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Service d'Ophtalmologie - Hôpital Jules Gonin de Lausanne

Management of long-term Coats' disease

THESE

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par

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RAPPORT DE SYNTHESE

La maladie de Coats est une vasculopathie non héréditaire. Elle est caractérisée par la présence de télangiectasies rétiniennes idiopathiques, d'exsudats lipidiques intrarétiniens et sousrétiniens, une rétinopathie ischémique et un décollement de rétine exsudative. Cette maladie se présente typiquement dans l'enfance, elle est unilatérale et atteint les hommes dans la majorité des cas. Nous décrivons un cas atypique d'un patient avec une maladie de Coats qui a récidivé 30 ans plus tard malgré un traitement initial efficace. Ce cas illustre l'évolution de la maladie de Coats à long terme.

L'enjeu de ce cas est de faire une étude exhaustive des différents traitements possibles de cette maladie.

Nous avons réalisé une révision de la littérature des cas de la maladie de Coats qui ont récidivé à long terme. Il y a peu de cas décrits dans la littérature avec un long suivi.

En conclusion, la maladie de Coats doit être considérée comme une maladie chronique qui nécessite un suivi à long terme. Cette maladie peut se réveiller et récidiver dans des zones de la rétine, qui n'ont pas été atteintes auparavant, et plusieures décennies plus tard.
Le traitement standard de cette maladie est la réalisation d'une cryothérapie et du laser argon dans les zones atteintes de la rétine. Dans les cas où l'exsudation rétinienne est très importante il peut s'avérer de faire un traitement chirurgical avec drainage du liquide sousrétinien, ce qui a été réalisé sur ce patient.

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CASE REPORT

Reemergence of dormant Coats disease after 30 years

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PURPOSE. *We describe an atypical case of a patient with Coats disease that re-emerged after 30 years, illustrating a previously poorly understood long-term evolution of the disease.*

METHODS. *A 20-year-old man consulted for visual acuity (VA) decrease in the left eye (LE) to 0.3. Fundus examination revealed an exudative lesion with telangiectasias in the superior peripheral retina compatible with the diagnosis of Coats disease.*

RESULTS. *The patient was treated with cryotherapy and argon laser. Visual acuity improved to 0.5 and remained stable during a 1-year follow-up. The patient did not seek further clinical follow-up. Thirty years later, he returned complaining of a progressive VA decrease in the LE. Snellen VA was measured to counting fingers. Fundus examination revealed stage 3A Coats disease with macular exudation and a serous retinal detachment in the inferior quadrants requiring the placement of an encircling band, external drainage, and cryotherapy of the vascular lesions. After 10 additional sessions of argon laser on the vascular malformations, exudation regressed further and best-corrected VA increased to 0.1 at the end of the follow-up period.*

CONCLUSIONS. *Coats disease must be considered as a chronic disease, which necessitates a very long-term follow-up even in the absence of subjective visual loss. The disease can reawaken and recur with force in previously unaffected areas of the retina several decades later. The gold standard treatment consists of cryotherapy and argon laser. However, in cases of very important retinal exudation, surgical management with subretinal drainage may be necessary.*

KEY WORDS. *Coats disease, Cryotherapy, Exudative retinal detachment, Intraretinal and subretinal lipidic exudates, Laser photocoagulation, Retinal telangiectases*

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INTRODUCTION

Coats disease is a non-hereditary vasculopathy characterized by idiopathic retinal telangiectases, intraretinal and subretinal lipidic exudates, ischemic retinopathy, and exudative retinal detachment (1). Coats disease typically presents in early childhood with vision loss, strabismus, or leukocoria. It usually occurs unilaterally in males (1). We describe an atypical case of a patient with Coats disease that illustrates the evolution of the disease over a 30-year period.

Case report

A 20-year-old man presented for the first time to our department in 1975 due to gradual visual loss. The ophthalmic examination of his right eye (RE) was normal. The left eye (LE) had a best-corrected visual acuity (VA) of 0.3 (–1.0/180°). Biomicroscopy showed no inflammation of the anterior segment and a normal intraocular pressure (IOP). On fundus examination, an exudative lesion with telangiectases could be seen in the superior peripheral retina and the diagnosis of Coats disease was made (Fig. 1, A-D). The

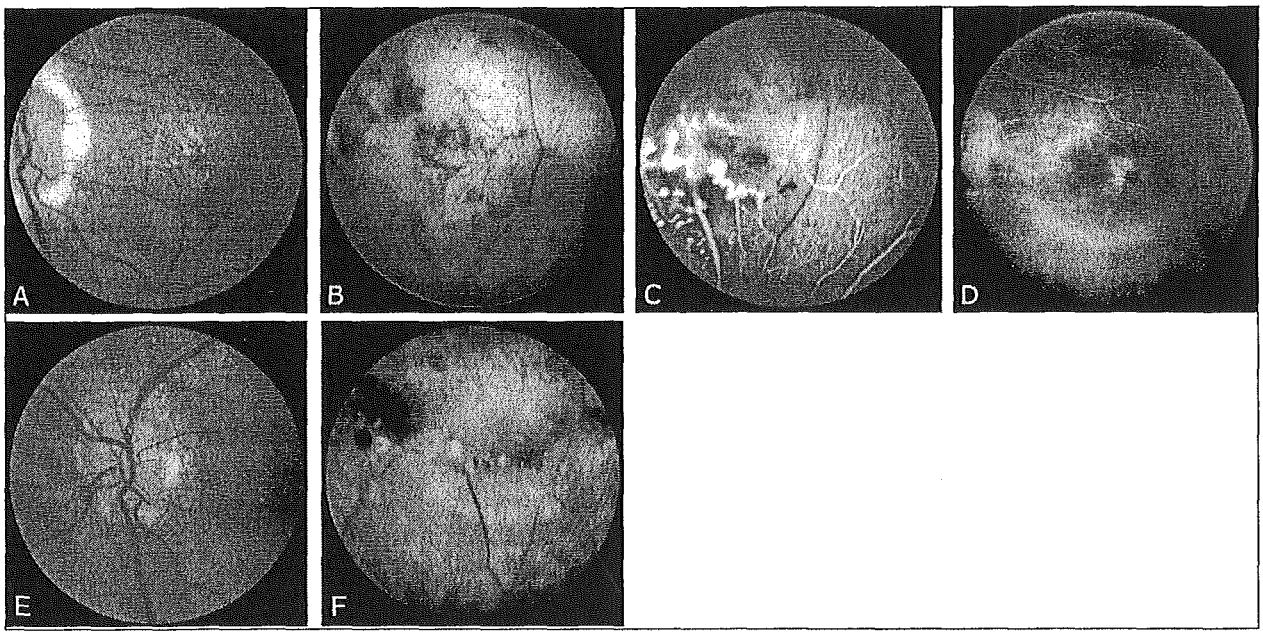

Fig. 1 - (A) *Fundus photograph at age 20 years showing peripapillary and macular exudates in the form of a macular star.* **(B)** *Peripheral exudates in the peripheral superior retina accompanied by hemorrhages and vascular telangiectases.* **(C)** *Fluorescein angiography showing capillary nonperfusion, telangiectases, and vascular malformations with hyperfluorescent aneurysmal dilatations.* **(D)** *In the end stages of the angiogram, cystoid macular edema accompanied by exudation in the peripapillary region and throughout the temporal inferior arcade can be seen. There were no untoward signs of exudation on the fluorescein angiography in the inferior fundus.* **(E, F)** *Fundus photography 6 months after the initial treatment with cryotherapy and argon laser.* **(E)** *Note the diminished exudates in the peripapillary and macular area.* **(F)** *Cicatricial chorioretinal scars in the superior quadrants with disappearance of the retinal exudation.*

exudative lesion was treated with cryotherapy and argon laser. Six months after the treatment, VA improved to 0.5, remaining stable during the ensuing year (Fig. 1, E, F). The patient was then lost to follow-up and, since VA remained stable, did not seek further medical help for the next 29 years.

Thirty years after the initial treatment, the patient returned to our department as he had a progressive loss of VA in his LE for about 1 month. The ophthalmic examination of the RE was normal, but VA of the LE had diminished to finger counting at 1 meter. Anterior segment biomicroscopy and IOP were normal. Fundus slit-lamp examination showed a retinal detachment in the inferior retina associated with marked peripapillary and macular exudation as well as typical peripheral areas with telangiectases and other vascular malformations compatible with stage 3A of Coats disease (Fig 2, A-D) (1). At that time, the retinal detachment was considered of serous origin, but due to the presence of an area of lattice degeneration with a possible small retinal dehiscence in the temporal detached retina, the differential diagnosis of a rhegmatogenous retinal detachment was retained as well.

Fluorescein angiography confirmed the presence of typical peripheral telangiectases as well as cystoid macular edema (Fig. 2E). The retinal detachment was treated with a silicone 240 encircling band (Labtician, Oakville, Ontario, Canada) placed under the 4 rectus muscles and fixed with a braided nonresorbable suture in all 4 quadrants. In addition, cryocoagulation and external drainage were performed, as the vascular malformations could not be treated otherwise due to the bullous detachment. Once the retina was attached peroperatively, cryotherapy could then be applied in the areas of vascular malformation. No retinal tear could be documented during surgery. At 2 months postoperatively, optical coherence tomography (OCT) did not show overt cystoid macular edema but only a diffuse thickening of the macula (Fig. 3A). During the 12 months after surgery the patient underwent 10 sessions of additio-

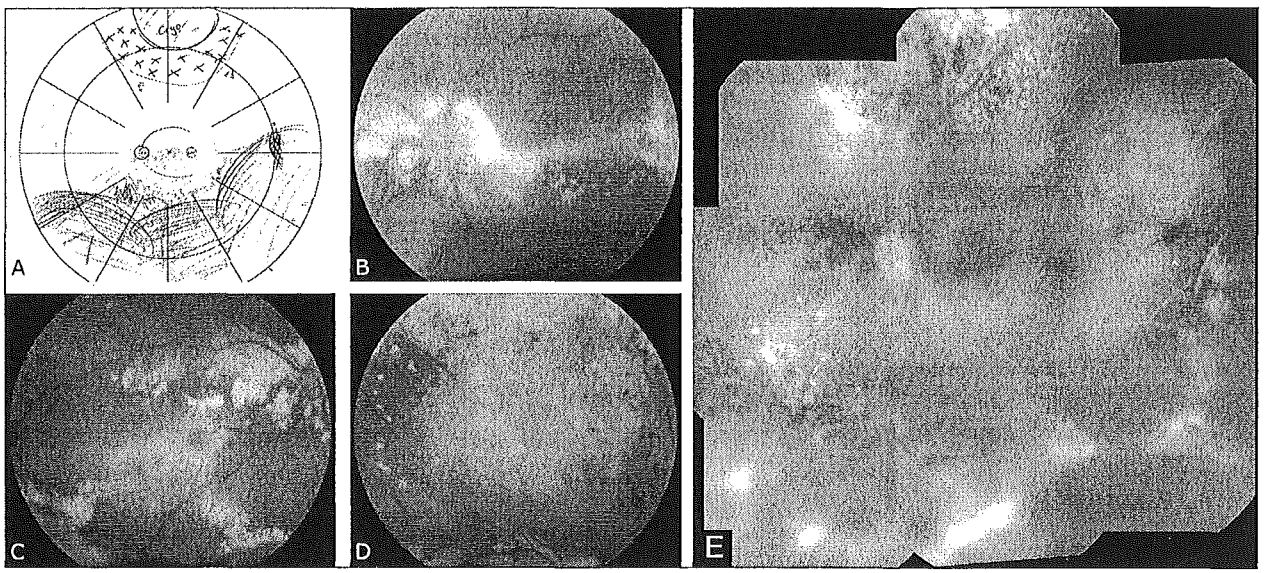

Fig. 2 - (A) *Pictorial diagram of the retinal changes at presentation 30 years after the first treatment. Note the large areas of exudation as well as the retinal detachment in the inferior retina.* **(B)** *Fundus photography showing new peripapillary and macular exudates with signs of retinal detachment inferiorly.* **(C)** *Massive intraretinal and subretinal lipidic exudates in the nasal retina with areas of telangiectasia in the periphery.* **(D)** *The chorioretinal scars in the superior quadrant are due to the initial treatment of cryotherapy and argon laser 30 years previously.* **(E)** *Fluorescein angiography showing the cicatricial peripheral superior retina without major leakage. The peripapillary and macular hypofluorescence corresponds to a blocking effect by the exudates. The nasal, inferior, and temporal hyperfluorescence corresponds to leakage from the typical telangiectases of Coats disease.*

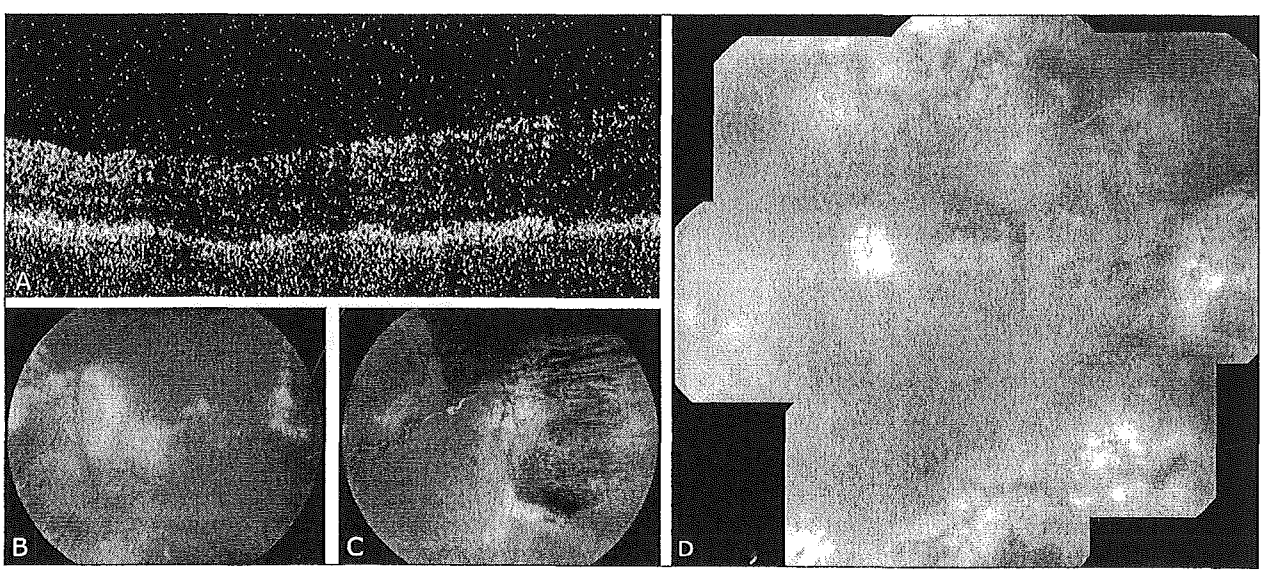

Fig. 3 - (A) *Optical coherence tomography of the foveolar area 2 months after retinal detachment surgery, which included external drainage of the subretinal fluid and cryocoagulation. Only diffuse macular edema can be seen.* **(B)** *Fundus photograph 12 months after the retinal detachment surgery and repeated laser application on the residual areas of telangiectases. Note the regression of the peripapillary and macular exudates.* **(C)** *Fundus photograph taken at the same time showing fibrosis both of the retina and retinal vessels in the temporal peripheral retina.* **(D)** *Composite images of the fluorescein angiography showing exudates in the nasal, inferior, and temporal retina. There is residual diffuse leakage in the macula although no frank cystoid edema can be seen. Note the laser scars in the periphery of the lower quadrants with a marked decrease of the leakage of the vascular malformations.*

nal argon laser treatment in the areas of persistent vascular malformations, which had caused continuing exudation. With time an almost complete attachment of the retina was achieved, which was associated with a minimal residual amount of retinal exudates (Fig. 3, B-D). Best-corrected VA at the end of the follow-up period was 0.1 (+2 –2.5 ×180°).

DISCUSSION

The present case is instructive as it depicts a previously poorly understood long-term evolution of Coats disease over a 30-year period. It is remarkable that even though a small telangiectatic retinal lesion was treated successfully, the disease could nevertheless reawaken and recur with force in previously unaffected retina several decades later. The mechanisms behind this late re-emergence of Coats disease are not well understood (2).

The first manifestation of the disease was successfully treated with simple cryotherapy and laser on the affected retinal areas. This combination treatment has remained the gold standard if there is little subretinal liquid (3). As the clinical manifestations of the second episode of the disease were complicated by a bullous inferior serous retinal detachment due to the marked exudation, drainage of the subretinal fluid and placement of an encircling band became necessary to reattach the retina to the retinal pigment epithelium, thus permitting subsequent cryotherapy or laser treatment.

In many cases it is essential to apply postsurgical argon laser in several sessions on areas of persisting vascular malformations. This can be performed even in a slightly detached retina, but it is advisable to employ energies of about 500 mW with a duration of 1000 milliseconds in order to coagulate the malformations completely. Recently, laser has also been used successfully with adjunct medical therapy such as intravitreal corticosteroids and other growth factor inhibitors (4). More invasive intravitreal surgery may become indispensable if persisting exudation triggers retinal detachment or the formation of important epiretinal membranes (5, 6).

In conclusion, the present case underlines the fact that it is possible for the disease to recur after several decades even if the initial lesions were treated successfully. The re-emergence of the disease can occur in previously unaffected areas of the retina. Coats disease must therefore be considered as a chronic disease, which necessitates a very long-term follow-up even in the absence of subjective visual loss (2, 7).

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