

Coats' Retinitis or Retinoblastoma in a 3-Year-Old Girl: A Case Report

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Summary. Coats' disease is an idiopathic disorder defined by an abnormal development of retinal vessels with a progressive deposition of intraretinal or subretinal exudates, leading to exudative retinal detachment. The most difficult task is to differentiate Coats' disease from retinoblastoma. We present a rare case of Coats' disease diagnosed in a 3-year-old girl. From the age of 6 months, the girl was followed up 2 times a year at the Department of Ophthalmology, Hospital of Lithuanian University of Health Sciences, due to congenital convergent strabismus and refractive errors. At the age of 3.6 years, a routine examination of the fundus of the right eye revealed hard exudates, telangiectasia and tortuosity, gray color lesion below the optic nerve disc, submacular exudation in the inferior nasal part of the retina, and exudative retinal detachment, which extended from the 7-o'clock position to the 4-o'clock position. Before this examination, no abnormalities were found in the fundus of her both eyes. The girl was not treated with laser photocoagulation, cryocoagulation, or intravitreal injections, as the diagnosis of retinoblastoma could not be excluded; therefore, only eye drops were prescribed. In order to exclude the diagnosis of retinoblastoma, ultrasonography, magnetic resonance imaging, and computed tomography were carried out, and an appointment to see an ophthalmic oncologist was scheduled.

Due to early and appropriate treatment, the progression of Coats' disease in patients could be arrested. However, in some cases, when the diagnosis is ambiguous, it is better to follow up the patient and to treat only with eye drops.

Introduction

In 1908, George Coats described a group of patients who developed subretinal exudation associated with retinal telangiectasia with no apparent etiology (1). Later on, light and electron microscopic studies demonstrated that the loss of the normal vascular endothelial barrier was the basic defect in Coats' disease (2).

Coats' disease is an idiopathic disorder defined by an abnormal development of retinal vessels with a progressive deposition of intraretinal or subretinal exudates, leading to exudative retinal detachment (3). The disease is almost isolated and unilateral, and affects mainly males aged 4 to 8 years (4). The disease manifests unilaterally in 80% to 90% of patients without any evidence of racial, genetic, or familial predisposition (5).

The diagnosis includes direct and indirect ophthalmoscopy, color fundus photography, fluorescein angiography (FA), ultrasonography, computed

tomography (CT), magnetic resonance imaging (MRI), and diaphanoscopy. The most difficult task is to differentiate the patient's condition from retinoblastoma and to make the correct decision: observe, treat, or enucleate.

This case report describes the pathology of the right eye in a 3-year-old girl and an attempt to make a correct decision, which might have an influence on the patient's future.

Case Report

A 6-month-old girl was referred to the Department of Pediatric Ophthalmology, Hospital of Lithuanian University of Health Sciences, by the local ophthalmologist for an evaluation of congenital convergent strabismus and refractive errors. No pathological changes were detected in the fundi of both the eyes.

The girl is the second child in the family. She was born by caesarean delivery, and her birth weight was 4700 g. Past medical and family histories were unremarkable. Visual acuity was not measured because of young age. Convergent strabismus of 5–10 degrees was documented. Slit lamp examination of

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the anterior segments of both the eyes disclosed no abnormal findings. Ophthalmoscopy of the right and left eyes showed no abnormal findings. Refractive errors of the right and left eyes were -6.00 D sph and $+2.00$ D sph $+2$ D cyl X 90° , respectively. Eyeglasses and alternate covering were prescribed. The girl had doctor's appointments at the Department of Ophthalmology twice per year. No changes in both eyes were detected until the age of 3.6 years.

At the age of 3.6 years, the angle of the strabismus remained the same without any correction, and only oblique strabismus of the right eye was detected. No strabismus was detected with correction. The best-corrected visual acuity was 0.1 for her right eye and 0.5 for her left eye. Ophthalmoscopy of the right eye revealed hard exudates, telangiectasia, and vascular tortuosity, gray lesion below the optic nerve disc, submacular exudation in the inferior part of the retina, and exudative retinal detachment, which extended from the 7-o'clock position to the 4-o'clock position (Fig. 1). The clinical diagnosis of Coats' disease was made, but the diagnosis of retinoblastoma was not rejected.

In order to eliminate the diagnosis of retinoblastoma, ultrasonography (Fig. 2), MRI (Fig. 3), and CT (Fig. 4) were carried out, and an appointment to see an ophthalmic oncologist was scheduled.

Laboratory values were within the reference ranges. The C-reactive protein level was 0.07 mg/L; blood glucose level, 4.42 mmol/L. Anti-*Toxoplasma gondii* IgG antibodies were not detected. The anti-cytomegalovirus IgG antibody level was found to be 30 mIU/mL. Therefore, the girl was suspected to have a congenital cytomegalovirus infection.

Following MRI and CT investigations, eye drops were prescribed: Flarex 2 times per day and Unichlophen 3 times per day. Another appointment was scheduled 1 month later. After 1 month, ultrasonography and examination of the fundus of both eyes were repeated. Ultrasound examination demonstrated the neoplasm to be reduced (Fig. 5), and no changes were detected in the fundus of the right eye (Fig. 6).

The girl was further examined every 4 weeks, but no changes were observed. She was treated only with eye drops due to the lesion localized near the macula. Visual acuity was stable, and progression of the disease was not noted, including the decreasing lesion, which was seen on the ultrasound scan.

Discussion

Retinoblastoma with a prevalence of 22%–24% and Coats' disease with a prevalence of 4%–16% are two common congenital intraocular lesions in children with leukocoria (6). Retinoblastoma manifests before the age of 5 years and accounts for 11% of all cancers during the first years of human life.

The peak prevalence of Coats' disease is at the age of 6 to 8 years, though the age range at its presentation is quite wide (5 months to 71 years) (7). Coats' disease affects males predominantly and accounts for about 69% of all the cases. It progresses gradually, affects central vision, and is usually unilateral (i.e., affects only one eye). The diagnosis of Coats' disease is often possible by employing clinical examination. Ultrasonography, CT, and MRI are helpful for excluding other potentially lethal conditions, such as retinoblastoma, especially in very young patients (8). However, both conditions can be associated with calcifications, and therefore, it is difficult to differentiate between them on imaging (8). Ocular ultrasonography enables the detection of intraocular mass and calcification, which are characteristic features of exophytic retinoblastoma (2). In the advanced stages of Coats' disease, ultrasonography demonstrates a linear echo typical of retinal detachment, and sometimes a few prominent echoes due to the presence of subretinal cholesterosis (9). CT is an extremely valuable diagnostic method because of its capability to detect intraocular calcification (10). More than 90% of advanced retinoblastomas show evident calcification on CT scans (11). MRI can also be very helpful in the diagnosis of Coats' disease because it provides multiplanar imaging with high contrast resolution that yields insights into the biochemical composition of the intraocular structure. The presence of proteinaceous subretinal fluid in Coats' disease leads to MRI of typical T1- and T2-weighted sequences without the need to use radiation (12).

Therefore, a careful examination of both eyes is recommended to all patients with symptoms in only one eye because bilateral involvement may occur in Coats' disease. Moreover, a continuous follow-up is necessary if late complications are to be identified in their early stages and are treated adequately to prevent visual loss. FA of the fundus provides important information as well because it demonstrates the location and extent of retinal telangiectasia as well as the degree of permeability alteration in the affected vessels; however, FA has contradicting reactions regarding small children (13). The area of vascular abnormality as seen with angiography is usually more extensive than the one appreciated by clinical examination.

If the disease is detected in its early stage, vision can usually be restored to some extent. If it is not diagnosed until its late stages, complete loss of vision may occur. At the final stages of the disease, enucleation (removal of the affected eye) is a probable outcome (14). In a study by Shields et al., 117 patients (124 eyes) were followed up for a mean period of 55 months. In this subset of patients with Coats' disease, poor visual outcome (20/200 or



Fig. 1. Color fundus photography of the right eye

Hard exudates, telangiectasia and tortuosity, a gray lesion below the optic nerve disc, submacular exudation in the inferior part of the retina, and exudative retinal detachment that extended from the 7-o'clock position to the 4-o'clock position are seen in the fundus of the right eye.

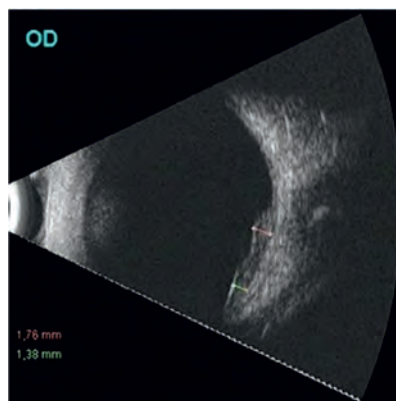


Fig. 2. B-mode ultrasound scan of the right eye

B-mode ultrasound scan shows a localized smooth lesion located in the aspect of the peripheral fundus. The neoplasm measured about $1.38\text{--}1.76 \times 10.92\text{ mm}$. Signal amplitude is not high and characteristic of retinoblastoma.



Fig. 3. Magnetic resonance imaging scan

T1- and T2-weighted images were obtained. An altered sector along the retina is about $\sim 1\text{ cm}$ in diameter and $\sim 0.15\text{ cm}$ in thickness in the posterior part of the right eye, and above it, contrast medium enhancement is present with its upper edge reaching the optic nerve disc. The changes are not specific because the lesion is very thin; however, the view is suggestive of Coats' disease.

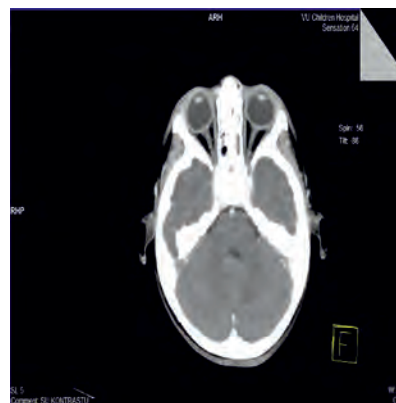


Fig. 4. Computed tomography scan

A very low nub of about 6.4 mm in length was noticed in the posterior-medial part of the right eye, but no contrast accumulation in this part was observed.

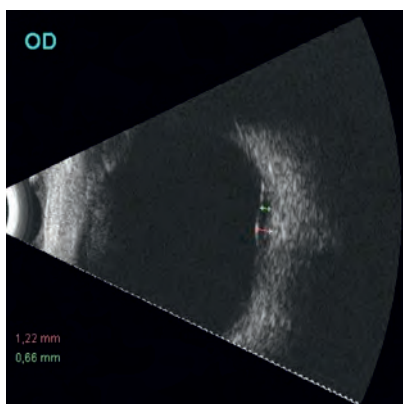


Fig. 5. B-mode ultrasound scan after 1 month

B-mode ultrasound scan after 1 month shows a localized smooth lesion located in the aspect of the peripheral fundus. The size of the neoplasm became smaller and measured $0.66\text{--}1.22 \times 6.18\text{ mm}$.



Fig. 6. Color fundus photography of the right eye after 1 month

Positive dynamics is observed and no changes were noticed in color fundus photography 1 month later.

worse) was found in 0% of eyes in stage 1, 53% in stage 2, 74% in stage 3, and 100% in stages 4 and 5 of Coats' disease. Enucleation was ultimately necessary in 0% of stages 1 and 2, 7% of stage 3, 78% of stage 4, and 100% of stage 5 disease (14).

Conclusions

Due to early and appropriate treatment, the progression of Coats' disease in patients could be arrested. However, in some cases, when the diagnosis

is ambiguous, it is better to follow up the patient and to treat only with eye drops. In our case, the decision was made to continue the treatment with eye drops as positive changes were seen and to follow up the girl every 2 weeks by employing B-mode ultrasound because this method is exceptionally informative and noninvasive.

Statement of Conflict of Interest

The authors state no conflict of interest.

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