PSEUDOINFLAMMATORY MACULAR DYSTROPHY*

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INTRODUCTION

IN 1949 SORSBY AND HIS ASSOCIATES DESCRIBED FIVE FAMILIES WITH AN EXUDATIVE and hemorrhagic maculopathy inherited in an autosomal dominant manner.[1] Since the late stages of this disorder resembled a postinflammatory process, the term pseudoinflammatory macular dystrophy has been used.[2] Two members of one pedigree had histologic studies performed which demonstrated dehiscences in Bruch's membrane, choroidal subretinal neovascularization, and glial cicatrization of the outer retinal layers.[3] However, in this family an autosomal dominant inheritance was not clearly established and the advanced age of these patients at the time of the histologic studies cast some doubt as to the direct cause-effect relationship.

This paper reports three generations of a family of Spanish descent with presumed pseudoinflammatory macular dystrophy in whom eight of eleven members had decreased vision occurring between ages 25 to 40.

METHODS

Four of the affected individuals had ocular examinations, two at the New York University Medical Center and two at the Barraquer Eye Clinic in Barcelona. Four individuals who were not examined had a bilateral decrease in vision in their late 20's or early 30's (Fig. 1).

The two patients examined at the New York University Medical Center underwent complete ophthalmological examination including fluorescein angiography, visual field examination, color vision testing, visual sensitivity testing, electroretinography (ERG), and electro-oculography

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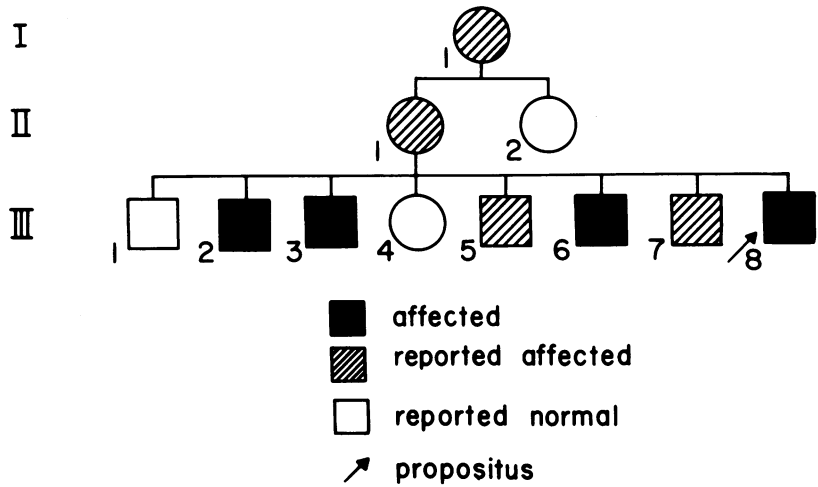

FIGURE 1
Pedigree of family with pseudoinflammatory macular dystrophy.

(EOG). The methods utilized in this laboratory are fully outlined in previous papers.[4, 5]

CASE PRESENTATIONS

CASE 1
(111-8) Propositus.
This 30-year-old man complained of decreasing vision in the right eye of six weeks duration. He had no previous ocular complaints and his general health was excellent. At his initial examination the following studies were performed.

Vision
RE: 6/30 (20/100) LE: 6/9 (20/30)

Fundus
RE: Hemorrhagic detachment of the retinal pigment epithelium (RPE) and the neurosensory retina in the macular area (1½ DD in size). LE: Small yellowish membrane just superonasal to the macula. In both eyes the optic nerves, retinal vessels, and retinal periphery were normal. (Figure 2a & b)

Fluorescein Angiography
RE: The choroidal fluorescence in the macula was obscured by the overlying hemorrhage (Figures 3 & 4). Within the hemorrhage was an area of subretinal neovascularization which leaked dye and partially covered the hemorrhage in the late phase. LE: A small subretinal neovascular net was seen just supero-nasal to

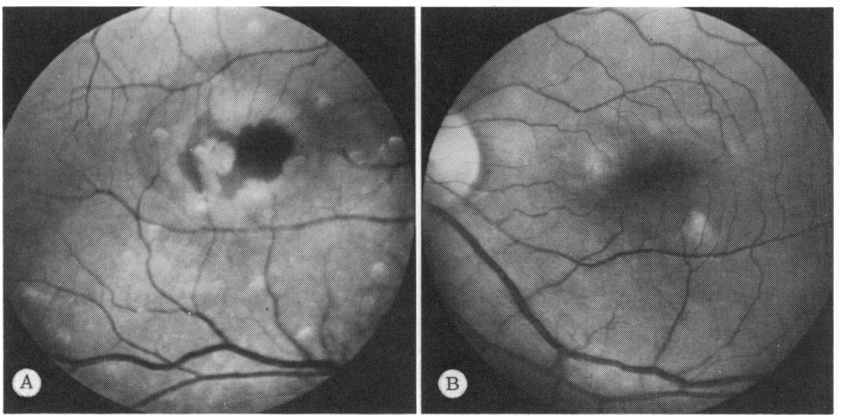

FIGURE 2
Case 1 A: The right eye has a hemorrhagic detachment of the retinal pigment epithelium and the neurosensory retina in the macula. B: The left eye has a small yellowish membrane just superonasal to the macula.

the avascular zone of the fovea. In the late stages, leakage of dye extended to the edge of the avascular zone of the fovea. The neovascular net was approximately 600μ from the nasal edge of the avascular zone of the fovea.

Electroretinogram
Normal photopic and scotopic responses.

Electro-oculogram
Normal light rise (230% O.U.)

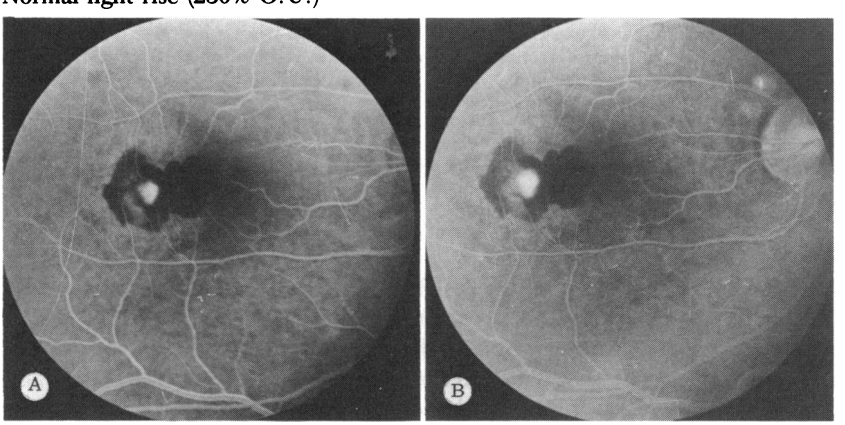

FIGURE 3
Case 1 Fluorescein angiography, right eye. The choroidal fluorescence is blocked by the overlying hemorrhage in the macula. Within this area, a neovascular network is present (Fig. 3A) which leaks dye into the subretinal space (Fig. 3B).

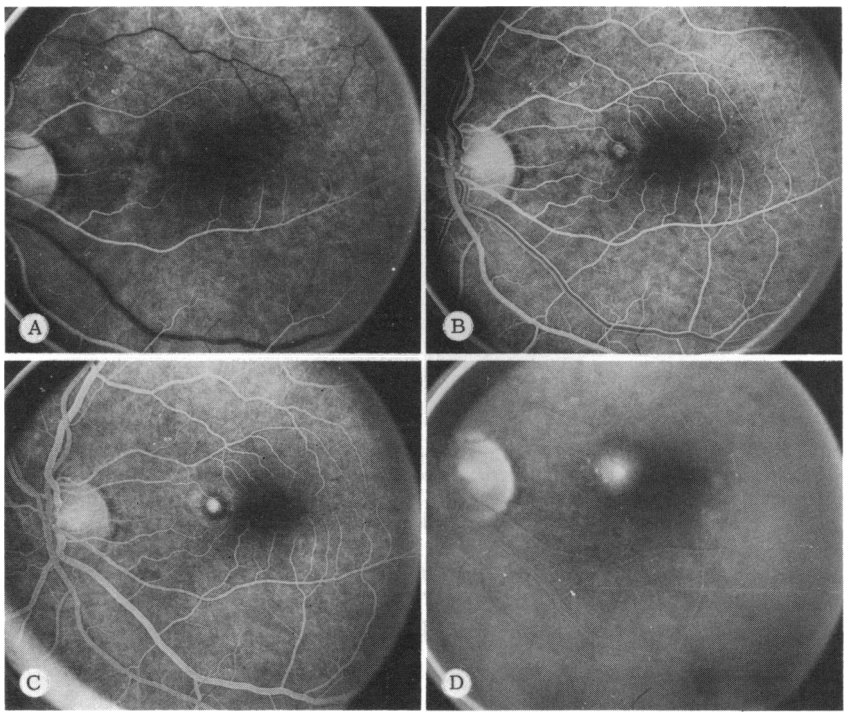

FIGURE 4

Case 1 Fluorescein angiography, left eye. A network of choroidal subretinal neovascularization is apparent in the early phase (Fig. 4A). The network is approximatley 600u from the nasal edge of the foveal avascular zone (Fig. 4B). These vessels leak dye (Fig. 4C) and an overlying sensory detachment is appreciated in the late phase (Fig. 4D).

Color Vision

AOHRR - mild R-G and B-Y defect. Farnsworth Panel D-15 - nondiagnostic color confusion.

Course

Over the two months following the initial examination there was a further decrease in the vision in the left eye to 6/15 (20/50). In view of the eccentric location of the subretinal net in this eye and because of the changes in the fellow eye he was treated with argon laser photocoagulation to the affected area. The following treatment parameters were utilized: spot size 100μ, time 0.5 sec., intensity 250 mw, number of applications 24. The treated area showed blanching indicative of a heavy burn intensity. Four weeks after treatment the vision dropped precipitously to 6/120 (20/400) in this eye and a large hemorrhagic detachment of the RPE and neurosensory retina involved the entire macula. Fluorescein angiography demon-

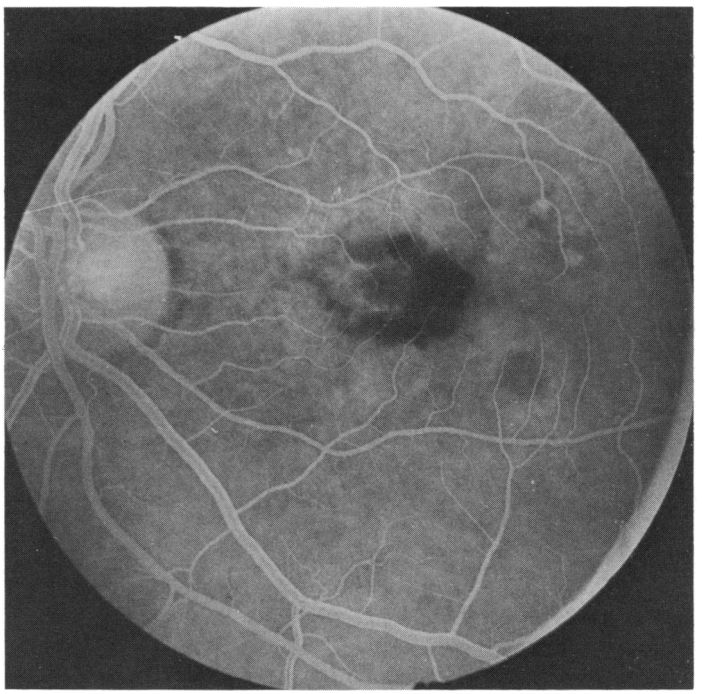

FIGURE 5
Case 1 Fluorescein angiography, left eye, following argon laser photocoagulation. The subretinal choroidal network has extended into the foveal avascular zone and is associated with a macular hemorrhage.

strated extension of the subretinal neovascularization temporally to include the fovea (Figure 5).

This patient was followed for two years and showed continued extensive hemorrhage of the posterior pole in each eye associated with a mild exudative response. At the present time the hemorrhages have absorbed and the patient exhibits bilateral glial cicatrization with visual acuity of 6/240 (20/600) RE and 6/120 (20/300) LE (Figure 6).

Follow-up psychophysical and electrophysiological tests confirmed that this was a local or geographic disease and that the peripheral retina was fuctionally normal.

CASE 2
(111-2)
The older brother of the above patient (Case 1) gave a history of poor vision in the right eye at age 35 and in the left eye two years later. Chronic simple glaucoma was diagnosed at age 38 and his intraocular pressures have been well-controlled

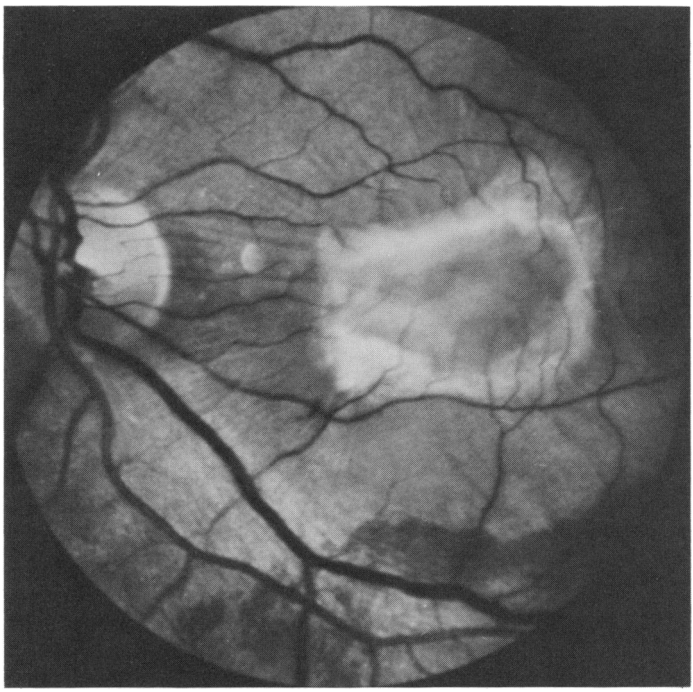

FIGURE 6
Case 1 Cicatricial stage, left eye. One year after laser coagulation the end result is glial scar formation.

on 4% pilocarpine. His general health was excellent. We first examined this man at age 44.

Vision
RE: 6/120 (20/400) LE: 6/15 (20/50)

Fundus
The macula in each eye showed gliotic changes associated with pigment accumulation and pigment epithelial atrophy (Figure 7). The right eye was more extensively involved. The optic nerve, retinal vessels and retinal periphery were normal.

Fluorescein angiography
The pigmentary accumulation and pigmentary atrophy resulted in the expected hypo- and hyperfluorescence respectively. There was no evidence of subretinal neovascularization or dye leakage (Figure 8).

Electroretinogram
Normal photopic and scotopic responses.

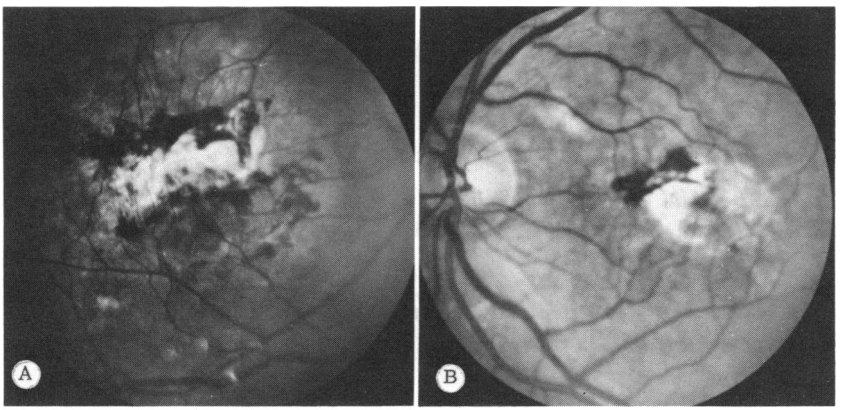

FIGURE 7
Case 2 The macula of each eye shows pigment accumulation, pigment atrophy, and gliosis.

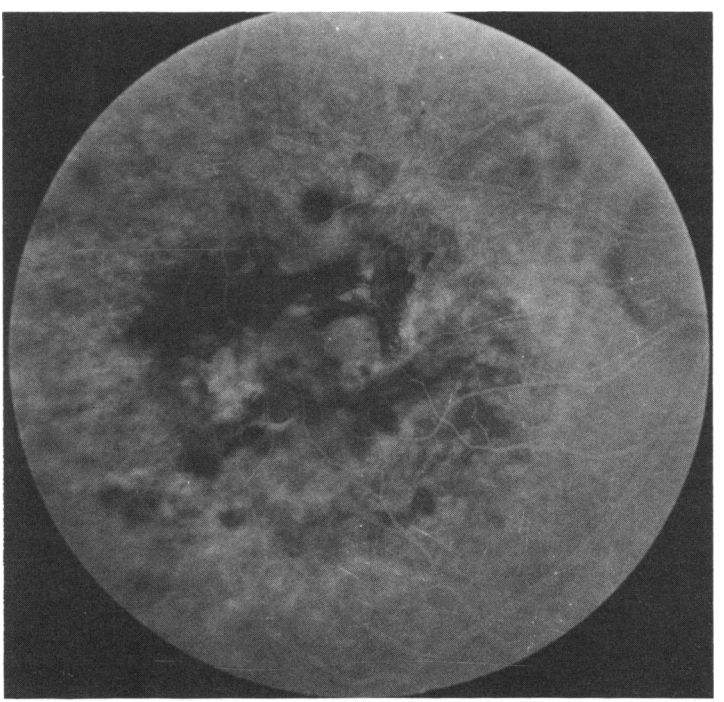

FIGURE 8
Case 2 Fluorescein angiography. Hyper- and hypo- fluorescence are a reflection of the pigment alterations. There is no evidence of subretinal neovascularization or dye leakage.

Electro-oculogram
Normal light rise (220% O.D.; 333% O.S.).

Color Vision
AOHRR - mild blue - yellow defect
Farnsworth Panel D-15 - nondiagnostic color confusion.

Goldmann Visual Fields (1 mm² white)
Bilateral central scotoma: O.D. 10-15°; O.S. 5° and slightly eccentric to fixation.

Retinal Sensitivity Profiles
O.D. elevated 3° to 10° nasally and temporally; O.S. elevated 3° temporally. Peripheral sensitivities normal.

Course
The patient was followed for two years during which time he demonstrated no change in his visual acuity, fundus appearance, or visual function tests.

CASE 3
(111-6)
This patient presented to the Barraquer Eye Clinic at age 35, six months after he noted decreased vision in his right eye.

Visual Acuity
RE: 6/200 (20/600) LE: 6/6 (20/20)

Fundus
RE: "post-hemorrhagic macular dystrophy" LE: "supramacular hemorrhage" (Figure 9).

The remainder of the ocular examination was normal.

Course
Examination throughout the next year showed a steady deterioration in vision in the left eye from 6/6 (20/20) to 6/12 (20/40) and finally to 6/18 (20/60) associated with a macular hemorrhage. The paramacular areas in both eyes were photocoagulated. Two years later there had been no further hemorrhage and the vision was RE 6/90 (20/300) and LE 6/18 (20/60).

CASE 4
(111-3)
This brother presented to the Barraquer Eye Clinic at age 33 with a two year history of bilateral visual loss, noted in the left eye and followed shortly thereafter in the right eye.

Vision
RE: 6/150 (20/500) LE: 6/150 (20/500)

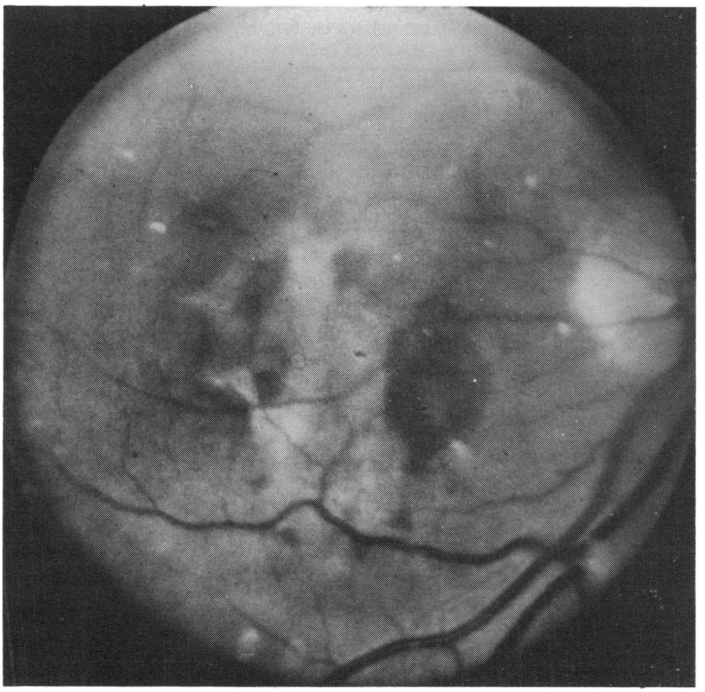

FIGURE 9
Case 3 Pigment atrophy, pigment accumulation and gliosis occupy the macular region. A subretinal hemorrhage is located between the macula and the optic disc.

Fundus
"macular dystrophy with posthemorrhagic chorioretinal atrophy."

Course
Over a 12 year period the condition has remained stable and the most recent visual acuity was RE 6/120 (20/400) and LE 6/60 (20/200).

RESULTS

The four members of this family demonstrated the essential features of pseudoinflammatory macular dystrophy (PMD) (Table 1). It is inherited as an autosomal dominant disorder with symptoms becoming evident in the third to fifth decade. Ophthalmoscopic examination shows a hemorrhagic and exudative maculopathy associated with pigment atrophy, pigment clumping, and eventually glial scar formation. Although visual symptoms

TABLE I: CHARACTERISTICS OF PSEUDOINFLAMMATORY MACULAR DYSTROPHY

1. Autosomal dominant
2. Onset - 3rd to 5th decades
3. Bilateral subretinal neovascularization
   a. Hemorrhagic and exudative maculopathy
   b. Subretinal gliosis with pigment proliferation and atrophy
4. Local (geographic) macular dystrophy
   ERG, EOG, and peripheral retinal sensitivities are normal

may initially be monocular, the other eye becomes symptomatic shortly thereafter and this is invariably a bilateral disease. Cases 1 and 3 demonstrated the early stage of this disorder and fluorescein angiography in one patient clearly demonstrated subretinal choroidal neovascularization. Since the net was located eccentric to the avascular zone of the fovea in the left eye we elected to treat that area with heavy argon laser burns. The rapid luxuriant growth of this neovascular network probably resulted from incomplete coagulation and was apparently the cause of the precipitous fall in vision. Case 3, in whom the data is less complete, had successful photocoagulation treatment and the vision has remained at a steady level for two years.

Follow-up examination of these four brothers was done over a period of 2 to 12 years. Once the cicatrical phase was reached there was virtually no progression in the size of the lesion nor in the visual symptoms. Visual function tests at this stage confirmed that this was a localized geographic disorder confined to the macular region and not part of a generalized retinal degeneration.

Psychophysical and electrophysical tests done on two family members (Cases 1 and 2) show that in both the initial and later stages of this disorder the ophthalmoscopically normal retina is functionally normal. This is evident by the normal peripheral retinal sensitivity, a normal ERG, and a normal EOG.

DISCUSSION

In 1949 Sorsby, Mason, and Gardner[1] described a hereditary hemorrhagic maculopathy in five family pedigrees which was characterized by autosomal dominant inheritance, onset in the 4th and 5th decades, and progression to a vision of 6/60 or worse in each eye. The initial appearance consisted of hemorrhage, exudate and edema confined to the macula. Progression of the lesion over a variable period of time resulted in pigmentary atrophy, pigmentary clumping, glial cicatrization, and choroidal sclerosis. These late changes were noted in the posterior

pole with extension into the more peripheral retina. The similarity between the late stages of this disease and a postinflammatory process has prompted the descriptive term "pseudoinflammatory" macular dystrophy.[2]

A search of the literature by Sorsby and associates[1] suggested that certain previously reported cases[6-11] may have had the same clinical entity, but a careful review of these cases by Deutman[12] failed to confirm placing any of them in the category of pseudoinflammatory dystrophy. Subsequent to Sorsby's initial report other families with a similar appearance were reported by Burn,[13] Francois,[14] Seedorff,[15] Rosen and Leighton,[16] Fraser and Wallace,[17] and Kalmus and Seedburgh.[18] Fluorescein angiography performed by Rosen and Leighton[16] in one affected individual showed hyperfluorescence related to extensive pigmentary defects in the macula and, in the periphery, a fine mottling related to changes in the pigment epithelium. There was no mention of subretinal choroidal neovascularization or dye leakage; however, the patient had advanced disease.

Ashton and Sorsby[3] examined histological sections from the eyes of two sisters, age 70 and 71, who were reported in Sorsby's initial publication.[1] The family history was incomplete since no other members were affected and their father had died at age 40. It is also interesting that Sorsby had previously reported these patients as examples of central and peripapillary choroidal sclerosis.[19] The histological findings in the four eyes were similar and demonstrated: (1) ruptures in Bruch's membrane, (2) subretinal neovascular tissue originating from the choroid and related to the dehiscences in Bruch's membrane, (3) destruction of the outer retinal layers and replacement with glial tissue, and (4) atrophy of the choriocapillaris with fibrous thickening of the intervascular stroma and chroidal vessel wall. Colloid bodies on Bruch's membrane were seen in one individual but not in the other. As the authors point out, the pathological picture closely resembled disciform degeneration.

Various and inconsistent ocular and systemic disorders have been noted in association with pseudoinflammatory macular dystrophy. Glaucoma was seen in two family members described by Sorsby and associates,[1] and in one of the patients (Case 2) in this report. The absence of glaucoma in other members of these pedigrees suggests a fortuitous association. Two reports[17,18] describe various color anomalies of nonspecific type in younger, as yet unaffected family members. Whether this anomaly will serve as a genetic marker to determine individuals at risk or whether the gene for the color defect segregates independently from the gene for pseudoinflammatory dystrophy awaits follow-up examinations.

Hereditary renal glycosuria has been reported with macular degeneration in two siblings.[15] unfortunately, there are no pictures available. The mother was said to be "weak-sighted" from the age of 30. The consanguinity of the parents, renal glycosuria in all 11 siblings and both parents, and nystagmus and poor vision in 3 other siblings make it unlikely that the gene for renal glycosuria is allelic for the retinal disorder, or indeed, that this is in fact a pedigree with pseudoinflammatory dystrophy.

Finally, there appears to be a high association of drusen or white dots in this disease. Sorsby and associates[1] noted this in members of three of the five pedigrees studied and they were verified on histologic section in one case.[3] Fundus drawings and descriptions in the pedigrees reported by Burns[13] and Rosen and Leighton[16] similarly indicated the presence of colloid bodies which were seen centrally or peripherally. Fluorescein angiography gave evidence to a "generalized abnormality of the pigment epithelium."[16] Only the fundus drawing of the Francois case,[20a] the photographs in the report of Fraser and Wallace,[17] and the family in this report do not show drusen. The relationship between drusen of Bruch's membrane and subretinal neovascularization has been well-documented.[21,22] It is certainly conceivable that some cases of presumed pseudoinflammatory macular dystrophy were in fact dominant drusen of Bruch's membrane.

A differential diagnosis of PMD would include those hereditary and acquired diseases associated with subretinal neovascularization (SRN). The presumed ocular histoplasmosis syndrome may have SRN located in the macula of one or both eyes. A positive histoplasmin skin test and the associated findings of peripheral atrophic spots and peripapillary atrophy should suggest this diagnosis. Subretinal neovascularization also has been seen in individuals without any predisposing causes. It can occur in the peripapillary region[23] or in the macula,[24] is usually unilateral (although bilateral cases are reported),[24] and shows no evidence of heredity.

The inherited ocular diseases associated with SRN are most likely to be confused with pseudoinflammatory dystrophy are listed in Table 2.

Angioid Streaks

The hereditary pattern of angioid streaks depends upon the systemic disease present. However, since the great majority of these patients have pseudoxanthoma elasticum[25,26] the mode of inheritance is usually autosomal recessive. The typical appearance of the streaks should pose no problem and fluorescein angiography may reveal subtle streaks that have been overlooked.[27]

TABLE II: SUBRETINAL CHOROIDAL NEOVASCULARIZATION IN HEREDITARY RETINAL DISEASES

1. Pseudoinflammatory macular dystrophy (AD)
2. Angioid streaks (AR,AD)
3. Vitelliform macular dystrophy (AD)
4. Drusen of Bruch's membrane (AD)
5. Optic nerve drusen (AD)
6. Myopia (AR,AD)

AD = Autosomal Dominant
AR = Autosomal Recessive

Vitelliform Macular Dystrophy

Vitelliform macular dystrophy is inherited as an autosomal dominant and ophthalmoscopically may resemble choroiditis, choroidal sclerosis, drusen, or disciform degeneration. The presence of SRN has been documented[28] and we have seen the propositus in a family pedigree of vitelliform dystrophy present as unilateral SRN.[29] The more typical vitelliform lesions seen in other family members and the finding of an abnormal EOG in these family members carrying the gene is necessary to confirm this diagnosis.

Drusen of Bruch's Membrane

Dominantly inherited drusen of Bruch's membrane may affect the macular region. The association of macular drusen with SRN and disciform degeneration has been well-documented by Gass.[21,30] With this in mind, a review of previously reported cases of pseudoinflammatory macular dystrophy indicates that the Ewbank family, the Kempter family, and the Cranston sibship described by Sorsby and associates[1] and the reports of Burns[13] and Rosen and Leighton[16] may represent dominant drusen with SRN. In our family there was no evidence of drusen on ophthalmoscopic or angiographic examination.

Optic Nerve Drusen

Drusen of the optic nerve head, also inherited as an autosomal dominant,[31] have been reported in association with a hemorrhagic maculopathy[32,33] and SRN extending into the macula from the disc has been seen on angiography.[23] The maculopathy may be seen at any age and has been reported in both a 4-year-old child[23] and an 11-year-old child whose mother had drusen and subretinal fibrosis.[33]

Myopia

High myopia is usually transmitted as an autosomal recessive[20b] but

autosomal dominant pedigrees have been reported.[20c] A hemorrhagic maculopathy with pigment accumulation (Fuchs' spot) or subretinal gliosis can occur. Fluorescein angiography has demonstrated the presence of SRN beneath the macula.[34] High myopia has not been reported in any cases of PMD although the late stages in several of the cases of Sorsby and associates[1] resemble a myopic chorioretinal degeneration of the posterior pole.

The possibility of efficacious treatment of SRN with photocoagulation has been outlined by others.[35,36,37] When the SRN is eccentric to the avascular zone of the fovea and the treatment is intense to the entire extent of the net, visual acuity may be preserved at a normal or near the pretreatment level.[38,39] The preservation of pretreatment vision in one patient (Case 3) following photocoagulation should lead others to consider this modality in similar cases in spite of the total lack of success in another patient (Case 1).

SUMMARY

A family with pseudoinflammatory macular dystrophy (PMD) is presented. This dominantly inherited macular dystrophy has its onset in the 3rd to 5th decades with the earliest manifestation being a macular subretinal neovascular network. Visual function tests (ERG, EOG, visual fields, retinal sensitivity) in the early and late stages indicates this is local or geographic disease.

This dystrophy should be differentiated from other hereditary causes for subretinal neovascularization (angioid streaks, vitelliform dystrophy, dominant drusen of Bruch's membrane, optic nerve drusen and myopia). It is suggested that treatment be directed at early obliteration of the subretinal neovascularization with intense photocoagulation since the outcome in virtually all cases of untreated PMD is legal blindness.

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DISCUSSION

Dr J. Donald M. Gass. The authors have presented the ophthalmoscopic and electrophysiologic findings in four members of a family with probable dominantly inherited disciform macular dystrophy. They choose to classify these patients as having Sorsby's pseudo-inflammatory macular dystrophy because of the onset of visual loss in the third to fifth decades of life, and because of the absence of any other associated ocular findings. I believe that the eponym, "Sorsby's pseudo-inflammatory macular dystrophy," is an inappropriate one for several reasons. Firstly, the hypertrophic diciform scars do not resemble most postinflammatory chorioretinal scars which are atrophic, for example, toxoplasmosis; they are not associated with postinflammatory changes in the overlying vitreous; and they resemble in every way the hypertrophic disciform scars seen in patients with a variety of macular dystrophies, for example, drusen of Bruch's membrane and angioid streaks. Secondly, three of Sorsby's five pedigrees had evidence of drusen in Bruch's membrane. Thirdly, histopathologic examination of the eyes of two sisters of one of his pedigrees showed the typical findings of senile macular dystrophy or degeneration. Fourthly, identical dominantly inherited disciform lesions unassociated with other intraocular findings may occasionally occur in patients of any age, including children. Fifthly, since the authors did not examine more than four members of their family, particularly older members, we cannot be sure that the family does nto have dominantly inherited drusen. The yellow lesions seen in the left eye of the authors' Case 1 (Fig. 2B) may represent drusen. It is well recognized that drusen in the macular area disappear after the development of disciform detachment of the macula. We cannot exclude the possibility that the patients illustrated in Figures 7 and 9 did not have drusen prior to their detachment. Normal EOG and ERG findings do not exclude the diagnosis of dominantly inherited drusen.

In summary, while I agree that this family has a dominantly inherited macular dystrophy I suggest that the term, "Sorsby's pseudo-inflammatory dystrophy," should be abandoned because it does not refer to a specific disease.

DR RONALD E. CARR. I cannot agree with Dr Gass that the patients we are dealing with have drusen of the macula with associated macular hemorrhage. There are certainly a number of entities in which macular hemorrhagic changes are seen, some of which are noted today. Merely because these are cases with an autosomal dominant heredity does not make them *de facto* cases of macular drusen. While I feel one must be careful to look for and think of drusen whenever such macular changes are seen. I likewise feel that there may well be another entity, inherited as an autosomal dominant, in which there are no associated drusen. Certainly, several of our cases which were seen in the early stages had no evidence of drusen, and as will be noted in the published text, there are a number of other cases in the literature in which no evidence of drusen was present.

I certainly agree with Dr Gass that the term "pseudoinflammatory macular dystrophy" is a poor one. However, one dislikes the constant addition of names to the literature even though a term such as "hereditary hemorrhagic macular dystrophy" would have more meaning. The former term, originally used by Duke-Elder, is merely an expression of the late-stage ophthalmoscopic picture.

Thank you.