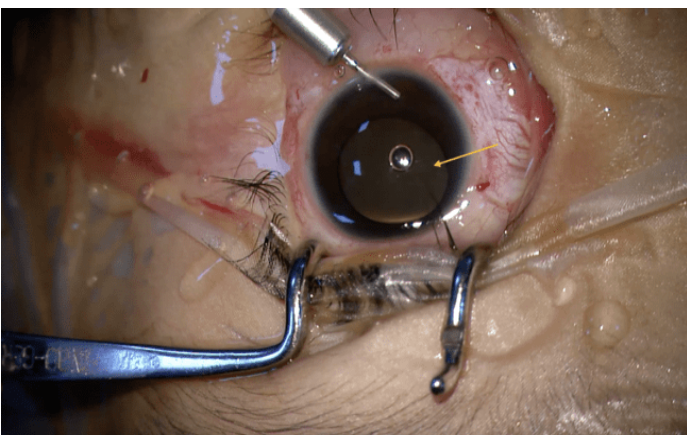


Figure 3. Retina settled intraoperatively (yellow arrow)

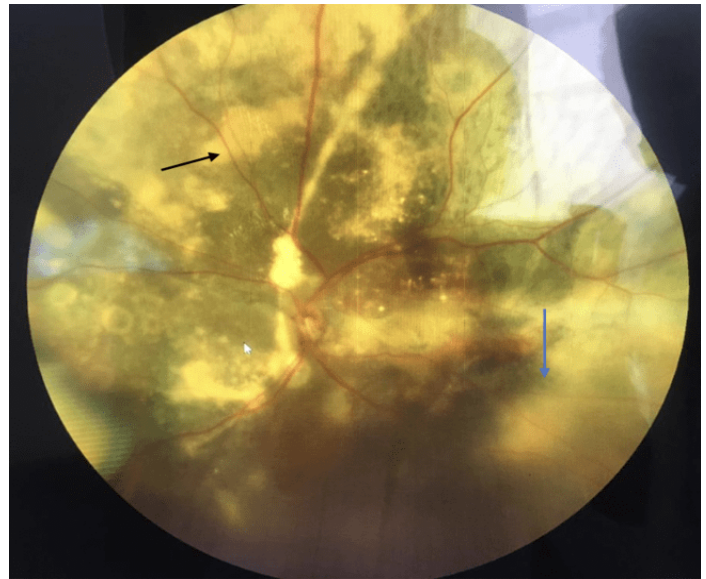


Figure 4. Fundus photo at 1 week followup. Retina attached with extensive exudates (black arrow). Inferior shallow subretinal fluid (blue arrow)

RE improved to 6/36 and retina was attached. The patient was asked to follow up after three months but patient did not turn up for followup.

DISCUSSION

Coats disease predominantly affects male population and it has unilateral presentation in most of the cases¹. It is staged into five categories as stage 1 (Retinal telangiectasia only), stage 2 (telangiectasia with exudation), stage 3 (Exudative retinal detachment), stage 4 (stage 3 with glaucoma) and stage 5 (Advanced end stage disease)^{2,3}. The treatment is indicated only in foveal exudation, exudative retinal detachment, leaking vessels and neovascular glaucoma^{3,6}.

In a study by P Rishi et al, retinal attachment after external subretinal fluid drainage along with cryotherapy was achieved in 57%⁶. In our case retina was completely attached at one week follow up but it needs long term follow up to completely cure the disease. In some studies selective laser photocoagulation over the telangiectatic vessels was effective in resolution of exudative retinal detachment. By ablating the abnormal vessels in the periphery, exudation was completely resolved by 4 weeks to 12 months⁷.

In our case the procedure involved external subretinal fluid drainage with fluid infusion via anterior chamber maintainer along with laser and cryotherapy. It is a simple technique except that the procedure has to be done under general anaesthesia. In a article by Desai SR, coats disease with bullous exudative retinal detachment was effectively managed with external subretinal fluid drainage along with cryotherapy⁸. It was a similar technique that we have managed in our case except that we did not use injection bevacizumab and active aspiration of subretinal fluid

with cutter. Similarly external drainage of SRF alone versus SRF drainage combined with pars plana vitrectomy proved more effective in the latter option of treatment⁹. However in our case external SRF drainage alone was effective in settling the retina. It is essential to differentiate between retinoblastoma and coats disease. Retinoblastoma will present as whitish fluffy mass in the retina either as endophytic or exophytic growth. The ultrasound B scan and CT scan will show hyperreflectivity due to high calcium content of the mass. The subretinal fluid analysis will show malignant cells in cytology. In histopathology, findings like fleurettes, Flexner -wintersteiner rosettes and Homer wright rosettes point towards retinoblastoma. In coats disease retinal telangiectasia and extensive exudation are hallmark of the disease. High calcium content and malignant cells are not seen in coats disease.

It was never reported in our country till date. This case report can serve as baseline reference for future research too.

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