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Vitrectomy techniques in late-stage Coats'-like exudative retinal detachment

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Abstract. Retinal telangiectasia is the hallmark of Coats' disease. In the late stages, leakage from these abnormal vessels can result in a total, bullous exudative retinal detachment with cholesterol-laden subretinal fluid. Secondary angle-closure glaucoma may result in a blind and painful eye which may require enucleation or evisceration. Surgical reattachment of the retina and destruction of the retinal telangiectasia may preserve these eyes. We have found that vitrectomy, internal drainage of subretinal fluid and cholesterol, direct treatment of the retinal telangiectasia with intraocular diathermy and intravitreal gas or silicone oil injection are effective surgical techniques for salvaging these severely damaged eyes.

Introduction

Coats' described unilateral retinal telangiectasia with subretinal exudate, cholesterol and hemorrhage which occurred more commonly in males in their first two decades of life [1]. This description was later revised to exclude retinal hemorrhage [2–3] and to include bilaterality [4–6]. In the earlier stages of the disease externally applied diathermy [4, 7], laser photocoagulation or cryotherapy alone may be effective in destroying the retinal telangiectasia [7–11].

In the later stages, a bullous, exudative retinal detachment may result in a painful, secondary angle-closure glaucoma necessitating enucleation (1, 4, 6–7, 10, 11]. Treatment in the past has utilized external drainage of subretinal fluid and scleral buckling [7–11] in conjunction with diathermy, photocoagulation or cryotherapy.

We utilized vitrectomy, internal drainage of subretinal fluid and cholesterol, direct application of intraocular diathermy to the retinal telangiectasia and gas-fluid exchange or intraocular silicone oil to treat late-stage Coats'-like disease.

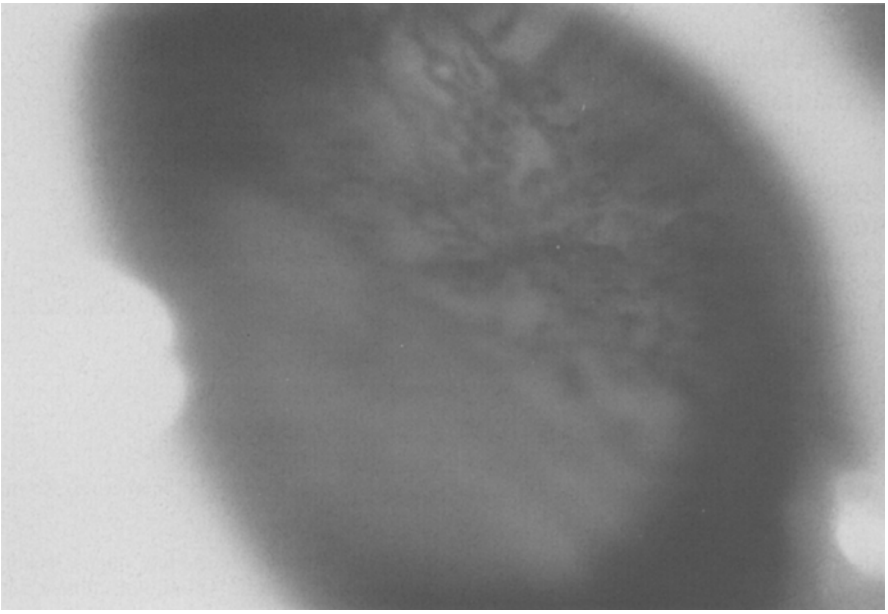

Fig. 1. Extensive area of retinal telangiectasia.

Materials and methods

Six patients treated surgically for extensive Coats'-like retinal telangiectasia (Fig. 1), subretinal cholesterol and bullous, exudative, total retinal detachments (Fig. 2) were reviewed retrospectively. Five of our cases were diagnosed with typical Coats' disease and one had a Coats'- like reaction due to juvenile retinoschisis (case 4). Five patients were male and one female. Their ages ranged from 2 months to 35 years. The follow-up period ranged from 3 months to 5 years.

All cases underwent only one operation: either a scleral buckling procedure alone or a vitrectomy combined with a scleral buckling procedure. No repeat operations except postoperative laser photocoagulation and cryotherapy (case 3) were performed in any eye.

All cases underwent external drainage of subretinal fluid and cholesterol, externally applied cryoablation of the retinal telangiectasia and scleral buckling.

In addition, four patients underwent vitrectomies (cases 2, 4, 5 & 6) and lensectomies were performed in two of these four cases (cases 5 and 6).

Only three (cases 4, 5 & 6) of the four patients who underwent vitrectomies had retinotomies for internal drainage of subretinal fluid and cholesterol with

Fig. 2. An exudative, bullous retinal detachment with subretinal cholesterol. The retina is displaced into a retrolental position by the accumulation of a large amount of subretinal fluid.

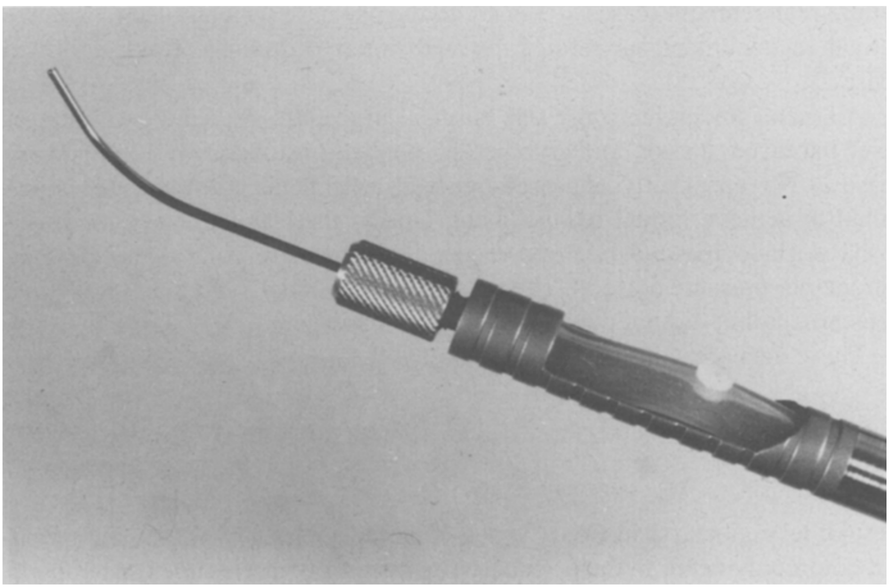

Fig. 3. Stainless-steel, curved, #20 extrusion cannula (Dutch Ophthalmic Research Corporation).

a tapered, silicone-tipped extrusion needle or curved, stainless-steel, #20 extrusion cannula[1] (Fig. 3).

Only two cases that underwent vitrectomies had intraocular diathermy applied directly to the retinal telangiectasia (cases 5 & 6).

Three cases underwent intravitreal gas-fluid exchanges with air, SF_6 or C_3F_8 (cases 2, 4 & 5). A five-year old child required intravitreal silicone oil in order to tamponade the retina after retinotomy because of the anticipated postoperative positioning difficulties (case 6).

Results

Four of the six cases in this study achieved retinal reattachment. Only one of the two cases which utilized only external drainage of subretinal exudate, cryoablation of the retinal telangiectasia and scleral buckling, achieved retinal reattachment (case 3). This case required multiple postoperative laser photocoagulation and cryotherapy treatments over a period of three months after the initial scleral buckling procedure in order to achieve retinal reattachment.

In contrast, three of the four patients who underwent vitrectomy and scleral buckling achieved retinal reattachment. In these successful cases, retinotomies with internal drainage of subretinal fluid and cholesterol, in addition to external drainage, cryoablation and scleral buckling, achieved retinal reattachment (cases 4, 5 & 6). In the one case which failed to achieve retinal reattachment, no retinotomy with internal drainage was performed (case 2).

All cases underwent only one surgical procedure with the exception of laser photocoagulation and cryotherapy applied postoperatively in one case (case 3). No repeat surgical procedures were attempted in the two cases which failed to achieve retinal reattachment. One of these failed cases continued to have glaucoma and required medical treatment to control the elevated intraocular pressure (case 2). The other failed case continued to have a retinal detachment, but did not develop glaucoma (case 1).

These data are summarized in Table 1.

Discussion

Retinal telangiectasia in Coats' disease can develop anywhere in the retina, but commonly occur in the periphery [6], especially in the superior-temporal

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Table 1.

Case number	1	2	3	4	5	6
Age	32 M	7 Y	2 M	35 Y	19 Y	5 Y
Sex	F	M	M	M	M	M
Preoperative Visual Acuity	LP	LP	LP	HM	LP	LP
Postoperative Visual Acuity	CF	LF	LP	LP	LP	20/300
External Drainage	X	X	X	X	K	X
External Cryotherapy	X	X	X	X	X	X
Scleral Buckle	X	X	X	X	X	X
Vitrectomy		X		X	X	X
Retinotomy & Internal Drainage				X	X	X
Intraocular Diathermy					X	X
Lensectomy					X	X
Intraocular Gas or Silicone Oil		SF_6		AIR	C_3F_8	SO
Retinal Attachment			X	X	X	X
Follow Up Period	6 M	3 M	6 M	8 M	5 Y	6 M

periphery [9]. Spontaneous regression may occur, but progression is more common [14].

Early in the disease, before the development of bullous retinal detachment, treatment of the telangiectasia with either laser photocoagulation or cryotherapy alone may prevent progression [7–11].

In the later stages of Coats' disease, the abnormal vascular permeability results in a bullous, exudative, retinal detachment with the accumulation of blood, cholesterol crystals, and lipid-filled macrophages in the subretinal space [1–4, 6–7,11–13]. The deposition of lipid in the subretinal space is usually proportional to the amount and duration of the retinal telangiectasia.

Secondary angle-closure glaucoma from iris neovascularization or gradual compression of the anterior chamber by a retrolental, bullous, exudative retinal detachment may ultimately result in a blind and painful eye necessitating enucleation [4, 6–7, 10–11].

Permanent reattachment of the retina and destruction of the retinal telangiectasia may help salvage these eyes.

External drainage has been traditionally employed to remove subretinal fluid and cholesterol in an attempt to reattach the retina [8–9, 11]. All of our cases underwent external drainage, but this maneuver alone was unsuccessful in reattaching the retina because accumulation of cholesterol deposits at the sclerotomy site frequently interrupted outflow. Multiple, often futile, attempts at external drainage were performed in order to drain residual subretinal fluid. In the two cases that employed external drainage alone (cases 1 & 3) only one (case 3) achieved retinal reattachment. Even when the fluid portion of the subretinal exudate could be drained externally, large accumulations of residual cholesterol deposits remained. These large plaques of crystalline cholesterol actually insulated the retinal telangiectasia from the freezing effects of externally applied cryoablation. In the case that successfully achieved retinal reattachment after using only external drainage and scleral buckling, multiple postoperative treatments with laser photocoagulation and cryotherapy were necessary to destroy the remaining retinal telangiectasia as the residual subretinal fluid slowly resorbed over a period of three months after the initial scleral buckling procedure (case 3). Large residual accumulations of subretinal cholesterol may also mechanically impede retinal reattachment even if the subretinal fluid portion of the exudate has been drained.

Vitrectomy is a powerful surgical technique in the treatment of late-stage Coats' disease. Vitrecomy was utilized in addition to scleral buckling techniques in four of our six cases (case 2, 4, 5 & 6). Only one of these cases failed to achieve reattachment of the retina (case 2). Although vitrectomy was performed in this failed case, no retinotomy and internal drainage of subretinal fluid and cholesterol was performed.

We have found that extraction of subretinal fluid and cholesterol can be best achieved by retinotomy and internal drainage and all eyes that underwent these procedures achieved retinal reattachment (case 4, 5 & 6). Internal drainage of subretinal fluid and cholesterol crystals through a retinotomy was determined to be a critical step in achieving permanent retinal reattachment. Two points must be emphasized when utilizing internal drainage in Coats'-like exudative retinal detachments:

1). The curved, stainless-steel, #20 extrusion cannula (Fig. 3) is recommended over the narrower bore, tapered, silicone-tipped extrusion needle. The narrower bore of the silicone-tipped extrusion needle became more easily clogged by larger cholesterol deposits. The curved, stainless-steel, #20 extrusion needle was particularly effective in sweeping the subretinal space and evacuating cholesterol accumulation far from the retinotomy site.

2). The retinotomy should be made in the superior retina, posterior to the equator. The retinotomy should be made much larger than the diameter of the extrusion needle in order to allow the intravitreal infusion fluid to flow back into the subretinal space and stir the suspended cholesterol crystals with eddy currents during internal drainage. If this stirring action is not achieved and only subretinal fluid evacuated, cholesterol deposits will coalesce into larger plaques that are difficult to remove. Very large crystalline cholesterol plaques occluded the tip of the extrusion needle and required mechanical extraction through a larger retinotomy opening (case 6).

We have found that vitrectomy techniques have other advantages. Compression of the vitreous body by a bullous, exudative retinal detachment squeezed vitreal humor out the vitreous body and pressed the vitreal collagen and posterior hyaloidal membranes upon the retinal surface creating epiretinal membranes [5]. Contraction of these membranes exerted tractional forces which prevented reattachment of the retina after drainage of subretinal fluid. Membrane peeling and excision may be necessary because epiretinal membranes can convert an exudative retinal detachment into a rhegmatogenous one by exerting tractional forces, especially during the evacuation of subretinal fluid when the retinal detachment is hydraulically pushed back into place. If the retinal tear is large, cholesterol crystals can escape into the vitreous body from the subretinal locus (case 5). The presence of cholesterol crystals in the vitreous body should alert one to the possibility of a combined exudative and rhegmatogenous retinal detachment, even if it is observed at the time of the preoperative examination.

Direct application of intraocular diathermy to the retinal telangiectasia was a very effective surgical technique for destroying the vascular lesions of Coats'-like disease (cases 5 & 6).

No extraordinary hemorrhagic or inflammatory response was noted in any case undergoing either scleral buckling or vitrectomy with or without intraocular diathermy.

Lensectomy may be necessary if cataracts or severe posterior synechiae limit visualization of the retina. Lensectomy may also be necessary to provide adequate space for intraocular surgical maneuvers if a bullous, total retinal detachment assumes a retrolenticular position, compresses the vitreous body, displaces the lens anteriorly and shallows the anterior chamber (cases 5 & 6). In most cases of late-stage Coats' disease, the loss of the lens will not adversely affect visual rehabilitation because of the limited visual potential in this commonly unilateral disease process. Our cases with successful retinal reattachment achieved postoperative visual acuity ranging from light perception to 20/300 (Table 1).

Certainly in the rarer bilateral cases of Coats' disease, every effort must be employed to preserve vision, but minimal visual recovery and ocular preservation are also important in the unilaterally affected young patient because enucleation or evisceration may lead to a lifetime of disfigurement. Vitrectomy, internal fluid drainage and directly applied intraocular diathermy to destroy retinal telangiectasia are powerful surgical techniques which are able to achieve an immediate and permanent retinal reattachment and salvage potentially lost eyes in late-stage Coats'-like disease.

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