MASSIVE CHOROIDAL DETACHMENT MASKING OVERLYING PRIMARY RHEGMATOGENOUS RETINAL DETACHMENT: A CASE SERIES

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SUMMARY

Choroidal detachment (CD) along with primary rhegmatogenous retinal detachment (RRD) as a presenting finding is a well-recognised association. The pathogenesis appears to revolve around the hypotony induced by the primary RRD and an unstable choroidal vascular system. We present 3 patients with massive CD with minimal or no overlying RRD, initially misdiagnosed as a CD due to other causes. Since the correct diagnosis may be delayed, such cases form a real challenge in differential diagnosis and management. The underlying cycle where the retinal defect of a primary RRD is buckled by a CD, followed by the regression of the latter and the recurrence of the RRD is demonstrated.

RÉSUMÉ

Un décollement de rétine rhegmatogène, se présentant avec un décollement choroïdien est une association bien connue. Le mécanisme pathogénique consiste en une hypotonie induite par le décollement de la rétine et une instabilité du système vasculaire choroïdien. Nous présentons 3 patients avec décollement choroïdien massif-avec ou sans décollement de rétine - lequel était erronément diagnostiqué comme décollement d'autre origine, ce qui pose un problème de diagnostic différentiel. Nous illustrons le mécanisme par lequel une déchirure rétinienne est fermée par le décollement choroïdien et est suivie d'une récidive de décollement rétinien lors de la résolution du décollement de la choroïde.

SAMENVATTING

Een preoperatief beeld van een chorioideale loslating met een rhegmatogene retinaloslating is een gekende klinische associatie. De pathogenese berust vermoedelijk op een door de retinaloslating veroorzaakte hypotonie en een instabiel chorioideaal vasculair systeem.
Wij beschrijven 3 patiënten, met een prominente chorioideale loslating die aanvankelijk de onderliggende retinaloslating geheel of gedeeltelijk maskeerde en zo de uiteindelijke diagnose vertraagde. Deze situaties vormen een differentiaal diagnostische uitdaging. Het onderliggend mechanisme waar een chorioidealoslating aanvankelijk een retinaal defect tamponneert, gevolgd wordt door een regressie van de chorioideale loslating en een recidief van de retinaloslating, wordt geïllustreerd.

KEY-WORDS

choroidal detachment, rhegmatogenous retinal detachment, differential diagnosis

MOTS-CLÉS

décollement choroïdien, décollement de rétine rhegmatogène, diagnostic différentiel

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received: 14.06.01
accepted: 10.09.01

INTRODUCTION

Choroidal detachment (CD) refers to a collection of fluid in the potential space of the suprachoroid, which is external to the main structural and functional layers of the choroid and ciliary body. This has been well documented following surgery for cataract, glaucoma or retinal detachment. Other underlying aetiologies to consider in CD are inflammation (scleritis, Vogt-Koyanagi-Harada syndrome), tumours (primary or metastatic), nanophthalmos, uveal effusion syndrome and carotid-cavernous fistula. Several authors have also described choroidal detachments along with an underlying primary rhegmatogenous retinal detachment (RRD) as the presenting finding (2-5, 7, 10)
Gottlieb (2) and Seelenfreund (5) reported incidence rates of 4.5% and 2 % respectively. As the CD may be initially misdiagnosed as being part of a different underlying condition and the correct diagnosis delayed by massive CD obscuring the underlying retinal tear, such cases form a real challenge in differential diagnosis and management. The pathogenesis of this complication is not clear but hypotony, initiated by the detachment of the retina may play an important role in its development. We present three cases of combined choroidal and retinal detachment as a primary finding, to help in diagnosing the complex at an early stage.
The underlying cycle, where the retinal defect of a primary RRD is buckled by a CD, followed by the regression of the latter and the recurrence of the RRD, will be illustrated.

CASE REPORTS

Case 1

An 86-year old man was referred to the glaucoma clinic in September 2000 with a choroidal detachment in his better right eye following uncomplicated phacoemulcification 3 months before. The CD had been considered elsewhere to be due to postoperative hypotony. He had a past ocular history of bilateral primary open angle glaucoma, for which he had undergone a right trabeculectomy in February 1992.
On presentation there was no ocular discomfort and the visual acuity, which had become worse 6 weeks before, was hand movements on both eyes. Slitlamp examination showed a right quiet pseudophakia with normal anterior chamber depth and a Seidel negative filtration bleb with a loose conjunctival flap. The anterior segment on the left was unremarkable apart from some nuclear sclerosis and a peripheral iridotomy. The intraocular pressure (IOP) was 4 mm Hg on the right and 14 mm Hg on the left. Funduscopy not only revealed a CD involving all quadrants with the exception of the inferior one, but also a previously undiagnosed U-tear in the temporal retina. Ultrasound examination confirmed the CD and excluded an underlying posterior scleritis. After referral to the vitreoretinal unit he underwent a vitrectomy with suprachoroidal drainage and silicone oil tamponnade under general anaesthesia. Postoperatively the CD was resolved and the retina was flat. On the last follow-up visit in January 2001 the visual acuity was 6/60 on that eye and the retina remained flat.

Case 2

In September 2000 a 55-year old afro-caribbean man presented to our casualty with a history of a painful red right eye since 4 days. His past ocular and medical history was unremarkable. The visual acuity was hand movements on the right and 6/12 with a pinhole on the left. The right pupil was fixed. Slitlamp examination of the right eye showed a very deep anterior chamber with marked flare, but no cells. Anterior segment on the left was normal. The IOP was 4 mm Hg and 19 mm Hg, on the right and left respectively. Funduscopy initially revealed a massive choroidal detachment involving the nasal and temporal quadrants on the right and a normal retina on the left. He was referred and seen in the vitreoretinal department the same day. On funduscopy, in addition to the CD, a retinal detachment was suspected but no retinal breaks were found. Ultrasound examination confirmed a massive CD with shallow overlying posterior inferior RD and excluded an underlying solid mass (fig. 1). A treatment of atropine 1% and Pred Forte® drops was started. One day later, as the view of the posterior segment cleared, an inferior retinal detachment with 2 U-tears temporal and superotemporal on the vitreous base was discovered on funduscopy. There were also some vitreal

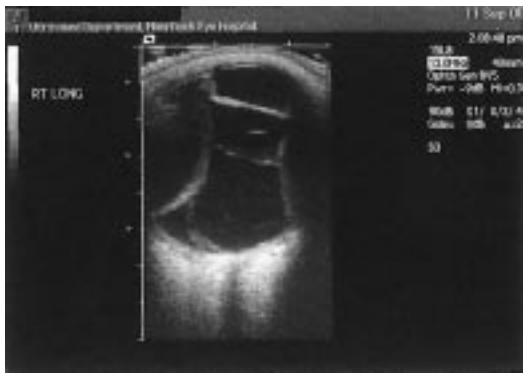

Fig 1. Ultrasound of the right eye of patient 2: Massive choroidal detachments involving the nasal and temporal quadrants and concealing an inferior posterior shallow retinal detachment.

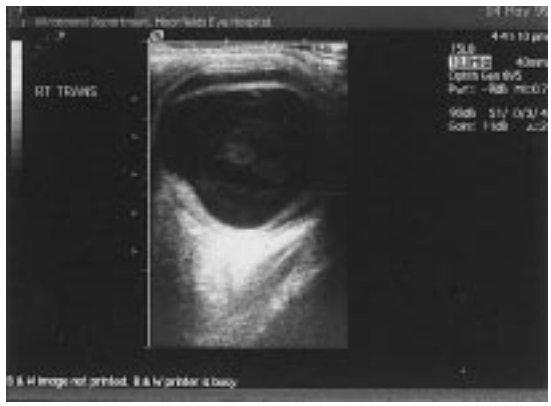

Fig 2. Ultrasound of the right eye of patient 3 on initial presentation: Nasal and temporal choroidal detachment without evidence of retinal detachment.-

cells and a cyclitic membrane present. Blood tests including full blood count (FBC), urea and electrolytes (U&E), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), angiotensine converting enzyme (ACE) were normal. Syphilis serology was positive, but was interpreted as a past treponemal inactive infection. Chest X-ray was normal. In order to bring the intraocular inflammation down, an oral prednisolone course of 60mg per day was given, tapered down gradually by 10 mg each week in addition to the topical treatment. On the follow-up visit in October the retinal detachment had become complete and was complicated by proliferative vitreoretinopathy (PVR). A vitrectomy was offered but the patient refused any surgical intervention in view of the very guarded prognosis.

Case 3

An 11-year old boy, originally from Pakistan, presented to our casualty department in May 1999, complaining of reduced vision in his right eye and sore eyes since 3 weeks.

Since early childhood he and his brother were regularly followed up in Moorfields Eye Hospital for bilateral microphthalmos, dislocated cataractous lenses, extensive pigmentary retinopathy involving the posterior pole and divergent squint. His vision used to be 6/18 bilaterally with + 7.5 spherical diopter and - 0.50 cylindrical diopter at 180° correction.

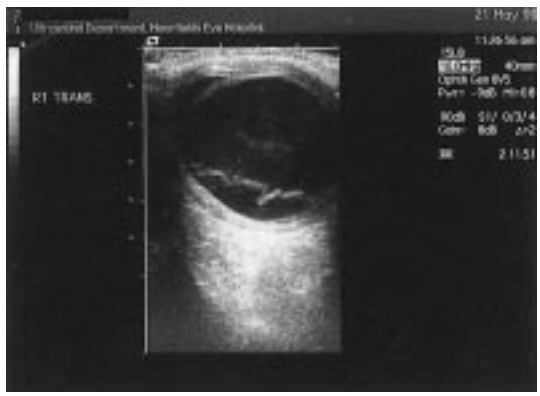

Fig 3. Ultrasound of the right eye of patient 3 two weeks later: Choroidal detachment in regression and bullous inferior half retinal detachment.

His vision had now dropped to hand movements on the right and slitlamp examination showed bilateral quiet anterior chambers with upwards and inwards dislocated cataractous lenses. The eye pressures were 0 and 16 mm Hg, on the right and left respectively. The pupils dilated poorly. Funduscopy was difficult and revealed nasal and temporal choroidal detachments on the right without any retinal detachment. There was no retinal detachment present on ultrasound B-scan (fig 2). A treatment with local steroid drops and atropine 0.5 % was started. Two weeks later however, ultra sound B-scan demonstrated the diminution of the CD and revealed a bullous retinal detachment inferiorly (fig 3). In June 1999 a vitreolensectomy combined with cryo-buckle and SF_6 gas

tamponnade was performed. Peroperatively a total retinal detachment was present with multiple small dialyses. The patient underwent further vitrectomies and in February 2001, the vision was counting fingers on the right and the posterior pole was attached with persisting subretinal fluid and PVR in the inferior retina.

DISCUSSION

A choroidal detachment (CD) combined with rhegmatogenous retinal detachment (RRD) has been considered a major preoperative complication of RRD. The predisposing factors include high myopia, aphakia or pseudophakia and old age and it is more common in non-caucasean population. The clinical presentation can occasionally mimic uveitis, including ocular pain, red eye and markedly impaired vision, as is the case in patient 2. Slit lamp examination can demonstrate a deep anterior chamber with occasionally intense reaction. Usually a marked hypotony is present. The CD can occasionally be massive as illustrated in figure 1. Vitreous haze with cells and CD make the detection of retinal defects difficult, leading to a delay in diagnosis of days, weeks or even months as illustrated in our cases. In case 1, a retinal detachment was most likely the underlying cause of the CD rather than pure postoperative hypotony, since in the latter, the detachments as a rule subside spontaneously in a short time. The retinal detachment was missed for probably many months. In case 3, there was a delay in the final diagnosis of 14 days.

Such CD can easily be misinterpreted as coming from other underlying inflammatory or tumoral aetiologies or being caused by nanophthalmos, uveal effusion syndrome and carotid-cavernous fistula. An exudative RD can accompany such CD. Although an underlying syphilitic intermediate uveitis causing retinal tears cannot be ruled out completely in case 2, the massive CD seen in this patient is most likely due to the RD. Massive CD are very unusual for intermediate uveitis.

The pathogenesis of this condition is not clear but appears to revolve around the profound hypotony and an unstable choroidal vascular system.

RRD is probably the primary event and decompensates the aqueous production (1) by a combination of ciliary body oedema (11) and an increased absorbing surface of the retinal pigment epithelium exposed to the subretinal fluid. The resulting acute hypotony disturbs the equilibrium in the choroidal circulation between intraocular, intravascular and colloidal pressure, the latter exerted by plasma proteins. In normal circumstances, the pressure in the choroidal space is about 2 mm Hg less than the IOP, according to animal studies (9). In response to the hypotony the choroidal arterioles will dilate, which results in transudation of protein rich fluid into choroidal and suprachoroidal space since the choroidal capillary and venous system operate at near-capacity-volume-load (8). This will lead to further oedema and detachment of the ciliary body enhancing the process. The degree of CD may be related to the compressibility of the vitreous, the permeability of the sclera, and the status of the choroidal vessels, explaining the higher occurrence in an older age group (5). The CD often buckles the retinal hole or tear, the subretinal fluid is absorbed by the increased surface of RPE and the retinal detachment is temporarily repaired. With flattening of the retina and restoration of the normal anatomy together with the original vitreous traction forces, the retinal hole or tear may reopen, causing a redetachment of the retina and creating a cycle. We believe this mechanism took place in patients 2 and 3, where after a short topical steroid treatment, the choroidals started to resolve, revealing the retinal defects in fundo clearly at a later stage. In the third case this process has even been documented on B-scan by a senior ultrasound operator (fig 2 and 3). The hypotony-related breakdown of the blood-retinal barrier, which is responsible for the CD, releases increased amounts of serum components in the intra ocular fluids. This factor, in combination with the delay in surgical intervention because of difficulties in detecting retinal holes, keeps the eye in a microenvironment for cellular proliferation, possibly leading to postoperative PVR (6). This explains the guarded prognosis of such retinal detachment cases, as illustrated in our case series.

In conclusion, an accurate diagnosis of this type of choroidal detachment concealing an overlying primary RRD is a challenging necessity.

Careful follow-up is needed in order to avoid any delay in diagnosis and treatment.

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