Anterior Chamber Cholesterolosis in a 1-Year-Old Patient with Coats' Disease. A Case Report

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SUMMARY

Aim: To present a rare case of anterior chamber cholesterolosis in a patient with Coats' disease with emphasis on the differential diagnosis, to compare the clinical findings and therapeutic approach with previously reported cases; and to clarify the pathogenetic mechanisms and associated complications of anterior chamber cholesterolosis, based on an analogy with previously published cases.

Case report: A 14-month-old boy was brought to our clinic with a painful, irritated right eye. Anterior chamber cholesterolosis was identified, associated with acute glaucoma, total retinal detachment, and vitreous hyperechogenicity with suspected calcifications on ultrasound (US). Coats' disease with anterior chamber cholesterolosis or retinoblastoma-associated masquerade syndrome were considered in the differential diagnosis. Magnetic resonance imaging (MRI) did not confirm retinoblastoma. The patient underwent anterior chamber lavage, followed by intracameral and intravitreal injection of an anti-vascular endothelial growth factor (anti-VEGF) agent. A cytological analysis of the anterior chamber aspirate showed no evidence of malignant cells. Although the treatment achieved the expected effect, the eye gradually progressed to phthisis bulbi. Nevertheless, the eye has remained painless, it has shown no signs of irritation and the intraocular condition has remained stable over the long term.

Conclusion: Anterior chamber cholesterolosis is a rare complication of Coats' disease, reported in less than 3% of patients. Exclusion of retinoblastoma-associated masquerade syndrome is essential in the differential diagnosis. In comparison with previously published cases of anterior chamber cholesterolosis as a complicating factor in Coats' disease, we have observed a similar character and course of the disease in patients with onset in early childhood. However, a difference can be observed in the therapeutic approach. In all the previously reported cases, the painful eye was enucleated. In our case, preserving the blind eye through a more conservative surgical approach proved beneficial.

Key words: Coats' disease, anterior chamber cholesterolosis, anti-VEGF

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INTRODUCTION

Coats' disease is an idiopathic retinal vascular disorder manifested especially in childhood age [1]. The disease is typically sporadic, without systemic abnormalities, and predominantly affects boys and young men. Women and girls are affected in approximately 16% of cases. [2]. It is considered to be a unilaterally manifested disorder, although recent studies also indicate the possibility of bilateral manifestation in more than 18% of patients with Coats' disease [3]. Most cases are diagnosed in the first and second decade of life. The severity of manifestation and the visual prognosis are generally worse in younger patients.

Characteristic findings are telangiectasia (dilated retinal blood vessels of irregular caliber, small to medium in size), tortuosity, micro- and macro-aneurysms, and dilated arteriovenous anastomoses [2,4]. These changes are located in the region of the equator and the periphery of the retina, most commonly in the inferior temporal quadrant. Upon progression of the disease, intraretinal and subretinal exudation takes place, up to subsequent serous retinal detachment [5]. Subretinal exudations are rich in foam cells (macrophages accumulating lipids, in particular LDL cholesterol) and cholesterol crystals. The presence of cholesterol is therefore limited to the subretinal space. Shields et al. were the first to describe a clinical-pathological correlation of massive cholesterolosis of the anterior chamber, which occurred in a patient with Coats' disease [6]. According to the data available to us, only 4 case reports have been published to date in which Coats' disease was complicated by the presence of cholesterol crystals in the anterior chamber [6-9]. In addition to these publications, Shields et al. in their retrospective study described anterior chamber cholesterolosis in 4 patients out of a cohort of 150 patients [10].

CASE REPORT

The mother of a 9-month-old boy noticed a whitish reflection in the pupil of her son's right eye. The

infant was examined at the local eye clinic, and was subsequently referred to a more specialized center. On the basis of an examination conducted under general anesthesia (GA), and according to magnetic resonance imaging (MRI) of the head, the boy was diagnosed with Coats' disease in the right eye with neovascular glaucoma and bullous retinal detachment at the age of 11 months. The finding corresponded to stage 4. Local antiglaucoma therapy was commenced, with a recommendation to monitor the finding.

The infant's general condition of health was good. On MRI of the head a chance finding of venous aneurysm was described on the left side. From the medical history of the boy's family we learned only that his mother suffered from myopia and had undergone laser surgery, otherwise the family medical history was without remarkable features. At the age of 14 months the patient was brought to the Department of Pediatric Ophthalmology at the University Hospital Brno due to pain redness of the right eye, three months after his diagnosis with Coats' disease. An objective macroscopic examination showed pronounced irritation of the right eye; presence of mixed conjunctival injection. The anterior chamber was filled with yellowish matter and the red reflex was absent. The right eye was without light perception.

There followed an examination under GA. A microscopic examination revealed a diffuse mass of yellowish crystalline deposits filling the entire anterior chamber of the right eye. Figure 1. This finding prevented any view of the pupil and other structures, including the posterior segment. Intraocular pressure was 38 mmHg. Upon US examination of the right eye dense echogenic mass was visible, with suspected calcifications virtually throughout the entire vitreous cavity. Figure 2. In the differential diagnosis it was necessary to exclude atypical retinoblastoma-associated masquerade syndrome, of which calcifications are pathognomonic. The finding on both the anterior and posterior segment of the left eye was physiological, intraocular pressure was within the norm. The dosage of local antiglaucoma therapy in the right eye was increased, and a carbonic anhydrase inhibitor – acetazolamide – was administered generally. On the MRI scans the image did not correspond to retinoblastoma, but to exudative retinitis; post-contrast no pathological saturation was evident. Figure 3B.

Following partial reduction of intraocular pressure, lavage of the anterior chamber was performed, with a sample taken for a cytological examination. During lavage the iris became visible, with pronounced rubeosis and neovascularizations in the iridocorneal angle (ICA). The lens was subluxated with diffuse opacity, and leukocoria was evident. It was not possible to differentiate the posterior segment. Figure 4A. Application of the anti-VEGF agent bevacizumab into the anterior chamber was subsequently indicated, performed immediately after lavage.

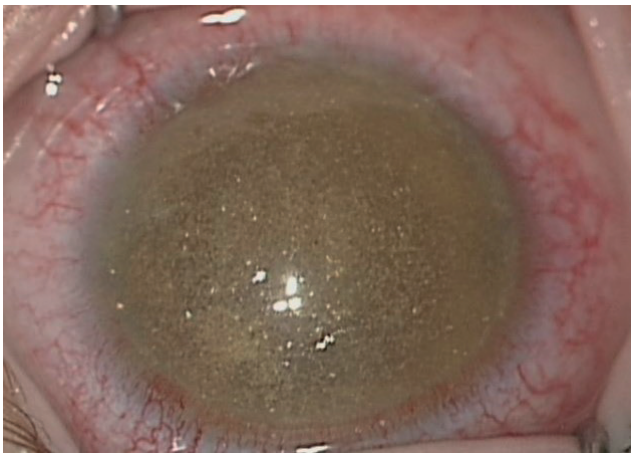

Figure 1. Findings on the anterior segment of the right eye during the first examination at our clinic. Mixed conjunctival injection, anterior chamber completely filled with a mass of shimmering "golden" cholesterol crystals

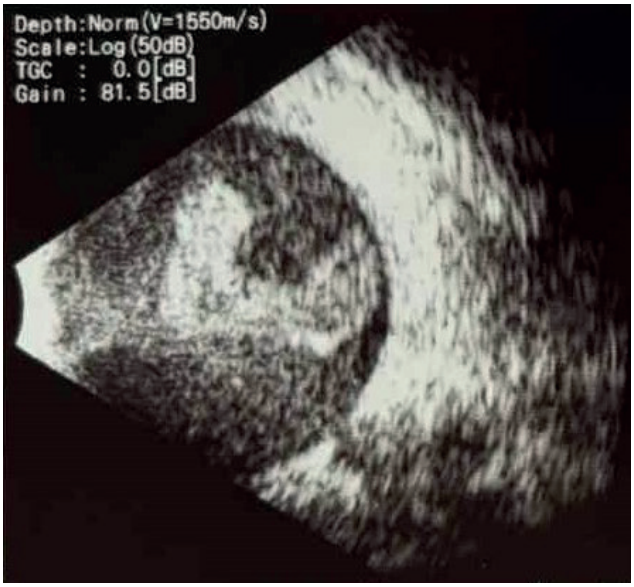

Figure 2. B-scan ultrasound of the right eye reveals a highly echogenic masses in almost the entire vitreous cavity, suspicious for calcifications

A cytological examination of the anterior chamber punctate fluid demonstrated inflammatory changes with a content of neutrophiles, lymphocytes, erythrocytes, lymphoid cells, without any finding of malignant cells. The following diagnosis was determined: Coats' disease – stage 4 with anterior chamber cholesterolosis and neovascular glaucoma. Two further doses of an anti-VEGF agent were planned.

A significant regression of neovascularization of the iris and ICA was recorded one month after the first application of the pharmaceutical bevacizumab, and intraocular pressure was normalized at 12 mmHg. Isolated fluctuating cholesterol crystals were visible in the anterior chamber, the pupil was unresponsive, in irregular wide mydriasis. Figure 4B. The patient was administered a second, intracameral and intravitreal

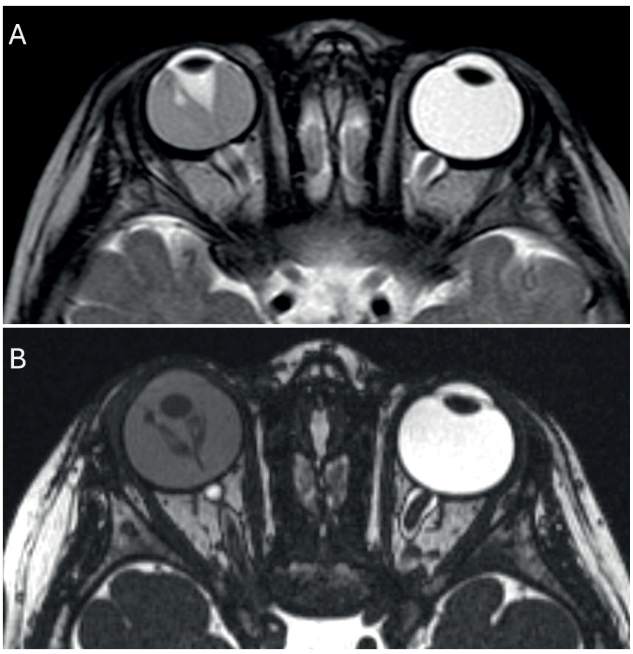

Figure 3. Axial section through the patient's orbits on T2-weighted MRI. At the time of diagnosis of Coats' disease, there is retinal detachment in the right eye with hypointense subretinal material; the remaining vitreous cavity, as well as the posterior and anterior chambers are hyperintense, and the lens is not subluxated (**A**). Subsequently, with the onset of anterior chamber cholesterolosis, the entire content of the affected right eye becomes hypointense, indicating protrusion of cholesterol material from the subretinal space into the vitreous and into the posterior and anterior chambers, accompanied by lens subluxation (**B**)

MRI – magnetic resonance imaging

dose of anti-VEGF agent. Antiglaucoma therapy was discontinued.

After one more month, examination under GA demonstrated complete regression of neovascularizations of the iris and ICA. Only a small residue of cholesterol crystals persisted in the anterior chamber and on the anterior surface of the lens. Figure 4C. The eyeball was hypotensive, with intraocular pressure of 5 mmHg. With reference to the satisfactory finding, administration of a third dose of anti-VEGF agent was not indicated.

Further follow-up examinations were conducted in outpatient care. Over the course of time the eyeball diminished in size. However, it remained pacific and painless. The cornea is diminished in size, with central zonular keratopathy. No cholesterol crystals are evident in the anterior chamber. The iris is pacific and the red reflex is absent. The ocular finding has remained stable for a period of five years, without signs of activity of the disease.

DISCUSSION

Coats' disease is a rare disorder characterized by the appearance of idiopathic retinal telangiectasias associated with intraretinal and/or subretinal exudations, and numerous serous retinal detachments without manifest retinal or vitreous traction [10]. Shields et al. proposed

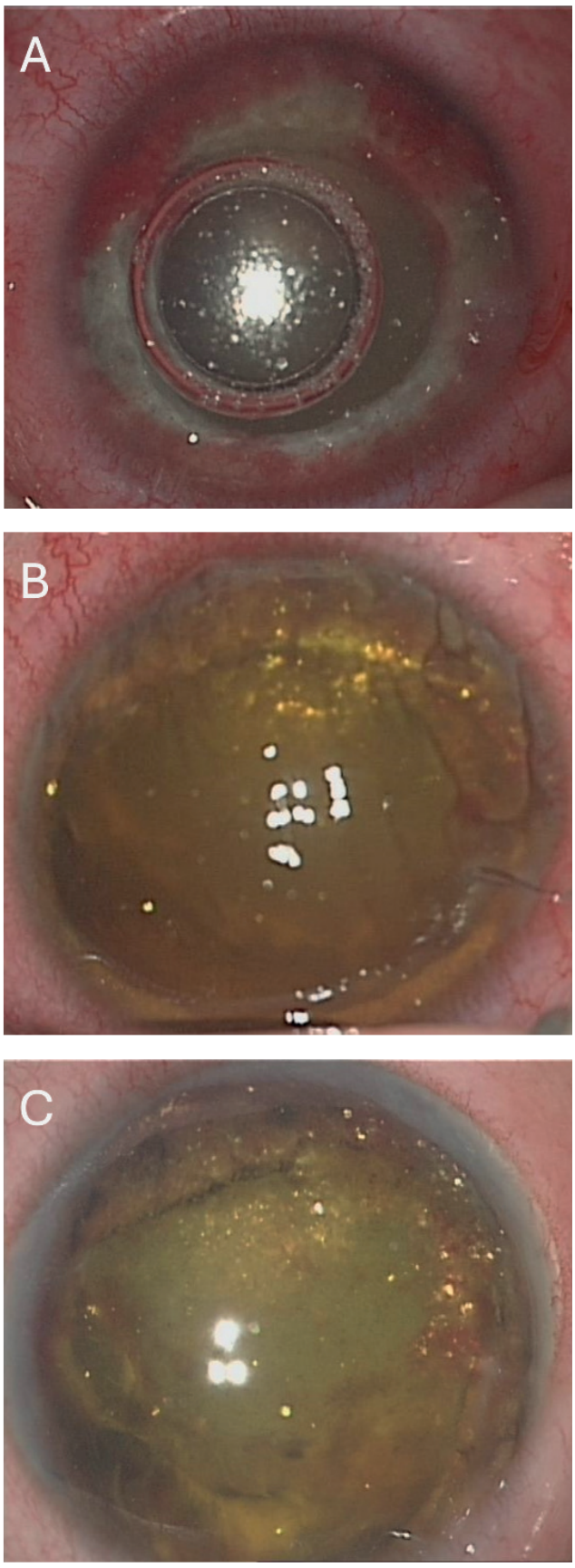

Figure 4. Anterior segment findings immediately after anterior chamber lavage (**A**), one month after the first application of anti-VEGF (**B**), and one month after the second anti-VEGF injection (**C**)

Anti-VEGF – anti-vascular endothelial growth factor

a classification of Coats' disease on the basis of an observation of clinical symptoms in 150 cases of their retrospective study [4,11]. This "staging" system was subsequently updated. This classification is illustrated in Table 1 [12]. In the advanced stages, manifestations of the anterior segment may also appear, such as cataract, neovascularization of the iris and ICA, or anterior chamber cholesterolosis, which is a very rare complication of Coats' disease, reported in fewer than 3% of patients with Coats' disease [7,13].

From the beginning of 1994 to the end of 2024 a total of 28 patients with a diagnosis of Coats' disease were examined and subsequently monitored at our clinic, out of whom anterior chamber cholesterolosis was detected in only one patient. The precise pathogenesis of this phenomenon has not yet been entirely clarified. It probably occurs as a consequence of the migration of cholesterol crystals from exudations in the subretinal space into the anterior chamber. For this to occur, structural changes must take place in the retina, which would enable the transfer of cholesterol into the vitreous [7]. In all five cases published to date, including the case of our patient, chronic serous retinal detachment was present [6-9]. The appearance of a tear or atrophic hole within the terrain of the detached retina, which would additionally aid the migration of cholesterol from the subretinal space into the vitreous, is also presumed [8]. However, in our case no defect in the retina was demonstrated.

An US B-scan showed a mass of cholesterol crystals as hyperechogenic opacities filling virtually the entire vitreous cavity. Chronic inflammation and abnormalities of the suspensory ligaments of the lens, which are also potential findings in patients in the advanced stages of Coats' disease, may contribute to the transfer of the mass of cholesterol from the vitreous to the posterior and anterior chamber [7]. In our patient the lens was subluxated, which was also confirmed by the finding on the MRI scans.

The aforementioned finding on the US B-scan also caused us to suspect calcifications, which are pathognomonic of retinoblastoma as the most common primary intraocular malignancy in childhood age [1]. MRI did not demonstrate calcifications or post-contrast pathological saturation, which is typical of retinoblastoma. To date only one case of a patient with bilateral retinoblastoma complicated by anterior chamber cholesterolosis unilaterally is referred to in the literature [14].

Secondary glaucoma is a common serious complication in patients with Coats' disease [10]. In our patient Coats' disease was diagnosed at the age of 11 months, and was then already in stage 4, which means that retinal detachment and secondary neovascular glaucoma were present. However, three months later the patient suffered an acute glaucoma attack, and in addition a new finding was present – aforementioned anterior chamber cholesterolosis. It is possible to assume that cholesterol crystals caused an obstruction of the trabecular meshwork, which led to a glaucoma crisis. The intraocular finding, as well as the course of the disease, indicate that this concerned a combination of two types of glaucoma, namely neovascular glaucoma in the advanced stage of Coats' disease and acute open-angle glaucoma triggered by obstruction of the trabecular meshwork by cholesterol crystals. We find a similar cause of the disease in two previously published cases [6,7]. In all three cases, including our own patient, Coats' disease was diagnosed in stage 3B-4, and subsequently, within a time frame of 1–13 weeks a new finding appeared – anterior chamber cholesterolosis and acute glaucoma with manifestation of a painful, irritated eye. All patients were aged under two years at the time of diagnosis.

Table 1. Classification of Coats' disease

Stage	Clinical findings
Stage 1	Retinal Telangiectasia only
Stage 2	Telangiectasia and exsudation A. Extrafoveal B. Foveal 1. Without subfoveal nodule 2. With subfoveal nodule
Stage 3	Exudative retinal detachment A. Subtotal retinal detachment 1. Extrafoveal 2. Foveal B. Total retinal detachment
Stage 4	Total retinal detachment and secondary glaucoma
Stage 5	Advanced end-stage disease

A difference can be observed in the approach to the treatment of the ensuing acute condition. Previous authors chose the approach of enucleating the painful eye, thereby at the same time obtaining material for a more detailed histopathological examination and description of this rare condition [6,7]. In our case we chose a less invasive therapeutic strategy, consisting in the lavage of a mass of cholesterol following partial reduction of intraocular pressure by pharmacotherapy and subsequent intracameral and intravitreal application of anti-VEGF treatment with bevacizumab.

It is assumed that vascular endothelial growth factor (VEGF) plays the chief role in the progression of Coast's disease. A number of studies have demonstrated raised levels of VEGF in the intraocular fluids of eyes with Coats' disease. For this reason anti-VEGF therapy is used as an adjuvant treatment for Coats' disease [15–17].

Therapy was administered in 2019, when treatment using anti-VEGF agents in pediatric patients was indicated outside of approved use (off-label), and as a result the informed consent of the infant's parents was essential for the application. At the time in question no standardized protocol was available for the administration of anti-VEGF agents to pediatric patients [18]. The treatment was targeted primarily at the anterior segment, where a significant regression of neovascularizations of the iris and ICA

was recorded one month after the first application. One month after the second application, regression was complete, the intraocular finding was satisfactory, and the eye was painless, without signs of irritation. Our aim was to choose a procedure that would lead to the removal of anterior chamber cholesterolosis, which appeared to be the trigger of the glaucoma crisis, and to eliminate the neovascularizations of the iris and ICA, resulting in the preservation of the eyeball. With reference to our stated aim we consider the approach we chose to be effective.

Over the course of time the eye progressed to phthisis bulbi, but is painless, and the ocular finding has now remained stable for several years.

CONCLUSION

Anterior chamber cholesterolosis is a rare complication of Coats' disease, reported in fewer than 3% of cases. In our case report we presented the case of a one-year-old boy with stage 4 Coats' disease, in whom anterior chamber cholesterolosis accompanied by acute glaucoma had developed. We emphasize the importance of differential diagnosis, in particular differentiation from retinoblastoma as the most common primary intraocular malignancy in childhood age. In comparison with previously published cases of anterior chamber cholesterolosis as a complication of Coats' disease, we found a similar character and course in patients with manifestation of the disease in infancy. By contrast, we see a difference in the therapeutic approach. In all previously published cases the eye was enucleated. From our perspective it was beneficial to preserve the eye, and for this reason we chose a far more conservative surgical procedure, namely lavage of the anterior chamber, and intracameral and intravitreal application of an anti-VEGF agent. In doing so we also wish to highlight the effectiveness of anti-VEGF agents as a supplementary therapeutic modality in the treatment of Coats' disease.

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