

A Case of Coats Disease with Bullous Retinal Detachment Treated Successfully by One Selective Ophthalmic Arterial Injection of Melphalan

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Financial Interest

None

Purpose

Here, we present successful eye-preservation in a case with advanced Coats disease with bullous retinal detachment using one selective ophthalmic arterial injection of melphalan using a balloon catheter.

Background

Coats disease is an idiopathic condition characterized by telangiectatic and aneurysmal vessels with intraretinal and subretinal exudation and fluid⁽¹⁾. It is a pseudoretinoblastoma and is one of the most difficult conditions to differentiate from retinoblastomas. Treatments of its advanced stages require surgical interventions. However, their outcome is not always favorable, and enucleation is often inevitable.

We experienced one case of Coats disease with bullous retinal detachment. Its diagnosis was difficult, and the patient was referred to the first author for eye-preservation therapy of retinoblastoma. We performed a therapeutic diagnosis using selective ophthalmic arterial injection of melphalan (SOAI)⁽²⁾ because of the safety and efficacy of this treatment⁽³⁾.

Patient & Clinical Findings

Patient: A 20-month-old boy.

C.C.: Leukokoria (OD)

History of present illness:

March 2012

The parent found leukokoria in the patient's right eye. This was diagnosed as unilateral advanced retinoblastoma with total retinal detachment at Miyazaki University Hospital.

April 2013

He was referred to the first author at Teikyo University Hospital for eye-preservation therapy of retinoblastoma.

Clinical findings

Visual acuity:

OD = light perception.

I.O.P: OD = 12 mmHg, OS = 15 mmHg.

Cornea, anterior chamber, iris, lens:

OU: no abnormality.

Vitreous:

OD: bullous retinal detachment.

OS: no abnormality.

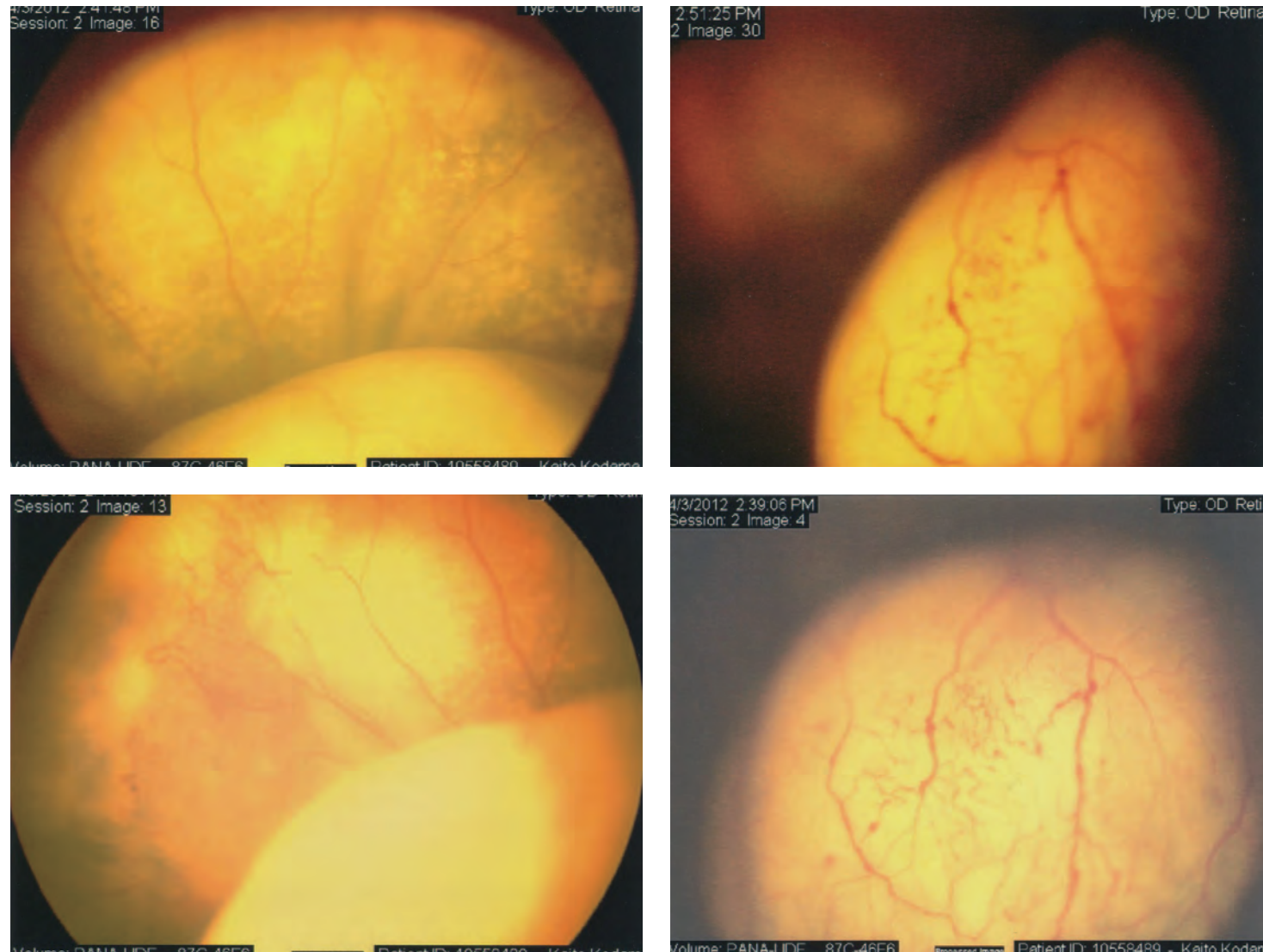
Retina:

OD: abnormal telangiectatic vessels and intraretinal exudation.

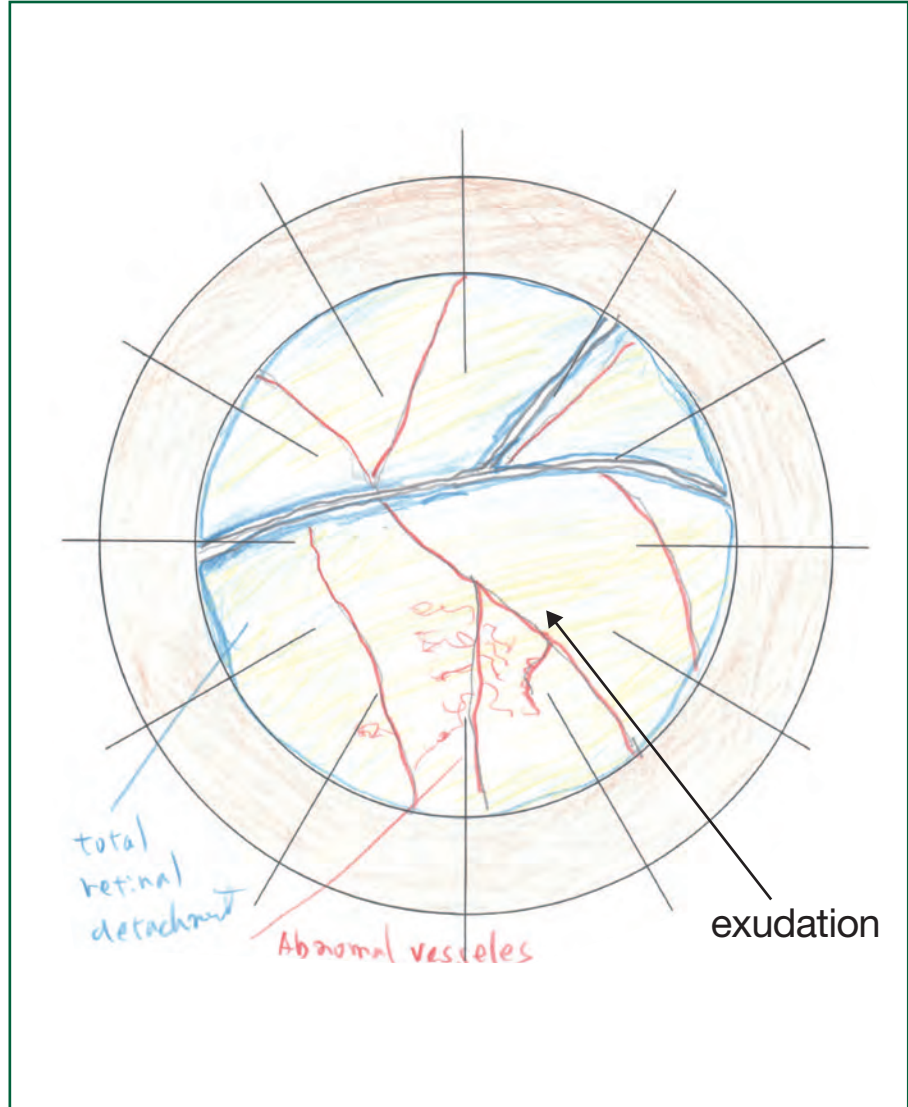
OS: no abnormality.

Fundus Images Before SOAI

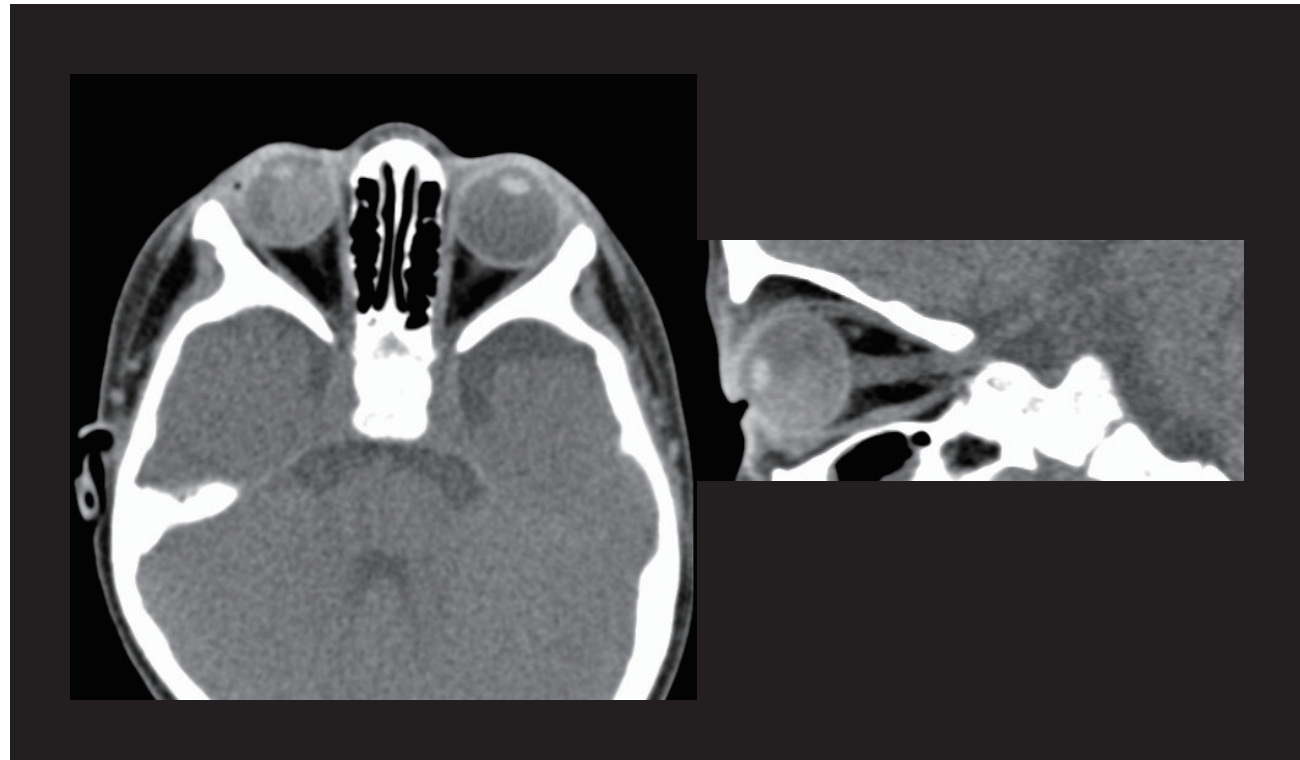
4/3/2012



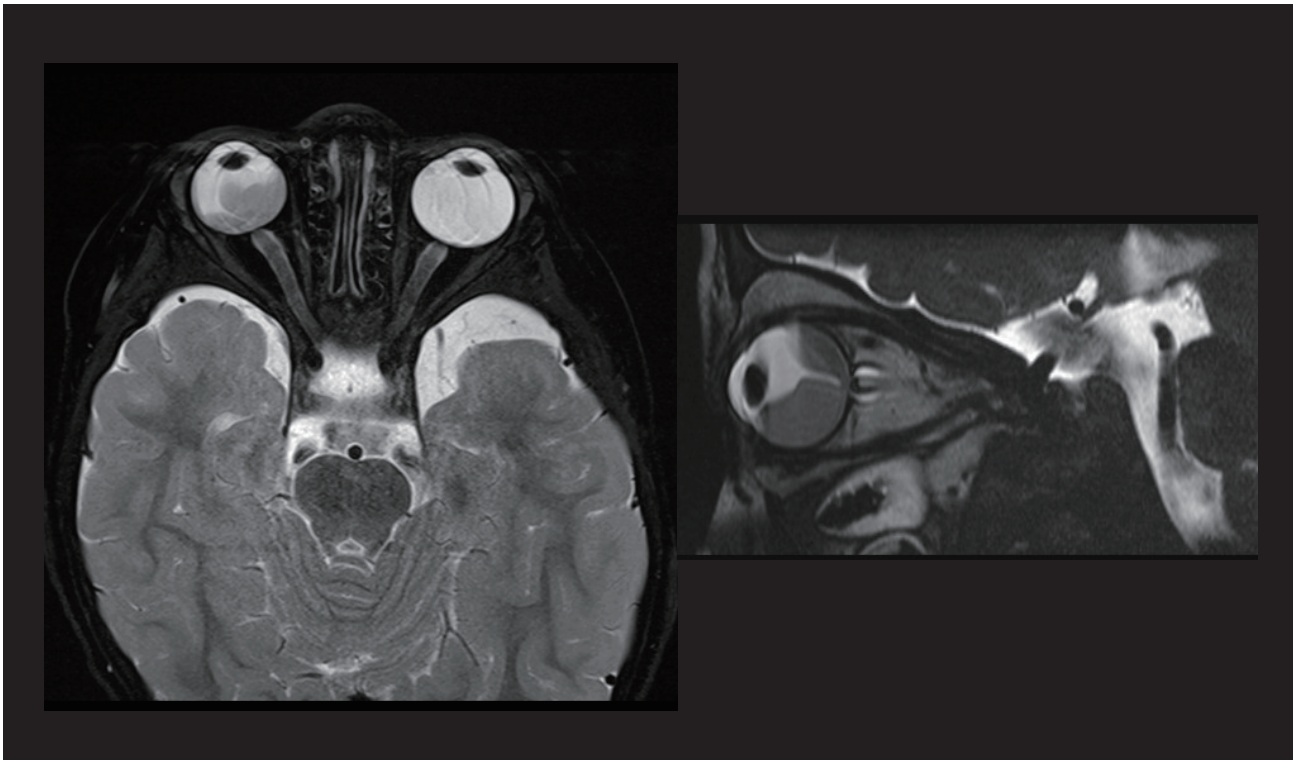
Fundus Sketch Before SOAI



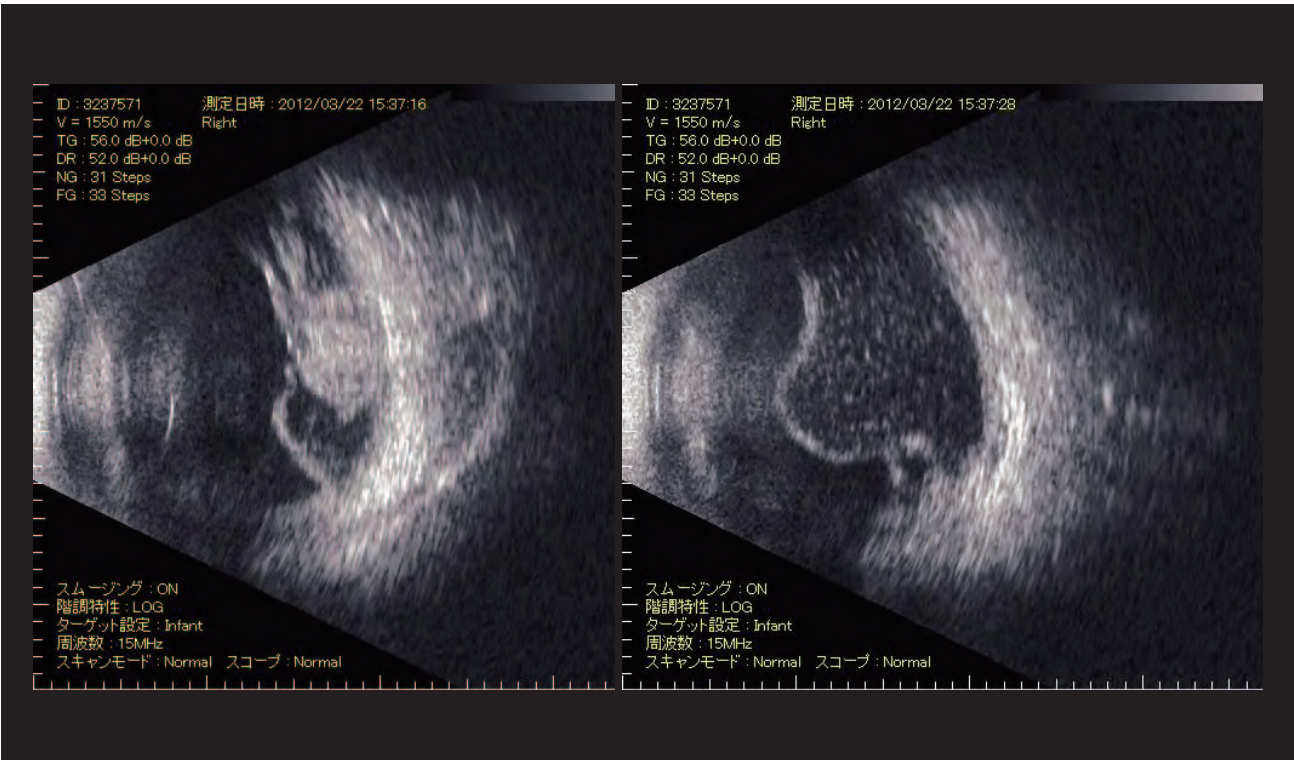
CT



MRI



ECHO



Method of Treatment

Under general anesthesia, a 4-F catheter was introduced from the femoral artery by the Seldinger technique and advanced into the right internal carotid artery. A microballoon catheter was then passed through the vascular catheter to a position beyond the orifice of the ophthalmic artery, and temporal occlusion was achieved at the portion just distal to the orifice by inflating the balloon. A total of 5 mg of melphalan and 1 mg of dexamethason were injected within several seconds using the introduced catheter, and the drugs flowed into the ophthalmic artery.

As routine, dexamethasone was injected to protect the endothelium of the artery from irritation by melphalan.

Angiogram of SOAI



Treatment & Course

Course:

April 3, 2012

Selective ophthalmic arterial injection of melphalan (5 mg) using a balloon catheter (SOAI) was performed on OD.

April 17, 2012

No change of the fundus condition was found.

July 2012

The bullous retinal detachment regressed extensively.

March 4, 2013

No retinal detachment was found.

August 21, 2013

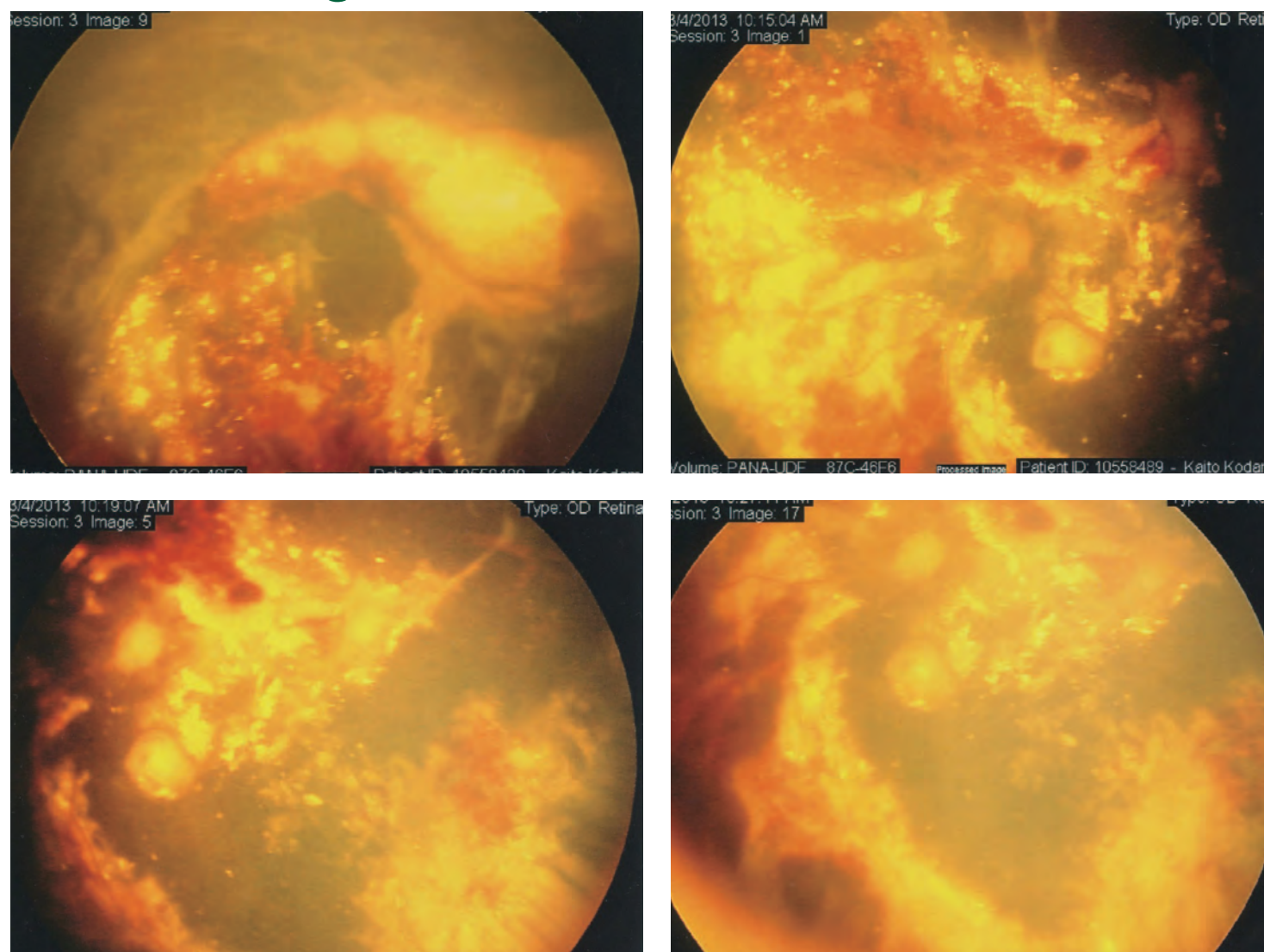
Stable

Visual acuity of OD = no light perception.

I.O.P = 15 mmHg = OU.

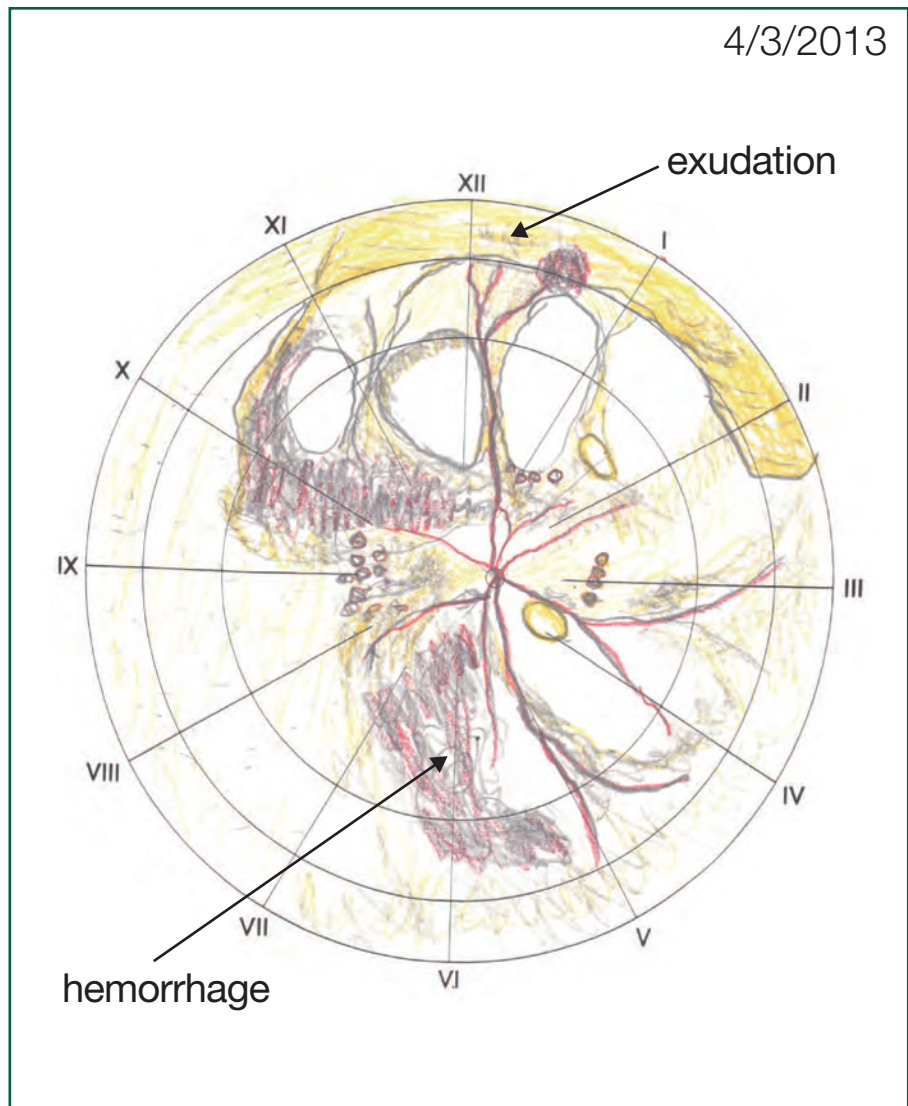
Fundus Images After SOAI

3/4/2013



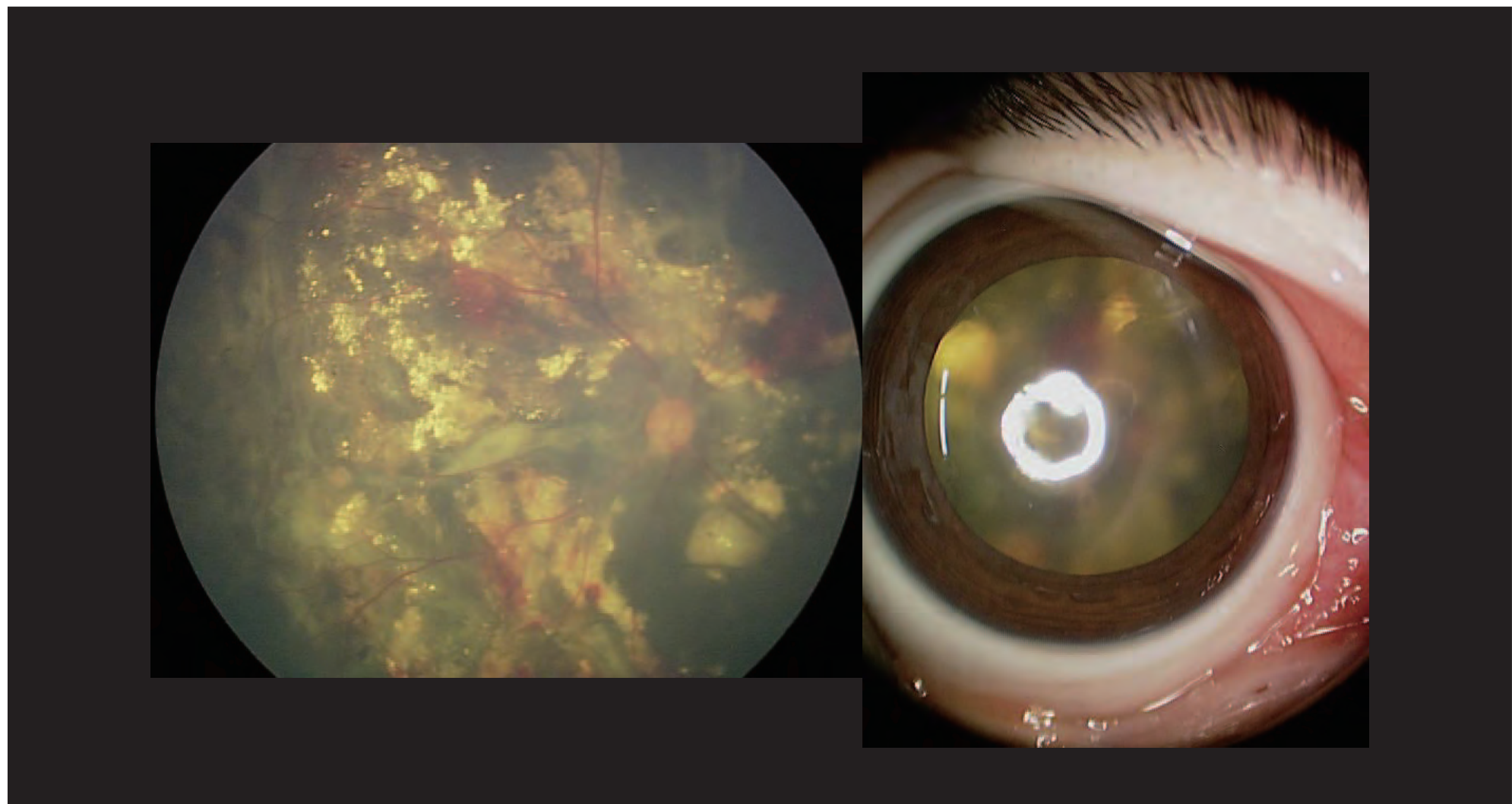
Fundus Sketch After SOAI

4/3/2013

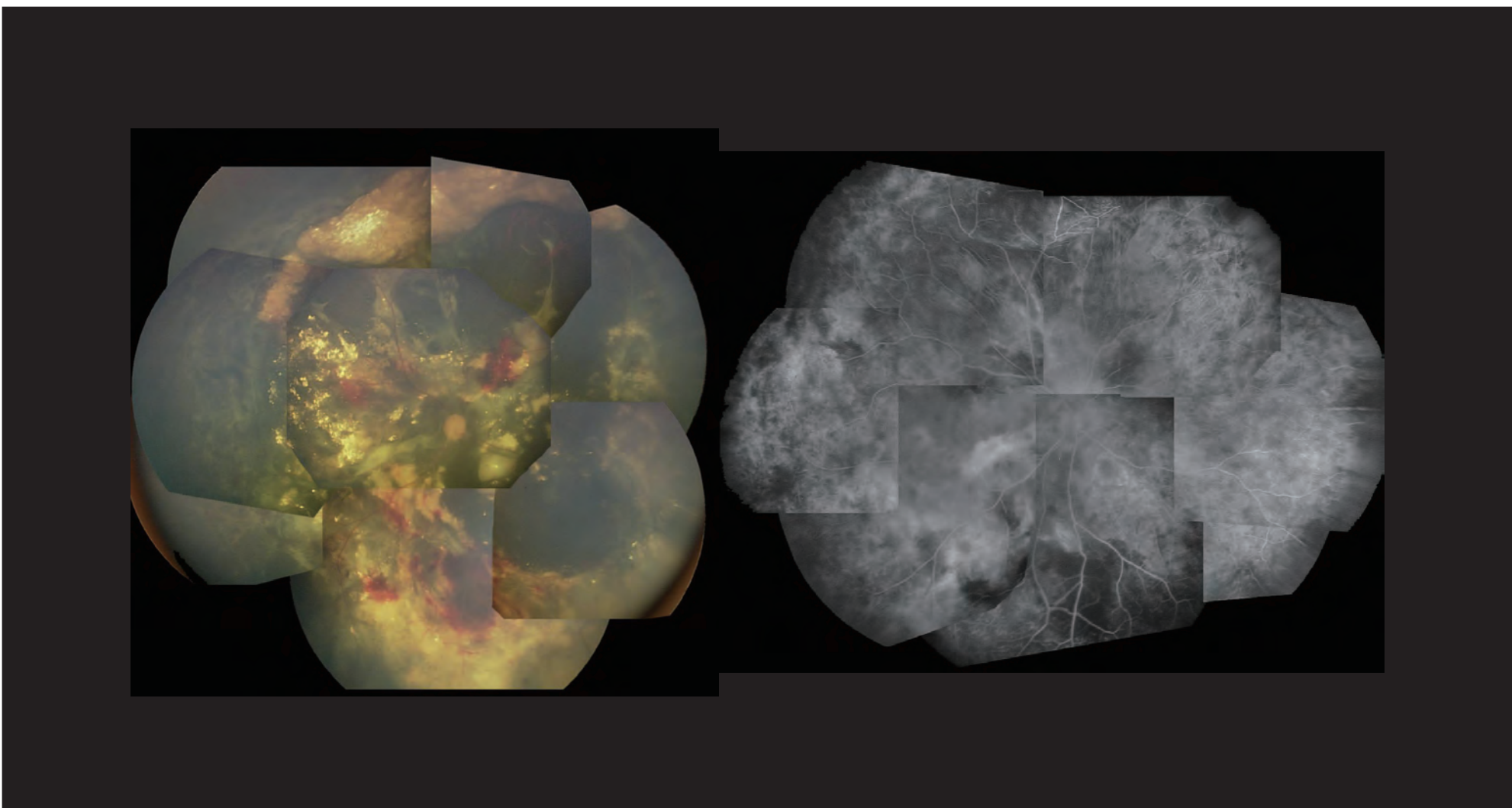


Fundus Image After SOAI

8/21/2013



Montage Fundus Images 15 Months After SOAI



Discussion

This is the first case report of successful treatment of Coats disease with bullous retinal detachment using the selective ophthalmic arterial injection of melphalan. Infrequent retinoblastomas showed no calcification despite extensive lesions. Therefore, surgical interventions must be very prudent in the case of Coats disease. The therapeutic diagnosis of this case was very appropriate, because of reported safety and efficacy of SOAI regarding retinoblastomas. However, almost all of them need SOAI several times until complete cure is achieved. Our results provide important evidence that Coats disease can be completely cured with only one time of SOAI. The mechanism of effectiveness of melphalan on Coats disease is estimated to be due to radiomimetic action of melphalan to abnormal vessels.

Conclusion

The selective ophthalmic arterial injection of melphalan is estimated to be useful for the treatment of Coats disease with bullous retinal detachment.

Reference

1. London, N.J.S. et al: Coats Disease 1058, Retina 5th Edition, Elsevier, 2013
2. Suzuki, S. et al: Selective ophthalmic arterial injection therapy intraocular retinoblastoma: the long-term prognosis. Ophthalmology 2011; 118:2081-7
3. Yamane, T. et al: The technique of ophthalmic arterial infusion therapy for patients with intraocular retinoblastoma. Int J Clin Oncol 2004; 9:69-73