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EC OPHTHALMOLOGY
Case Report

Bilateral Coats Disease in a Young Girl

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Abstract

We bring a case of Coats disease in a young girl of seven years with no particular history following the examination of her fundus, she was brought by her mother for an ophthalmology consultation at the CHU -IOTA for a progressive decrease in her visual acuity. Clinical examination found visual acuity limited to light perception at ODG. Examination of the anterior segment is normal. His fundus after papillary dilation shows in the right eye an intravitreal hemorrhage associated with a serous retinal detachment and a retinal detachment in the inferior temporal, in the left eye we also have an intravitreal hemorrhage with a total retinal detachment. Fluorescein angiography notes inferotemporal retinal detachment, hyperpermeability of capillaries, venous dilation, retinal neovessels on the right and on the left and total retinal detachment. Faced with these clinical and paraclinical signs, we concluded that the form of Coats disease in ODG is complicated.

Keywords: *Coats Disease; Bilateral; Young Girl*

Introduction

Coats disease is a rare retinal angiomatosis consisting of primary telangiectasias and exudative retinopathy that can be complicated by retinal detachment, vitreoretinal hemorrhages, neovascular glaucoma and ultimately loss of vision. Usually unilateral, it most often affects young boys in the first decade [1,2]. Its incidence is 0.09 per 100,000 inhabitants [3]. Of undetectable etiology probably congenital but not familial [4]. It is associated with a somatic mutation of the NDP gene located on chromosome Xp11.2 [5]. The diagnosis of Coats disease is clinical, due to the existence of numerous clinical forms varying according to the age of onset and the evolution of the exudative phenomenon. The diagnosis of this serious condition remains difficult on a practical level. To optimize the functional prognosis, treatment must be started early [3,6]. Coats disease is an important differential diagnosis of unilateral retinoblastoma in the presence of unilateral leukocoria and when growth of the lesion into subretinal layers mimics exophytic retinoblastoma [7,8].

Observation

We report the case of a seven-year-old female patient, with no previous history, brought to an ophthalmology consultation for progressive decline in visual acuity in both eyes. The examination revealed visual acuity limited to light perception. Ocular tone was measured at 8 mmhg in ODG. The anterior segment examination was normal in ODG. At the back of the eye, we found in the right eye an intravitreal hemorrhage associated with a serous retinal detachment and a retinal detachment in the inferior temporal, in the left

eye we also found an intravitreal hemorrhage with a total retinal detachment. Fluorescein angiography noted an inferotemporal retinal detachment, hyperpermeability of the capillaries, venous dilation, new retinal vessels on the right (Figure 1 and 3) and on the left a total retinal detachment (Figure 2 and 4). Treatment based on photo coagulation using emergency endolaser was offered to the patient's parents; in the absence of a technical platform, a referral to other authorized countries was suggested but could not be honored due to lack of financial means.

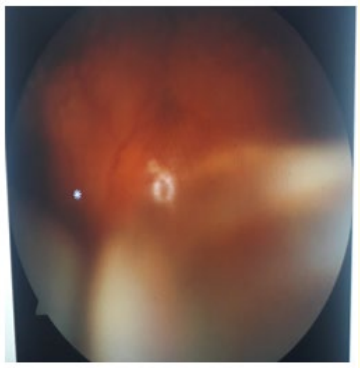

Figure 1: *AGF color image of the right eye.*

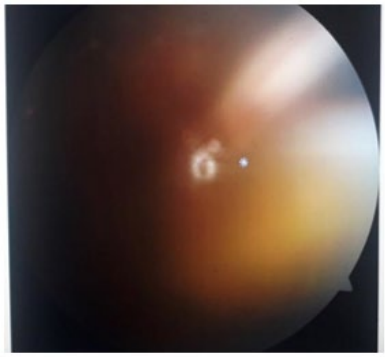

Figure 2: *AGF color image of the left eye.*

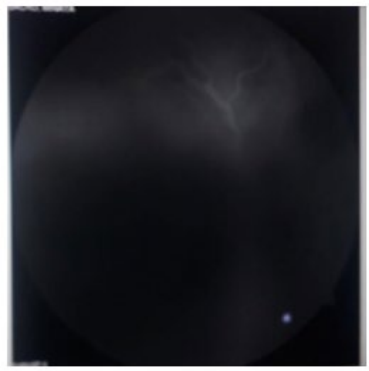

Figure 3: *Filtered AGF image of the right eye.*

Figure 4: *Filtered AGF photo from the left eye.*

Discussion

Characterized by peripheral retinal damage, Coats disease is generally temporal and/or inferior [1]. The essential signs on angiography are marked by localized microaneurysms, vascular ectasias, rarefaction and dilation of the capillary bed in the periphery [9]. Funduscopic examination reveals circinated exudates, more or less significant exudative maculopathy, intravitreal hemorrhage associated with serous retinal detachment and partial or total retinal detachment in severe forms. Telangiectasia is demonstrated by fluorescein angiography but can also be visible by biomicroscopy [10]. Bilateral localization being unusual 0 to 10% In some cases, a general pathology must be investigated [4-11]. Our patient presents the bilateral form and has no general pathology. It is essential to try to stop the progression of Coats disease by destroying the telangiectasias responsible for intra and subretinal exudation [12]. The treatment of choice is photocoagulation; recurrences remain possible, hence the need for regular monitoring [13]. In cases of significant retinal elevation, these coagulations can only be carried out after often difficult drainage of the subretinal fluid [14]. In our case, endolaser photocoagulation could only be performed in the right eye after vitrectomy, drainage of subretinal fluid and retinal detachment surgery or cryotherapy. Treatment remains particularly difficult at the stage of total retinal detachment [15]. Treatments remain difficult in advanced unilateral forms, requiring long-term monitoring of the adelphic eye; late asymmetric damage, although rare, is still possible [2,10]. The visual prognosis is reserved for the stage of complications, which explains the visual acuity of our patient with a complicated, bilateral form of ODG which risks progressing to blindness in the absence of treatment [16]. The disease can appear at any age but generally affects young boys and is unilateral in 95% of cases; our case is female with an age of discovery of seven years [17-19]. Analysis of the various publications made on Coats disease over the past 50 years seems to show that the later the syndrome is revealed, the more benign and less progressive the disease is [20]. Therapeutic management must be early, in a specialized environment; treatment started at an uncomplicated stage of the disease improves the chances of recovery [16]. In children, major forms have a poor functional and anatomical prognosis regardless of the technique chosen and/the existence of profound amblyopia considerably reduces a better anatomical result [15]. The main reason for consultation was the progressive decline in visual acuity, as is the case in the majority of cases.

Conclusion

Coats disease is a serious illness of unknown etiology, which can compromise the functional visual prognosis in complicated forms. Majority among male subjects. The bilateral location is unusual, it should lead to the search for a general pathology. Its diagnosis is clinical and management remains difficult in cases of total retinal detachment. However, early treatment started in a specialized environment at an uncomplicated stage of the disease improves the chances of recovery.

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