Other full case

Simultaneous extrusion and intrusion of a scleral buckle in a patient with Marfan's syndrome

K T Oyewole,¹ A J Shortt,² Y Ramkissoon,³ P M Sullivan⁴

¹Department of General Medicine, West Middlesex University Hospital, London, UK;

²Department of Cornea, Moorfields Eye Hospital, London, UK;

³Moorfields Eye Hospital, London, UK;

⁴Department of Vitreo-retinal Surgery, Moorfields Eye Hospital, London, UK

Correspondence to K T Oyewole, kehinde.oyewole@imperial.ac.uk

Summary

An Asian male in his late 50s with known Marfan's syndrome (MFS) presented with sudden deterioration in vision and loss of globe structure 13 years after the placement of a silicone scleral encircling band for the management of retinal detachment. Surgical exploration revealed simultaneous intrusion and extrusion of the encircling band resulting in globe perforation and collapse. This was managed by removal of the encircling band and a scleral patch graft over the defect. This case highlights that in patients with thin sclera, such as in MFS, scleral perforation is a potential complication that should be considered in patients undergoing scleral buckling. Management of this complication is also technically difficult because of impaired healing of the sclera in these patients.

BACKGROUND

Marfan's syndrome (MFS) is an autosomal, dominant, connective-tissue disorder, with an estimated incidence of between 1/5000 and 1/20 000.^{1–3} It is characterised by musculoskeletal abnormalities, cardiovascular disease and ocular abnormalities.⁴ Approximately 50% of patients with MFS are diagnosed by an ophthalmologist, and few patients may present with isolated ocular features that are suggestive of the syndrome.⁵

The most common ocular abnormalities associated with MFS are lenticular abnormalities (ectopia lentis, microspherophakia and lens opacity), corneal abnormalities (astigmatism, keratoconus and megalocornea), glaucoma, colobomata (iris and lens), retinal tears and retinal detachment.⁵ The incidence of retinal detachment in MFS ranges from 5% to 26%, and it is frequently bilateral. This is significantly higher than the normal population incidence.⁵

Scleral buckling is a commonly used technique to surgically repair retinal detachment. Complications of scleral buckling include refractive changes (myopic shift), infection, raised intraocular pressure, optic neuropathy, extraocular muscle imbalance (resulting in strabismus and diplopia), cystoid macula oedema, reduced corneal sensitivity, anterior segment ischaemia, posterior segment ischaemia, choroidal detachment, re-detachment of the retina, extrusion and scleral intrusion causing globe perforation.⁶

We report the management of a case of simultaneous extrusion and scleral intrusion of an encircling band in a patient with MFS. This led to spontaneous scleral perforation and proved to be a challenging case to manage. This case highlights the difficulties and potential complications in the management of retinal detachment in patients with MFS.

CASE PRESENTATION

An Asian male in his late 50s with known MFS and bilateral, long-standing, ectopia lentis underwent a left vitrectomy, lensectomy, internal drainage, cryotherapy, encirclement and 20% SF6 gas injection for the management of retinal detachment. An intraocular lens was not implanted. This was successful in re-attaching the retina. Thirteen years following this surgery, he re-attended complaining that his eye had suddenly gone soft, and his vision had deteriorated. There was no history of precipitating trauma.

TREATMENT

The patient underwent surgical exploration under general anaesthesia (video 1). Exploration revealed a 276 encircling band with end-to-end sutures in the superonasal quadrant. The encircling band was removed, and the globe reformed with BSS via a corneal paracentesis. The scleral defect was over sewn-in layers using 6/0 Vicryl. At the end of the procedure, the eye was formed and firm, and no leak was visible by Siedel test.

Video 1 Exploration of ruptured globe, removal of explant and primary repair of globe. [10.1136/bcr.08.2009.2182v1](https://doi.org/10.1136/bcr.08.2009.2182v1)

Postoperatively the intra-ocular pressure was initially 0 mm Hg and did not rise above 1 mm Hg. Aqueous was found to be leaking from the conjunctival wound. This failed to resolve, and 3 months following the initial procedure, a second exploration was performed (video 2). The sclera in the area of the leak in the superonasal quadrant was felt to be too thin to suture directly, and, therefore, a 3×5 mm scleral patch graft was sutured onto the defect, using three 8/0 Ethilon mattress sutures and the conjunctiva closed over the scleral patch using 7/0 vicryl.

Video 2 Repair of scleral defect using scleral patch graft. [10.1136/bcr.08.2009.2182v2](https://doi.org/10.1136/bcr.08.2009.2182v2)

OUTCOME AND FOLLOW-UP

For 1 year following the second procedure, the intra-ocular pressure as has remained stable at 3 mm Hg. No choroidal effusions have occurred, though the visual potential of the eye is limited by the previous retinal detachment, aphakia and macular RPE changes. The left eye remains Siedel positive, but only a very small leak from the posterior edge of the scleral patch graft has been identified. This is being managed with prophylactic chloramphenicol 0.5% twice daily.

DISCUSSION

MFS is caused by a mutation of the FBN-1 gene on chromosome 15 that codes for fibrillin-1, a microfilament that is the main component of extracellular microfibrils. These microfibrils contribute to tissue elasticity and strength. Fibrillin has been immunolocalised to many ocular connective tissues. Until recently it was thought that the defective FBN-1 gene negatively influenced microfibrillar assembly, preventing normal elastogenesis and thus weakening the mechanical properties of connective tissue. More recent studies suggest that fibrillin is predominately involved in the function rather than the assembly of elastic microfibrils.

In the sclera of the eye, fibrillin is normally located between the collagen lamellae. The sclera of MFS patients is unusually thin, and few patients can manifest gross scleromalacia.^{5 7} This may be related to the generalised increase in the size of the eye as indicated by the above-average axial lengths seen in these patients. This combination of a structurally thinner sclera and a functionally less elastic tissue makes scleral perforation more likely.

Scleral perforation is a recognised but rare complication of scleral buckling for the treatment of retinal detachment, even in patients who do not have MFS. Its incidence has been estimated to be 1 in 1110 cases in the general population.⁸ Other risk factors for scleral buckle intrusion include thin sclera, high myopia, glaucoma, extreme scleral buckle tension and extensive retinopathy.⁹ A single case of this complication in a patient with MFS was previously reported by Deramo *et al.* The authors reported hypotony secondary to scleral perforation in a 33-year-old woman with MFS.¹⁰ This occurred 17 months following retinal detachment repair with a silicone sponge and encircling band. This was managed by removal of the explant and suturing of the scleral defect, which was successfully closed. The authors concluded that this complication was related to the scleromalacia associated with MFS.

Scleral perforation as a result of previous intraocular surgery in MFS was previously described by Rodríguez-Ares *et al* who successfully repaired the scleral perforation with a combination of preserved allogeneic sclera and amniotic membrane.¹¹ Spontaneous scleral perforation has also been described in MFS. Law *et al* reported spontaneous bilateral scleral perforation in a 18-year-old man, which was successfully treated using scleral patch grafting.¹²

This case differs from the existing cases described in the literature. Although the existing two reports of this condition have been successfully treated with relative ease, the

Learning points

- ▶ The incidence of retinal detachment is higher in patients with MFS than in the normal population. The repair of retinal detachment in these patients may involve the use of a scleral explant or buckle.
- ▶ Patients with MFS have abnormal sclera. This predisposes them to scleral perforation and resulting hypotony.
- ▶ Scleral perforation may occur at incisional sites or secondary to intrusion of the silicone explant through the sclera into to globe.
- ▶ Scleral perforation may occur many years after the initial procedure, is challenging to manage and frequently requires scleral patch grafting.

treatment of this case proved rather more challenging, and the outcome was only partially successful. Unlike the case described by Deramo *et al*,⁹ primary suturing of the sclera, tenons and conjunctiva in layers was unsuccessful in treating the perforation, and the eye remained hypotonous. The next step was to use allogeneic sclera as described in the case reported by Rodríguez-Ares *et al*.¹¹ However, we did not concurrently transplant amniotic membrane. Whereas Rodríguez-Ares *et al* reported a successful outcome, the result of our using a scleral patch alone was only partially successful.

This case demonstrates the management of this condition in patients with MFS is extremely challenging and that in cases where primary suturing of the scleral defect is impossible or unsuccessful, scleral patch grafting alone may not be sufficient to restore scleral integrity.

Competing interests None.

Patient consent Not obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Oyewole KT, Shortt AJ, Ramkissoon Y, Sullivan PM. Simultaneous extrusion and intrusion of a scleral buckle in a patient with Marfan's syndrome. *BMJ Case Reports* 2011;10.1136/bcr.08.2009.2182, date of publication

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