CASE REPORT

Acute retinal detachment induced by the Valsalva manoeuvre in morning glory disc anomaly

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Accepted 11 January 2018

SUMMARY

We present a case in which a large, bullous, predominantly inferior, serous retinal detachment developed acutely after the Valsalva manoeuvre (from a coughing fit) in an eye with morning glory disc anomaly. We postulate that a rapid alteration in intracranial pressure was transmitted through the cavitary disc defect. This allowed a sudden influx of cerebrospinal fluid and/or liquefied vitreous into the subretinal space. This previously unreported case provides important evidence for the role of intracranial pressure fluctuations in the pathogenesis of macular schisis and neurosensory detachment secondary to optic disc cavitations.

BACKGROUND

Optic disc pits, optic disc colobomas and morning glory disc anomalies are closely related congenital malformations, which together comprise a clinical spectrum of cavitary disc anomalies.^{1 2} Histologically, these conditions are characterised by a cavitary defect of the optic disc lined by herniated, dysplastic retinal tissue.^{3 4} In optic disc pits, the cavitation is small and localised, while in colobomas and morning glory anomalies, the defect can be extensive, involving most of the disc area.² The anomalous configuration of the optic disc in these conditions allows potential communications to arise between the intraocular and extraocular spaces, specifically the vitreous cavity, the subretinal space and the subarachnoid space, which may be variably interconnected.^{1 2}

Cavitary disc anomalies are associated with maculopathy in 25%–75% of cases.^{5–7} Maculopathy typically begins as macular schisis with or without a neurosensory detachment. In severe cases, total retinal detachment (RD) may ensue. The cause of maculopathy and detachment in these cases is poorly understood. Clinical and histopathological evidence have variously implicated both the vitreous^{8 9} and the cerebrospinal fluid (CSF)^{4 10} as the source of fluid. Moreover, the mechanisms which induce fluid accumulation are unclear, with conflicting theories suggesting a role for either peripapillary traction^{11 12} or CSF pressure fluctuations.⁹

We report a case of a patient with morning glory disc anomaly in which an extensive, bullous, serous RD developed acutely after a coughing fit. To our knowledge, this is the first reported case of acute RD in any eye due to the Valsalva manoeuvre. Perhaps more significantly, this case provides the

most convincing evidence yet that variations in CSF pressure play an important role in the pathophysiology of retinal fluid accumulation in cavitary disc anomalies.

CASE PRESENTATION

A 42-year-old man with no medical or family history presented to the eye casualty department. His only ocular history of note was mild left amblyopia with best corrected visual acuity from his last optician's report 6 months previously of 6/5 right, 6/9 left. He had never before seen an ophthalmologist.

The patient supplied without prompting a detailed, convincing history of the sequence of events leading up to his presentation. Five days before presentation, he explained that he awoke early in the morning with “the worst coughing fit of my life”. Within seconds of the coughing fit, he became aware of a dense, central scotoma in the left eye. Over the next 6 hours, the scotoma enlarged into a monocular, centre-involving, complete superior visual field defect. Over the ensuing days, the visual field defect remained stable after which he sought medical attention.

Visual acuity at presentation was 6/6 right, 6/24 left. Right eye examination was entirely normal, including a normal optic disc. In the left eye, he had a morning glory disc anomaly. There was an extensive area of RD, equivalent to one quadrant of the fundus, extending from the optic disc to the equator from three o'clock temporally to six o'clock inferiorly (figure 1A). The detachment was bullous near to the optic disc inferiorly and temporally, and became more shallow peripherally. No vitreous pigment migration or retinal breaks were found. The vitreous was clinically attached (figure 2A). Optical coherence tomography (OCT) of the macula showed macular schisis and a large neurosensory detachment extending to the periphery (figure 2B).

OUTCOME AND FOLLOW-UP

Surgical options including vitrectomy were discussed with the patient at his initial visit and at subsequent visits but the patient declined intervention. The patient was averse to surgery given the small associated risks and given that subretinal fluid may improve or resolve spontaneously in some cases of detachment related to cavitary disc anomalies.^{1 2} At his most recent review at 21 months' postpresentation, the area of detachment had shifted slightly temporally compared with his initial presentation

To cite: Fajgenbaum MAP, Antonakis SN, Membrey L, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223131

Figure 1 Fundus photograph of the left eye (A) at presentation and (B) 21 months following presentation showing in both cases extensive neurosensory detachment of the retina extending from the optic disc to beyond the equator. The detachment shifted temporally over time but did not diminish in extent, which is demarcated by the black arrows.

but the fluid had not resolved (figure 1B) and visual acuity was 6/24. The patient remains under active review.

DISCUSSION

The aetiology of intraretinal and subretinal fluid accumulation in congenital optic disc cavitations has not been fully established. Traction is a commonly proposed mechanism. Evidence in support of a tractional aetiology is circumstantial and includes the following: breaks have been identified in dysplastic retina overlying disc cavitations¹¹; abnormal membranes and fibroglial tissue often overlie disc cavitations¹; maculopathy tends to develop in the third and fourth decades when vitreous has liquefied and posterior vitreous detachments (PVDs) develop⁶;

vitrectomy and gas has a high reported success rate, potentially by relieving vitreous traction¹²; both internal limiting membrane (ILM) peeling and trimming of peripapillary fibroglial tissue at the time of vitrectomy have a high reported success rate, although there are no randomised comparisons to determine the additional benefit of these manoeuvres over standard vitrectomy.^{1 2} On the other hand, some circumstantial evidence points away from a tractional aetiology. Retinal fluid often accumulates in eyes with cavitory anomalies in the absence of a clinically evident parapapillary break and without the sorts of exuberant tractional membranes typically associated with retinal fluid accumulation in other tractional pathologies.^{1 2 13} Moreover, retinal fluid can persist or reaccumulate after a vitrectomy, when all vitreous traction at the disc should have been relieved.¹⁴

An alternative theory to peripapillary traction is that extra-ocular CSF pressure fluctuations play the primary role in fluid accumulation. This was most convincingly discussed in a paper by Johnson and Johnson.⁹ They proposed that physiological variations in perineural CSF pressure, as can occur during everyday postural changes of the head, for example, may be transmitted across the cavitory disc anomaly to affect the intra-ocular compartments. They postulated that in cases where the posterior collagenous capsule of the cavitation or pit is non-porous (figure 3), CSF pressure variations cause the capsule itself to act like an enclosed pipette bulb which expands and contracts, sucking vitreous fluid into the subretinal space. On the other hand, in eyes where the posterior capsule of the cavitation is porous, CSF itself can enter and leave the subretinal space directly during pressure fluctuations. In this way, retinal fluid can be recruited from either the vitreous or the CSF, depending on the porosity of the posterior collagen capsule and depending on the anatomic interconnections present in the particular anomaly.

The model proposed by Johnson and Johnson is convincing on a number of levels. First, it unifies the apparently contradictory evidence in the literature—where in some case reports, the retinal fluid has convincingly emanated from the vitreous^{8 9} and in others the CSF.^{4 12} In their model, retinal fluid may arise from either source, depending on the anatomy of the particular disc.

Their model also helps to explain the bimodal age of presentation of optic cavitation maculopathy. Many cases occur in the third and fourth decades, when vitreous liquefaction and PVDs develop. Other cases present in childhood, when the vitreous has not yet liquefied and tractional forces should be negligible. Johnson and Johnson suggested that cases occurring in childhood are likely to have a porous posterior capsule allowing a

Figure 2 (A) Optical coherence tomography (OCT) image of the left optic disc at presentation. The cross-section runs inferonasal to superotemporal along attached retina through the disc and shows the cavitation defect. (B) OCT of the left macula at presentation showing schisis and neurosensory detachment.

Figure 3 Schematic representation of the anatomy of a cavitory optic disc defect, as occurs in optic disc pits and morning glory anomaly. The cavitory defect is lined internally with herniated, dysplastic retinal tissue. The posterior border comprised a collagen-rich sac extending into the subarachnoid space through a breach in the lamina cribrosa. It is theorised by Johnson and Johnson⁹ that fluctuations in intracranial pressure (ICP) cause ingress of intraretinal and subretinal fluid. In (A) and (B), the posterior collagenous capsule of the cavitory defect is intact. ICP pressure fluctuations cause the capsule to expand and contract akin to a pipette bulb, which transmits the pressure gradient to the intraocular space, sucking vitreous fluid into the retina. In (C), the posterior capsule is porous so fluctuations in ICP cause the cerebrospinal fluid to move directly into or out of the subretinal space (reproduced with permission from Johnson and Johnson⁹).

direct communication between liquid CSF and the retina. Meanwhile, cases that develop in later adulthood are likely to have a non-porous capsule, which therefore relies on the recruitment of aged, liquefied vitreous into the subretinal space by a pipette effect.

On a purely mechanical level, the model by Johnson and Johnson provides the only convincing explanation for why fluid in a larger compartment—such as the vitreous cavity—should overcome its inherent surface tension to migrate through a narrow fissure or channel into the retina. Such a phenomenon, as has been described in several cases where intravitreal gas and oil migrated subretinally through an optic disc pit,¹⁹ could only conceivably occur if a pressure gradient were present between the vitreous and the subretinal space. Such pressure gradients do not exist physiologically within a closed eye, and could only be explained by CSF pressure fluctuations being transmitted across the cavitation to the intraocular compartments.

Our case provides perhaps the most convincing evidence yet in support of Johnson and Johnson's model of CSF fluctuation. In our case, there was no evidence of abnormal traction at the disc, no prominent fibroglial tissue on the disc surface and no evidence of peripapillary retinal breaks. The acute onset of symptoms immediately following the Valsalva episode supports a causal relationship. The abnormally large volume and rapid accumulation of fluid in our case compared

Learning points

- ▶ This unique case shows that the Valsalva manoeuvre can cause an acute retinal detachment in patients with congenital optic disc cavitations.
- ▶ This case provides strong support for the theory that variations in cerebrospinal fluid pressure are responsible for the insidious maculopathy (macular schisis and neurosensory detachment) associated with congenital optic disc cavitations.

with typical optic disc cavitation maculopathy suggests that the inciting forces which create maculopathy in the first place, such that they are, were exaggerated. There is no evidence that the Valsalva manoeuvre exacerbates intraocular traction. The Valsalva manoeuvre does, however, cause a sudden increase in perineural CSF pressure, which was estimated in one study to be 11 mm Hg above and beyond baseline physiological fluctuations.¹⁵

In keeping with Johnson and Johnson's model, we propose that the subretinal fluid in our case may have conceivably emanated from either liquefied vitreous or the CSF, depending on the porosity of the posterior capsule of the cavitation anomaly.

Currently, optic disc cavitation maculopathy is treated either conservatively (since fluid may spontaneously improve) or with vitrectomy and tamponade, with or without adjuvant treatments such as ILM peeling and parapapillary laser. Cases which fail to improve despite repeated vitrectomies pose a real therapeutic challenge. In the future, as the aetiology of this condition becomes better understood, it may be that surgical or medical therapy aimed at reducing CSF pressure fluctuations, or their transmission to the intraocular compartment, may prove beneficial. We recommend advising patients with congenital disc cavitations that the Valsalva manoeuvre may be associated with progression of maculopathy.

Contributors MAPF: conception, design, patient management, data collection, drafting paper and approval of final script. SNA and LM: conception, design, patient management, data collection and approval of final script. DAL: conception, design, patient management, data collection, editing and approval of final script.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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