RHEMATOGENOUS RETINAL DETACHMENT COMPLICATED BY SEVERE INTRAOCULAR INFLAMMATION, HYPOTONY, AND CHOROIDAL DETACHMENT

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INTRODUCTION

THIS REPORT CONCERNS AN UNUSUAL FORM OF PRIMARY (RHEGMATOGENOUS) retinal detachment which differs in many ways from the routine, uncomplicated case of detached retina encountered by the ophthalmologist. Unlike most cases of primary retinal detachment, patients with this syndrome have a severely inflamed, painful, red eye. The eye manifests a severe panuveitis, deepening of the anterior chamber, concentric folding and posterior bowing of the iris, posterior synechiae, iridophakodonesis, hypotony, and choroidal detachment, in addition to the detached retina. Retinal tears are present but may be difficult to identify. Massive periretinal proliferation is often present, manifested by star folds and vitreous traction bands. The surgical and visual prognosis is poor.

Such cases present problems in differential diagnosis and in management. They are, indeed, rhegmatogenous detachments and hence are potentially curable by surgery, but the correct diagnosis may be masked by the severe uveitis, small pupil, hazy media, and choroidal detachment. These cases may be confused with simple choroidal detachment following cataract surgery, the uveal effusion syndrome, secondary retinal detachment, or malignant melanoma of the choroid.

Once correctly diagnosed, initial therapeutic efforts must be directed toward reducing the severe inflammatory response and the choroidal detachment prior to surgery, as ill-timed surgery may precipitate phthisis bulbi in these severely inflamed eyes.

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HISTORICAL REVIEW

Beigelman[1] credits Schnaubel (1876) with the first description of this unusual symptom complex associated with retinal detachment. Schnaubel was following a highly myopic patient with retinal detachment and one day noted that the eye had suddenly become soft to finger palpation, the anterior chamber was much deeper than normal, and the iris was drawn far posteriorly. He postulated a spontaneous rupture of a posterior staphyloma, but this proved to be an erroneous diagnosis because the globe later regained its normal tension and appearance. According to Beigelman,[1] Leber was so impressed with the association of hypotony and retinal detachment that he proposed it as a separate distinct clinical entity.

Since Gonin initiated the "modern era" of retinal detachment surgery 50 years ago, only four significant papers on this peculiar variant of rhegmatogenous retinal detachment have appeared in the literature. Because of this paucity of published information, it seems appropriate to review these articles in some detail.

1. Gonin[2] recognized the occasional association of marked hypotony with retinal detachment, and commented on the presence of choroidal and ciliary body detachment in such cases. Severe uveitis and inflammation are not included in his description of such cases, however. He stressed the poor surgical prognosis and attributed it to a progressive disease of the vitreous body.

   In his paper, published in 1934, Gonin addresses the question, "Is the hypotony the cause of the retinal detachment, or does it result as a consequence of the detachment?" He argues that neither is the case; rather, both the retinal detachment and the hypotony are explained by the same process, ie, a progressive retraction of the vitreous body. Where the vitreous is firmly attached to the retina at the equator, vitreous retraction produces retinal holes and subsequent detachment of the retina. He believed that progressive vitreous retraction then caused the ciliary body and choroid to separate from the sclera, just as had the retina.

   Gonin's careful clinical observations convinced him that the hypotony did not cause the detached retina, as some authors believed at that time. He correctly noted that the hypotony was not pronounced at the onset of the detached retina but became apparent several days to several weeks after the detachment became manifest. Gonin observed that other oph-

thalmologists, including Lindner and Arruga, believed that the hypotony was caused by the absorption of subretinal fluid by the choroidal vasculature and that this process was enhanced when large retinal tears were present. Again, this was contrary to Gonin's clinical experience. He had observed cases of hypotony with retinal detachment in which retinal tears were very small and difficult to find, while other cases of detached retina with very large retinal tears did not exhibit hypotony.

Finally, Gonin realized that treatment with thermo-cautery was beneficial in some of these cases. He attributed the improvement to an adherence between the sclera and ciliary body produced by the thermo-cautery.

2. In a report from England in 1958, Graham[4] cited two cases of "unusual evolution of retinal detachment." He observed that such cases had been well described in the classic ophthalmic literature, but had received scant attention in recent years and thus were apt to be missed by clinicians.

In both of his reported cases, the disease began as a "routine" case of detached retina, and in each case a retinal tear was visualized ophthalmoscopically. After two weeks, in one case, and four days in the other, the clinical picture changed abruptly. Both patients developed aqueous flare, concentric iris folds, deepened anterior chamber, posterior synechiae, hazy vitreous, and detachment of the ciliary body and choroid. One eye was considered inoperable and the other eye was enucleated because malignant melanoma could not be ruled out. Pathologic findings in the enucleated eye included a generalized retinal detachment with retinal folding and a localized preretinitis (sic). The venous channels were moderately dilated and uveal congestion was present. The anterior choroid was detached, as was the ciliary body to the scleral spur. In other pathologic specimens of similar cases, the findings "have varied from massive round cell infiltration of the uvea to very minor round cell infiltration of the iris root, associated with uveal congestion."

Graham attributed the iridodonesis and backward displacement of the lens-iris diaphram to two factors: (1) detachment of the ciliary body with consequent slackening of the zonules, and (2) vitreous shrinkage and contraction. He states that the hypotony and uveal congestion must be assumed to be a con-

sequence of the pre-existing retinal detachment. He reasoned that vasodilatation caused by "an ill understood toxic effect in retinal detachment may be the basic cause of the whole clinical picture." He cites experimental evidence that vitreous in contact with retinal pigment epithelium gives rise to choroidal exudation, and he states that histamine has been found in the subretinal fluid in some cases of retinal detachment.

3. In the first important paper on this subject since the indirect ophthalmoscope became widely used in the diagnosis and management of detached retina in the United States, Gottlieb[5] presented 36 cases of combined choroidal and retinal detachment. These occurred in a consecutive series of 754 eyes with primary retinal detachment, a 4.5% incidence. He stressed the importance of iritis as an accompanying clinical sign and indicated that diagnostic confusion with other clinical entities might occur in such cases. He includes the following in his differential diagnosis of combined choroidal and retinal detachment: (1) choroidal detachment following cataract or glaucoma surgery; (2) massive uveal effusion; (3) choroidal and retinal detachment associated with scleritis; (4) malignant melanoma; (5) metastatic carcinoma to the choroid.

In one of his case reports, a two week delay ensued between the time of diagnosis of detached retina and hospitalization for surgery. In that interval, the patient developed a red eye, hypotony, folds in Descemet's membrane, uveitis, posterior synechiae, and iridodonesis, plus choroidal detachment. Treatment with topical corticosteroids resulted in resorption of the choroidal detachment and a rise in the intraocular pressure. The eye was subsequently operated on successfully. In another case report, the choroidal elevation was interpreted as a malignant melanoma and the patient was scheduled for a ^{32}P test; the choroidal elevation subsequently cleared and the retinal detachment became more extensive. The ^{32}P test was not performed. The eye eventually developed rubeosis iridis, ectropion uveae, and a heavily vascularized pupillary membrane and was enucleated. In his series, the surgical reattachment rate was 35%.

4. In a paper combining the experience of the Retina Service at the Massachusetts Eye and Ear Infirmary in Boston and the Mt. Sinai Medical Center in New York, Seelenfreund et al[6] reviewed 50 cases of choroidal detachment associated

with primary retinal detachment. This represents approximately 2% of cases of rhegmatogenous retinal detachments managed by the authors. The disease is more common in older patients and in those exposed to surgical trauma (58% had had prior cataract surgery). There was a high incidence of retinal pathology in the fellow eye; in eight cases (16%) a retinal detachment had occurred, and in four others (8%) retinal tears were present.

The surgical prognosis was poor, with only 52% successful reattachment in the 42 cases available for follow-up study. Similarly, the visual results were poor, with only four patients achieving 20/70 vision or better. Massive periretinal proliferation developed in 90% of the 20 eyes which were surgical failures and occurred in 36% of the total eyes in this series. This incidence of MPP is considerably higher than would be expected in a series of rhegmatogenous retinal detachments. Their data did suggest a slightly better prognosis if the choroidal detachment had resolved prior to retinal reattachment surgery.

The authors consider hypotony to be the major clinical feature of this entity and do not consider ocular inflammation in their discussion. Hypotony is said to be the essential element in the development of choroidal detachment, and acute hypotony in turn is related to increased permeability of the choroidal capillaries. They postulate the following sequence: "(1) occurrence of a retinal detachment in an eye with an underlying deficiency in choroidal circulation; (2) decreased aqueous production due to edema of the ciliary body; (3) acute hypotony; (4) dilation of choroidal arterioles, with transudation of protein-rich fluid into the choroid and the suprachoroidal spaces; and (5) further edema and detachment of the ciliary body, enhancing the process."

MATERIALS AND METHODS

Because of the striking clinical presentation of these eyes, the difficulty in preparing them for surgery, and the relatively poor surgical and visual results, these cases constitute a most difficult challenge to the retinal surgeon. I have observed 47 such cases over a 12 year period, and these 47 cases constitute the basis of this report.

For purposes of comparison, the records of 665 cases of uncomplicated retinal detachment managed by me since 1971 have been reviewed. Several striking differences exist between the type of retinal detachment herein described and an uncomplicated retinal detachment. These will be presented and discussed.

CLINICAL FINDINGS

Every eye in this series had a rhegmatogenous retinal detachment. The macula was detached in every case at the time of my initial examination.

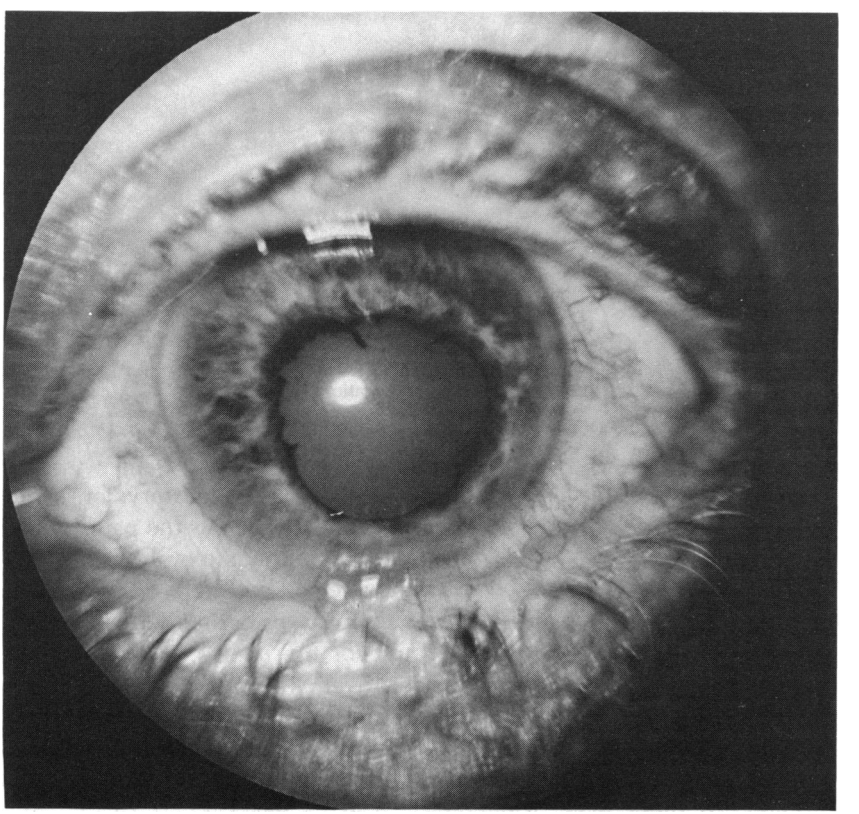

FIGURE 1
Right eye of patient with rhegmatogenous retinal detachment complicated by severe uveitis and choroidal detachment. The eye was painful and tender to the touch.

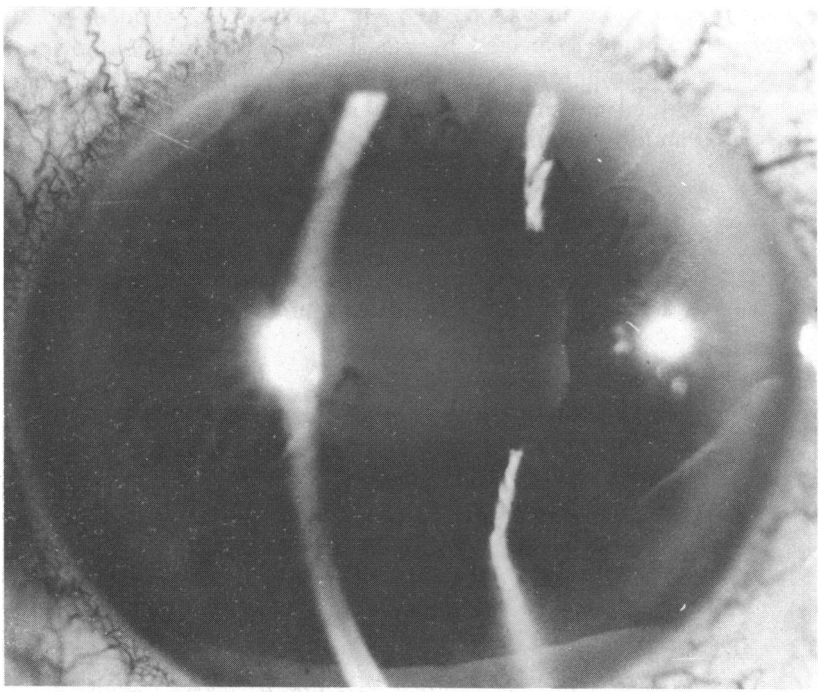

FIGURE 2
Severe anterior and posterior segment uveitis with retinal detachment. Note posterior synechiae formation.

In some patients the detachment had been present, by history, for several weeks prior to the onset of a red and painful eye. It was the pain, as well as the diminished vision, which prompted ophthalmic evaluation.

On examination, the involved eye is injected and tender to the touch (Fig 1). A severe panuveitis is present (Fig 2). There is no evidence of focal retinitis or choroiditis to account for the uveitis. The intraocular pressure, measured by applanation, is invariably low, varying from zero to 4 mm Hg. In one notable case, the eye was so hypotonous that the cornea was actually concave (Fig 3). In phakic eyes, the anterior chamber is abnormally deepened, with backward bowing of the iris in a concentric fashion. In these eyes, iridodonesis may be present, introducing the possible misdiagnosis of subluxation of the lens. Because of the intense anterior segment inflammation, posterior synechiae may form, making visualization of the fundus difficult.

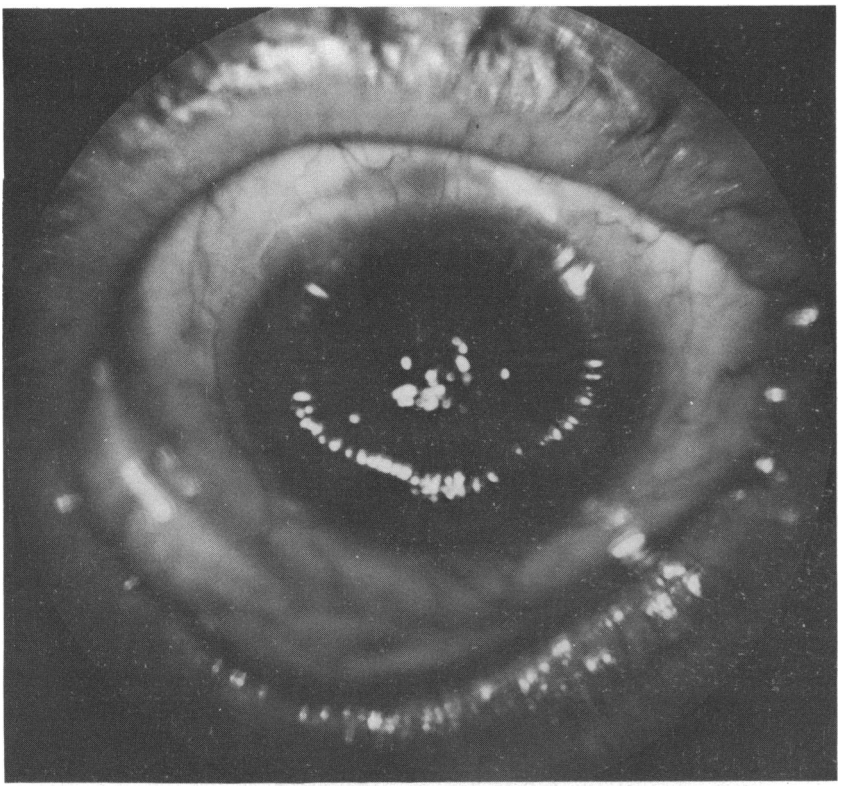

FIGURE 3
In this eye with rhegmatogenous retinal detachment and severe inflammation, the hypotony was so marked that the cornea was actually concave in configuration.

In most of these cases, a choroidal detachment of varying size and extent is present, in addition to the retinal detachment. The choroidal detachment may be extremely large and may totally dominate the funduscopic appearance.

Retinal tears are invariably present. Funduscopic examination is very difficult because of the severe uveitis, poor pupillary dilatation, posterior synechiae, and hazy media, and adequate examination is often impossible until the severe inflammatory response has subsided under treatment. In general, the retinal breaks are horseshoe tears. They tend to be large, are located quite far posteriorly, and have rolled, everted edges (Fig 4). Frequently, multiple horseshoe tears occur. Fixed retinal folds, star

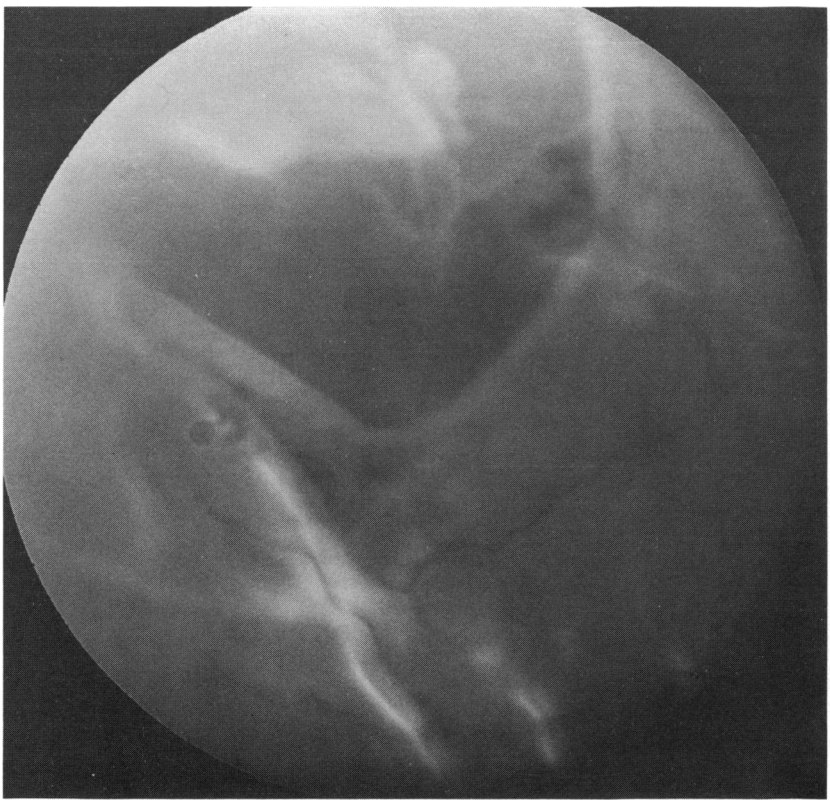

FIGURE 4
Huge, gaping horseshoe tear with everted edges, located posterior to equator. Massive periretinal proliferation is present.

folds, and meridional and circumferential retinal folds are commonly present, attesting to the presence of massive periretinal proliferation. The

TABLE I: CHARACTERISTIC SIGNS AND SYMPTOMS

1. Rhegmatogenous retinal detachment
2. Red, painful eye
3. Severe, generalized uveitis
4. Hypotony
5. Iridodonesis
6. Posterior bowing of the iris
7. Concentric folds in iris
8. Posterior synechiae
9. Choroidal detachment
10. Massive periretinal proliferation

features of this particular form of rhegmatogenous retinal detachment are summarized in Table I.

ASSOCIATED FINDINGS

Age

The age incidence, by decades, of the study group may be found in Table

TABLE II: AGE BY DECADE OF STUDY PATIENTS

DECADE	AGE (YR)	NO. OF PATIENTS	% OF PATIENTS
1	0–10	2	4
2	11–20	4	9
3	21–30	5	11
4	31–40	1	2
5	41–50	4	8
6	51–60	10	21
7	61–70	12	26
8	70+	9	19
Total		47	100

II. The majority of these cases occur in the 6th, 7th, and 8th decades of life, as do "uncomplicated" cases of retinal detachment (Table III).

Race

Of the 47 patients, 29 (62%) were white, but blacks accounted for an unexpected 38% of patients with this syndrome. In marked contrast, blacks constituted only 6% of the control population with uncomplicated retinal detachment.

TABLE III: AGE BY DECADE OF 665 UNCOMPLICATED CASES OF RHEGMATOGENOUS RETINAL DETACHMENT

DECADE	AGE (YR)	NO. OF PATIENTS	% OF PATIENTS
1	0–10	15	2
2	11–20	32	5
3	21–30	48	7
4	31–40	36	6
5	41–50	79	12
6	51–60	181	27
7	61–70	188	28
8	70+	86	13
Total		665	100

Sex

There were 30 men and 17 women in the study population (64% and 36%). In the control detachment population, men accounted for 58% and women 42% of the patients.

Eye

In 29 cases the right eye was the involved eye; in 18 cases the left eye was involved. There was one case of bilateral involvement.

Medical

In 11 cases, systemic hypertension was present. There were no other medical ailments common to this group of patients.

Status of Lens

In 25 patients (53%), the involved eye was phakic, while the eye was aphakic in 21 patients (45%). In one patient, the lens had been traumatically dislocated and an unsuccessful attempt had been made to remove it surgically. In the control series, 426 (64%) of the detachments occurred in phakic eyes, while 239 (36%) of the eyes were aphakic.

Status of Fellow Eye

In six cases the fellow eye was totally blind for the following reasons:

1. Inoperable retinal detachment (two eyes)
2. Traumatic cataract (one eye)
3. Enucleation after cataract operation (one eye)
4. Retrolental fibroplasia (one eye)
5. Trauma (one eye)

In two other cases, the fellow eye was legally blind (vision 20/400) because of a macular scar. In addition, significant ocular pathology was present in the fellow eye as follows: (In some patients more than one condition existed for each eye.)

1. Retinal detachment, successfully repaired surgically (two eyes)
2. Subclinical retinal detachment, successfully treated (two eyes)
3. Lattice degeneration without holes (three eyes)
4. High myopia (greater than -9.00) (five eyes)
5. Glaucoma (seven eyes)
6. Aphakia (14 eyes)
7. Immature cataracts (two eyes)

SURGICAL RESULTS

In five patients, the detachment was considered inoperable and surgery was never performed. In the other 42 patients, the eye was treated

preoperatively with topical, subtenon, and/or systemic corticosteroids, as well as with cycloplegics and mydriatics, in an effort to reduce the severe inflammation, dilate the pupil, and break the posterior synechiae. Although a few patients were operated on within a day or two of diagnosis, most patients were treated vigorously with cycloplegics and corticosteroids for two to three weeks and sometimes longer before surgery was performed.

Scleral buckling procedures were performed in the 42 patients who underwent surgery. Ophthalmoscopically monitored cryocoagulation was used to seal any existing retinal tears. An encircling silicone element was used with an additional solid, molded silicone piece sutured to the intact sclera as an exoplant when large retinal tears were encountered. Subretinal fluid was drained in most cases, and in some cases Ringer's solution or air was injected into the vitreous cavity to restore the volume of the globe and to tamponade large retinal breaks.

Surprisingly, intraoperative complications were encountered infrequently and no more often than in operations on "routine" cases of detached retina. However, the postoperative course was often stormy. One eye in which subretinal fluid was not drained developed a massive hyphema postoperatively and two other eyes quickly went into phthisis bulbi postoperatively, with a severe postoperative inflammation. In several cases, a very dense cataract developed after the detachment had been repaired.

Anatomic reattachment was achieved in 26 cases, while the retina failed to reattach in 16 cases; thus the anatomic success rate in the operated eyes is 62%. This compares with a 94% reattachment rate in the control series of uncomplicated retinal detachment cases. The reason for the comparatively poor reattachment rate is the pronounced tendency for massive periretinal proliferation in these eyes, perhaps stimulated by the severe intraocular inflammation.

VISUAL RESULTS

Even when the retina was anatomically reattached, the final visual acuity in these eyes was dismally poor. Of the 26 eyes achieving anatomic reattachment, only nine achieved 20/200 vision or better. There was a high incidence of macular disturbance in these eyes, including macular pucker, macular hole, or pseudo hole of the macula. In every eye, the macula had been detached preoperatively.

The preoperative and postoperative visual acuities in these 26 successfully treated patients is displayed graphically in Table IV. Table V gives

similar information for the larger, overall population of patients with retinal detachment; 94% of these patients (626 eyes) achieved anatomic reattachment following surgery.

TABLE IV: VISUAL ACUITY IN 26 "STUDY" PATIENTS WITH SUCCESSFUL ANATOMIC REATTACHMENT OF RETINA

POST-OP	20/20	20/25	20/30	20/40	20/50	20/60	20/70	20/80	20/100	20/200	20/400	CF	HM	LP	
LP															
HM										1		1	4	2	(8)
CF												3	2		(5)
20/400					1				1		1		1		(4)
20/200										1		2	1		(4)
20/100													1		(1)
20/80												1	1		(2)
20/70															
20/60												1	1		(2)
20/50															
20/40															
20/30															
20/25															
20/20															
PRE-OP					(1)				(1)	(2)	(1)	(8)	(11)	(2)	26

TABLE V: VISUAL ACUITY IN 626 PATIENTS WITH SUCCESSFUL REATTACHMENT OF RETINA IN UNCOMPLICATED RETINAL DETACHMENT

POST-OP	20/20	20/25	20/30	20/40	20/50	20/60	20/70	20/80	20/100	20/200	20/400	CF	HM	LP	
LP												2	0	1	(3)
HM											1	4	7	3	(15)
CF											1	8	11		(20)
20/400										2	9	12	12	4	(39)
20/200		1			3				1	7	7	18	28	7	(72)
20/100									3	4	4	6	12	1	(30)
20/80								3	4	3	3	7	11		(31)
20/70					4	1	2	2	2	4	3	10	11	3	(42)
20/60				1	2	2		4	3	7		11	12	1	(43)
20/50		1	2		4	2	3	2	2	5	6	13	21	2	(63)
20/40		1	3	18	1		7	4	3	12	3	17	19	2	(90)
20/30		2	20	16	7	9	2	1	2	7	4	10	15	1	(96)
20/25	4	9	6	2	2	3		1	1	3		3	5		(39)
20/20	24	5	3	3	1	2				1	1	2	1		(43)
PRE-OP	(28)	(19)	(34)	(40)	(24)	(19)	(14)	(17)	(21)	(55)	(42)	(123)	(165)	(25)	626

DISCUSSION

Every eye afflicted with a rhegmatogenous retinal detachment manifests some degree of intraocular inflammation, albeit small. Every ophthalmologist is accustomed to seeing a few cells in the anterior chamber and vitreous in such cases, along with cells and debris in the vitreous cavity. Similarly, the intraocular pressure in an eye with retinal detachment is usually lower than in the fellow eye, by 2 to 3 mm Hg. This is due to diminished aqueous secretion by the ciliary body.[7-10] In the cases under discussion, there is a pronounced exaggeration of these tendencies toward uveitis and inflammation, and the hallmark of this syndrome is severe intraocular inflammation and marked hypotony.

It is my belief that this unusual ocular syndrome is precipitated by a rhegmatogenous retinal detachment as the causal event. As in most cases of rhegmatogenous retinal detachment, a retinal tear forms idiopathically and a detached retina ensues. For reasons as yet unknown, the eye then exhibits a marked inflammatory response to the detached retina, leading in turn to increased vascular permeability, leakage of fluid and protein into the extracellular spaces, hyperemia of the choroid and ciliary body, eventual ciliary body and choroidal detachment, and subsequent hypotony (Fig 5).

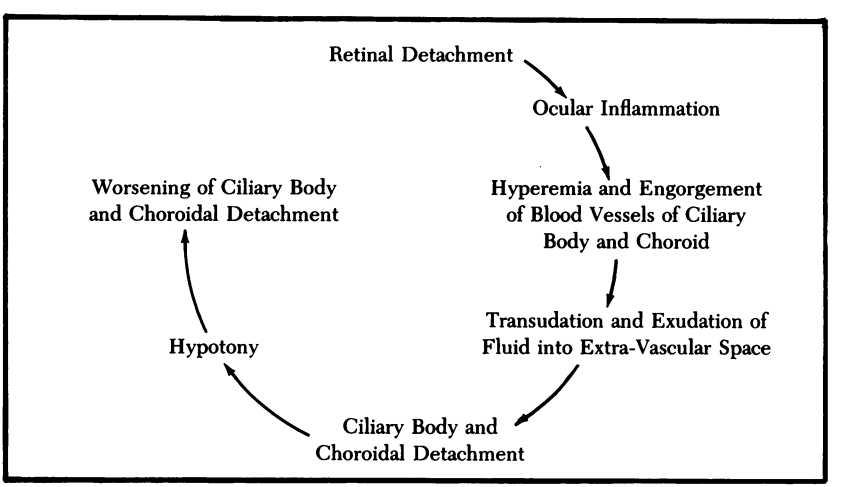

FIGURE 5
Postulated sequence of events leading to choroidal detachment and hypotony.

This sequence of events is in marked contrast to postinflammatory retinal detachment,[11] in which the initial eye disease is a severe uveitis. A variety of disease entities, including toxoplasmosis, pars planitis, or toxocariasis may be responsible for the ocular inflammation. In these cases, the severe uveitis and vitritis causes the formation of vitreous bands which then exert traction on the retina. A retinal tear forms as a result of this traction, and a retinal detachment ensues. In postinflammatory retinal detachment, then, ocular inflammation eventually causes retinal hole formation and retinal detachment. In the cases I am describing, this sequence of events is reversed.

Clinical observation suggests that the retinal detachment itself in some way initiates the sequence of events which culminates in inflammation and hypotony. In none of these patients was there a prior history of ocular inflammatory disease, nor were there signs of this on ocular examination. Cataract extraction had been performed in 21 patients, but in most instances the eye had healed from that surgery and vision had been restored until the onset of the detached retina. Most of these patients presented with the fully developed syndrome of detached retina, severe uveitis, hypotony and choroidal detachment when first examined by me. However, in two instances, the inflammatory component was observed to develop after the initial examination, while arrangements were being made to admit the patient for surgery. Gonin,[2] Graham,[4] Gottlieb,[5] and Seelenfreund[6] all report similar experiences.

DIFFERENTIAL DIAGNOSIS

It is not surprising that mistaken diagnoses are often made when dealing with this clinical entity. The most obvious feature, severe generalized inflammation, leads to the diagnosis of uveitis, and the detachment may be completely overlooked. If the detached retina is observed, it may be considered an inflammatory, or secondary, retinal detachment such as is seen with scleritis,[12] or Harada's disease.[13] The condition must also be differentiated from the uveal effusion syndrome which is an inflammatory condition characterized by choroidal separation and secondary retinal detachment.[14-16] The distinction is important because a rhegmatogenous retinal detachment is potentially repairable surgically, whereas uveal effusion is not. The iridodonesis may be mistaken for subluxation or dislocation of the lens. The marked hypotony may lead the ophthalmologist mistakenly to suspect a ruptured globe, as it did Schnaubel. If the eye is aphakic one might suspect a wound leak[17] or an epithelial downgrowth with fistulization of the cataract wound.[18] One of our patients did indeed have an epithelial downgrowth in addition to the detached retina.

The most serious misdiagnosis in such cases is malignant melanoma of the choroid. Unfortunately, several such eyes have been enucleated because this distinction was not made.[4,19] These enucleated eyes constitute the only pathologic specimens available for study in this entity. The clinical resemblance of this syndrome to melanoma may tax the most astute clinician. Both entities exhibit an elevation of the choroid in one or more quadrants, and eyes containing melanoma frequently have retinal detachment and may even have visible retinal holes.[20] Although choroidal detachment usually shows increased light transmission on transillumination, and melanomas usually block transmission of light,[21] this is not universally true and may be misleading.[22,23] The clinical course is helpful in making this differentiation, since a choroidal detachment should improve with time and/or treatment, whereas a choroidal tumor will either remain stable or slowly increase in size. The radioactive phosphorus uptake test has not been utilized in any of these cases.

MANAGEMENT

While immediate surgery is advocated in uncomplicated rhegmatogenous retinal detachment, it is contraindicated in the complicated cases under discussion. In this group of patients, the eye must first be treated with corticosteroids and cycloplegics in order to control the inflammation and clear the choroidal detachment prior to surgery. The presence of a choroidal detachment makes accurate localization of a retinal tear extremely difficult during the operation. In addition, Seelenfreund et al[6] have emphasized the uncertainty of adequate diathermy application to the swollen, edematous choroid, and he also stresses the difficulty in performing a lamellar scleral dissection in the boggy sclera of such eyes if one utilizes the technique of lamellar scleral resection. The risk of hemorrhage when draining subretinal fluid is heightened when one punctures the edematous, swollen, highly vascular choroidal tissue. Thus, it is advisable to reduce the inflammation, raise the intraocular pressure, and eliminate the choroidal detachment before subjecting these eyes to surgery. To achieve the desired condition in our patients, hourly applications of fortified corticosteroid drops topically, and subtenons injections of corticosteroids are used, along with mydriatics and cycloplegics. If the eye shows poor response to these measures, systemic corticosteroids are then added to the regimen. Surgery is usually delayed two to three weeks until the eye begins to show improvement.

With regard to surgical technique, we prefer the use of ophthalmoscopically monitored cryocoagulation to achieve chorioretinal scarring around the retinal holes. In this way, the surgeon is certain that the hole is

adequately treated, and this eliminates the uncertainty of unmonitored diathermy application to an undermined scleral bed. We also suture the molded silicone piece and encircling element to full thickness sclera, again eliminating the risk of dissecting the edematous sclera. Because localization of the hole may be difficult and uncertain if the choroidal detachment has not completely subsided, a scleral exoplant can easily be moved merely by repositioning the sutures. Drainage of subretinal fluid is mandatory in these cases. We agree with Freeman[24] that diathermy application to the choroid prior to perforation is helpful in avoiding complications at drainage in these most difficult cases.

DISTINGUISHING CHARACTERISTICS

There are a number of important differences between this type of retinal detachment and the "uncomplicated" rhegmatogenous detached retina usually encountered. In the latter group, the patient's only symptom is painless loss of vision when the detachment develops; in marked contrast is the patient who experiences a painful, red eye in addition to poor vision. Reparative surgery is undertaken as quickly as possible in the "routine" case of detached retina, whereas surgery must be postponed until the choroidal detachment subsides and the inflammation has lessened in the latter group. Rhegmatogenous retinal detachments are rare in blacks, accounting for only 6% in my personal series of retinal detachment, and 4.6% in another larger series.[11] Blacks, however, account for 38% of the cases under discussion. The reasons for the relative rarity of retinal detachment in blacks in general, and the propensity for blacks to develop inflammation, hypotony, and choroidal detachment when they do develop detached retina, are unknown.

The very high incidence of disease in the fellow eye in this group of patients must be stressed. In six patients the fellow eye was totally blind, and two others were legally blind in the fellow eye. Potentially blinding retinal disease was present in seven other fellow eyes. Glaucoma was also prevalent. In view of the poor surgical and visual prognosis in eyes with this type of detachment; the fellow eye deserves special attention. We recommend periodic examinations at six month intervals, and we instruct the patient to report any suspicious symptoms in the fellow eye at once.

The prognosis for successful anatomic reattachment is considerably poorer with this type of retinal detachment than for the "uncomplicated" retinal detachment: 62% versus 94%. Likewise, Table IV documents the very poor visual acuities achieved in the 26 patients with successful reattachments. These unfavorable visual results are due to an uncom-

monly high incidence of massive periretinal proliferation[25] in these patients. The ophthalmologist must recognize this entity to enable him to advise his patients intelligently concerning reparative surgery.

PATHOLOGY AND PATHOGENESIS

There are few specimens of this disorder available for pathologic study. One eye, mistakenly removed because melanoma was suspected clinically, has been examined in the pathology laboratory and subsequently reported.[19] Histologic features of this eye included: (1) deep anterior chamber due to retrodisplacement of the iris, (2) iris hyperemia, (3) detachment of the ciliary body and choroid, (4) detached retina, (5) marked dilatation of the choroidal vessels, and (6) small, inconspicuous foci of round cell infiltrates scattered throughout the uveal tract. These pathologic changes are non-specific and offer no clues as to the pathogenesis of this unusual condition.

Several excellent studies have been made of ocular hypotony and choroidal detachment.[22,26-29] Capper and Leopold[27] stressed the equilibrium maintained in the normal eye by balancing those factors which force fluid into the extravascular space (capillary arteriolar and venous pressure), with those factors which keep fluid within the vascular tree (intraocular pressure and colloid osmotic pressure). In our cases, clearly, this delicate equilibrium is upset. It is probable that the severe inflammation adversely affects capillary permeability, causing transudation and exudation of fluid into the extravascular space in the choroid and ciliary body. When the ciliary body detaches, aqueous secretion ceases, favoring further exudation of fluid out of the vessels into the tissue spaces. This concept is illustrated in Figure 5. This vicious cycle is broken only by decreasing the inflammatory reaction in the eye.

This missing link in this sequence of events is the trigger mechanism which incites the inflammatory response in the first place. All clinical evidence points to the detachment itself, but it is unclear how this is mediated. Kaufman and Podos[30] state that "vitreous has an irritative and degenerative effect on the choroid when they are brought into contact, and subretinal fluid, when injected into the eye, causes a uveitis." Dobbie[3] has suggested that a histamine-like substance might be elaborated from the mast cells in the uvea, and Graham[4] notes that histamine has been demonstrated in the subretinal fluid in some eyes. Further studies on the histochemical and immunological properties of the retina, choroid, and subretinal fluid will be needed to elucidate the mechanism of disease in these cases.

SUMMARY

An unusual type of rhegmatogenous retinal detachment is described and compared with a control group of patients with detached retina. Features of the condition, in addition to retinal detachment, include severe anterior and posterior uveitis, choroidal detachment, hypotony, deepened anterior chamber, posterior synechiae, iridophakodonesis, and a poor surgical and visual prognosis due to massive periretinal proliferation. The disease occurs in a disproportionately high ratio in blacks.

These cases may be mistaken for uveitis, Harada's syndrome, the uveal effusion syndrome, dislocated lens, or malignant melanoma.

It is postulated that the detached retina initiates a series of exaggerated pathophysiological changes in the eye, with the severe inflammation leading to choroidal detachment and hypotony. Such eyes are inoperable until the inflammation, hypotony, and choroidal detachment are reversed with corticosteroid treatment. The surgical and visual prognosis is very poor.

REFERENCES

1. Beigelman MN: Acute hypotony in retinal detachment. *Arch Ophthalmol* 1:463-467, 1929.
2. Gonin J: L'Hypotonie oculaire dans ses relations avec le decollement retinien. *Arch d'Ophtalmologic* 51:426-430, 1934.
3. Dobbie JG: Schepens CL, Regan CDJ (eds): *Controversial Aspects of the Management of Retinal Detachment*. Boston, Little Brown & Co, 1965, pp 73-74.
4. Graham PA: Unusual evolution of retinal detachments. *Trans Ophthalmol Soc UK* 78:359-371, 1958.
5. Gottlieb F: Combined choroidal and retinal detachment. *Arch Ophthalmol* 88:481-486, 1972.
6. Seelenfreund MH, Kraushar MS, Schepens CL, et al: Choroidal detachment associated with primary retinal detachment. *Arch Ophthalmol* 91:254-258,1974.
7. Regan CDJ, Rousseau AP: The intraocular dynamics of eyes with retinal detachment. *Am J Ophthalmol* 61:696-702, 1966.
8. Dobbie JG: A study of the intraocular fluid dynamics in retinal detachment. *Arch Ophthalmol* 69:159-164, 1963.
9. Smith JL: Retinal detachment and glaucoma. *Trans Am Acad Ophthalmol Otolaryngol* 67:726-732, 1963.
10. Sebestyen JG, Schepens CL, Rosenthal ML: Retinal detachment and glaucoma. *Arch Ophthalmol* 67:736-745, 1962.
11. Hagler WS, Jarrett WH, Chang MM: Rhegmatogenous retinal detachment following chorioretinal inflammatory disease. *Am J Ophthalmol* 86:373-379, 1978.
12. Sears ML: Choroidal and retinal detachments associated with scleritis. *Am J Ophthalmol* 58:764-766, 1964.
13. Duke-Elder S: Diseases of the uveal tract, in *System of Ophthalmology*. London, Henry Kimpton, 1966, vol 9, pp 373-383.
14. Schepens CL, Brockhurst RJ: Uveal effusion: clinical picture. *Arch Ophthalmol* 70:189-201, 1963.

15. McDonald PR, de la Paz V, Sarin LK: Non-rhegmatogenous retinal separation associated with choroidal detachment. *Trans Am Ophthalmol Soc* 62:226-247, 1964.
16. Rosen E, Lyne A: Uveal effusions. *Am J Ophthalmol* 65:509-518, 1968.
17. Chandler PA, Maumenee AE: A major cause of hypotony. *Am J Ophthalmol* 52:609-618, 1961.
18. Calhoun FP Jr: The clinical recognition and treatment of epithelization of the anterior chamber following cataract extraction. *Trans Am Ophthalmol Soc* 47:478, 1949.
19. Klien BA: Concerning conditions simulating an intraocular tumor. *Am J Ophthalmol* 20:812-819, 1937.
20. Robertson DM, Curtin VT: Rhegmatogenous retinal detachment and choroidal melanoma. *Am J Ophthalmol* 72:351-355, 1971.
21. Reese AB: *Tumors of the Eye*. New York, Harper & Row Publishers Inc, 1963, pp 269-271.
22. Brav SS: Serous choroidal detachment. *Surv Ophthalmol* 6:395-415, 1961.
23. Hagler WS, Jarrett WH, Humphrey WT: The radioactive phosphorus uptake test in diagnosis of uveal melanoma. *Arch Ophthalmol* 83:548-577, 1970.
24. Freeman HM, Schepens CL: Innovations in the technique for drainage of subretinal fluid: transillumination and choroidal diathermy. *Trans Am Acad Ophthalmol Otolaryngol* 78:829-836, 1974.
25. Machemer R, Laqua H: Pigment epithelium proliferation in retinal detachment: massive periretinal proliferation. *Am J Ophthalmol* 80:1-23, 1975.
26. Verhoeff FH, Waite JH: Separation of the choroid, with report of a spontaneous case. *Arch Ophthalmol* 55:38-53, 1926.
27. Capper SA, Leopold IH: Mechanism of serous choroidal detachment: a review and experimental study. *Arch Ophthalmol* 55:101-113, 1956.
28. Swan KC, Christensen L, Weisel JT: Choroidal detachment in the surgical treatment of retinal separation. *Arch Ophthalmol* 55:240-245, 1956.
29. Heath P: Ocular hypotony. *Trans Am Acad Ophthalmol Otolaryngol* 52:613-621, 1948.
30. Kaufman PL, Podos SM: Subretinal fluid butyrylcholinesterase. *Am J Ophthalmol* 75:627-636, 1973.