

Combined Transcranial-supraorbital and Transconjunctival Approach for Optic Nerve Coloboma with Ophthalmic Dysplasia Associated with Rheumatoid Arthritis

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We report a 59-year-old woman with optic nerve coloboma and ophthalmic dysplasia associated with rheumatoid arthritis. She experienced progressive visual dysfunction over the course of several years and presented with headache and pain in the left eye. Since infancy the visual acuity of her left eye had been compromised and her eyesight worsened gradually until she was blind in the left eye. Macroscopic observation showed a reddish lesion on the sclera thought to be due to rheumatoid arthritis (RA). Magnetic resonance imaging and computed tomography disclosed a well-defined cystic lesion at the left retro-bulbar optic nerve within the optic nerve sheath. We selected the combined transcranial-supraorbital and transconjunctival approach to remove the eyeball after detaching the optic nerve. This technique was successful and the placement of an ocular prosthesis was cosmetically acceptable.

Keywords: optic nerve coloboma, ophthalmic dysplasia, cloudy cornea, ophthalmectomy, ocular prosthesis

Introduction

Ocular coloboma is a rare congenital anomaly; its annual incidence is 2.4 cases per 100,000 population.¹⁾ It is attributable to incomplete closure of the embryonic fissure during the development of the optic cup. The resulting malformation can be a notch, gap, hole, or fissure in any of the ocular structures including the iris, zonulas and ciliary body, choroid, retina, and optic nerve.¹⁾ Visual impairment is most severe in

patients with optic-nerve colobomas; they can elicit complications such as subretinal exudation, extraretinal neovascularization, peripapillary choroidal neovascularization, and tractional retinal detachment.²⁾ The retinal detachment associated with optic nerve colobomas is typically extensive and bullous. Surgical repair is difficult^{2,3)} and enucleation may be indicated in patients whose blind eye is painful.⁴⁾ Scleral perforations have been observed in patients with systemic inflammatory conditions such as rheumatoid arthritis (RA)⁵⁾ which is characterized by antiglobulin antibody (rheumatoid factor, RF) in the systemic circulation. The main manifestation of RF is inflammation of the synovial membrane of the joints. Although the systemic accumulation of RF can result in damage to extra-articular organs, the eye is often damaged by RF. We encountered a patient with optic nerve coloboma and scleritis related to RA. She suffered traction retinal detachment and underwent ophthalmectomy via the combined transcranial-supraorbital and transconjunctival approach.

Case Report

The patient provided informed consent for the intended procedures; the expected treatment outcome was explained. She was 59 years old when she presented with compromised left-eye vision and severe pain. She had a past medical history of RA and was treated with methotrexate and non-steroidal anti-inflammatory drugs for pain in- and swelling of the wrist joint. Both were well-controlled by the medications. At the time of diagnosis, scleritis of the left eye was observed and she was referred to an ophthalmologist. Her left eyesight and pain gradually worsened and she developed clouding of the left cornea. Magnetic resonance imaging (MRI) performed elsewhere by a neurosurgeon revealed cystic lesions in the left orbit and she was referred to the ophthalmology department of our hospital. MRI and ocular ultrasound showed a cystic lesion communicating with the left eyeball. The diagnosis was coloboma accompanied by ophthalmic scleritis, retinal detachment, and bullous keratopathy related to RA. We referred her to our neurosurgery department for the treatment of painful bullous keratopathy attributable to elevation of the intraocular pressure accompanied by coloboma. Although coloboma can be an inherited autosomal disorder, she had no relevant family history. Her left cornea was cloudy (Fig. 1A) and the severity of her left eye pain had recently increased. Macroscopically there were reddish nodules thought to be due to acute scleritis.

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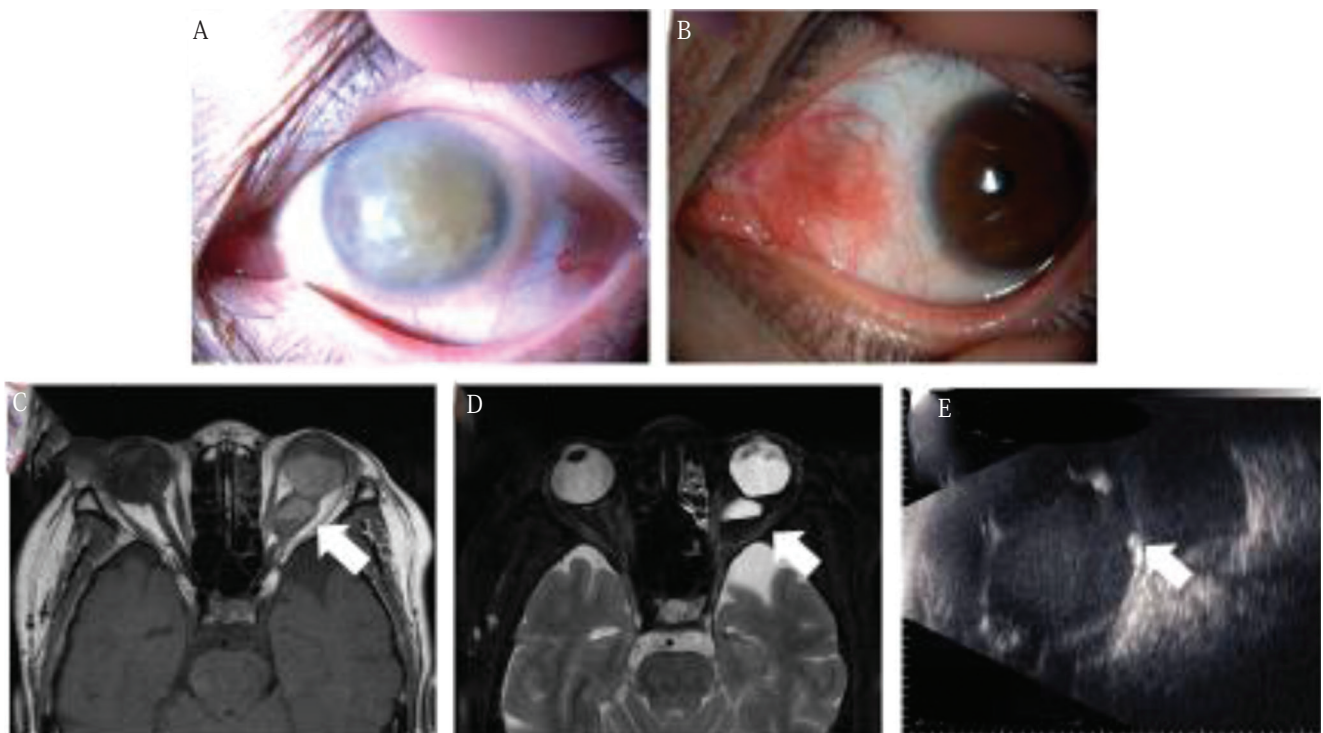


Fig. 1 Preoperative findings (left eye). (A) Cloudy cornea. (B) Macroscopically, a scleral lesion is seen around the limbus. The reddish nodules are thought to represent acute scleritis with marked scleral injection and edema. (C) Axial T1-weighted image (WI) showing an intraorbital high-intensity mass and a cystic mass behind the eyeball. Note the area of high intensity inside the optic nerve sheath (arrow). (D) Axial T2-WI revealing an intraorbital high-intensity mass reflective of bleeding (arrow). (E) Ultrasound image showing a hypoechoic lesion communicating on the posterior side of the eyeball (arrow).

We also observed marked scleral injection and edema (Fig. 1B). US revealed a hypoechoic lesion communicating on the posterior side of the left eyeball; we thought it to be the site of the optic nerve. Repeat MRI confirmed the presence of a thickened, cystic optic nerve and denaturation of the left eyeball (Figs. 1C and 1E).

As she was a candidate for ophthalmectomy and the placement of an ocular prosthesis, we sought the cooperation of neurosurgeons and ophthalmologists for planned retrobulbar surgery. As she refused the placement of a skin incision on her face, we chose the combined transcranial-supraorbital and transconjunctival approach to remove the eyeball after detaching the optic nerve. A neurosurgeon performed craniotomy and orbitotomy, and a retrobulbar intraorbital maneuver. After minimum shaving of the hair, a curvilinear frontotemporal skin incision was made and the temporal muscle was peeled off; the facial nerve was preserved. Superior and lateral orbitotomy was performed after a small frontotemporal craniotomy via one burr hole. The periorbita was opened under a surgical microscope, the enlarged optic nerve sheath was identified, and orbital fat was dissected (Figs. 2A and 2B). The cyst was punctured and blood components in xanthochromic fluid were aspirated. This decreased the volume of the optic nerve sheath. We then exposed the optic nerve sheath as far as possible to the orbital conus and cut the sheath and the atrophied optic nerve on the proximal side of the cyst.

The scalp was closed and the ophthalmologist performed transconjunctival eyeball removal under a microscope. After

incising the bulbar conjunctiva and before disconnecting each rectus muscle above, below, inside, and outside, the lower and superior oblique muscles were separated and dissected. The optic nerve sheath was detached in the conical direction and the cyst and the optic nerve sheath were removed. The inner, outer, upper, and lower rectus muscles were sutured consecutively. The bulbar conjunctiva was sutured to form the floor for the ocular prosthetic placed in the prosthetic bed; the eyelids were temporarily sutured. Subsequently, neurosurgeons released the temporary scalp suture and closed the wound. The periocular membrane was sutured as much as possible. Titanium plates were affixed to the orbital rim and skull, the bone defect was filled with hydroxyapatite, and the suture was closed.

The pathologists reported that the cystic mass was 1.2 cm in length; its diameter at each end was 1.0 cm and 1.0 mm. (Figs. 2C and 2D) Microscopically, the cystic lesion was attached to the optic nerve. The cyst wall consisted of fibrous connective tissue; some macrophages and pigmentation were observed. The retina was atrophic and the optic nerve was denatured (Figs. 3B–3E). These findings are compatible with coloboma. Postoperative MRI confirmed removal of the eyeball and coloboma. She was satisfied with the outcome and the aesthetic appearance of the ocular prosthesis (Fig. 3A).

Discussion

Our patient suffered RA; the systemic accumulation of RF can damage extraarticular organs. Ocular manifestations of RA

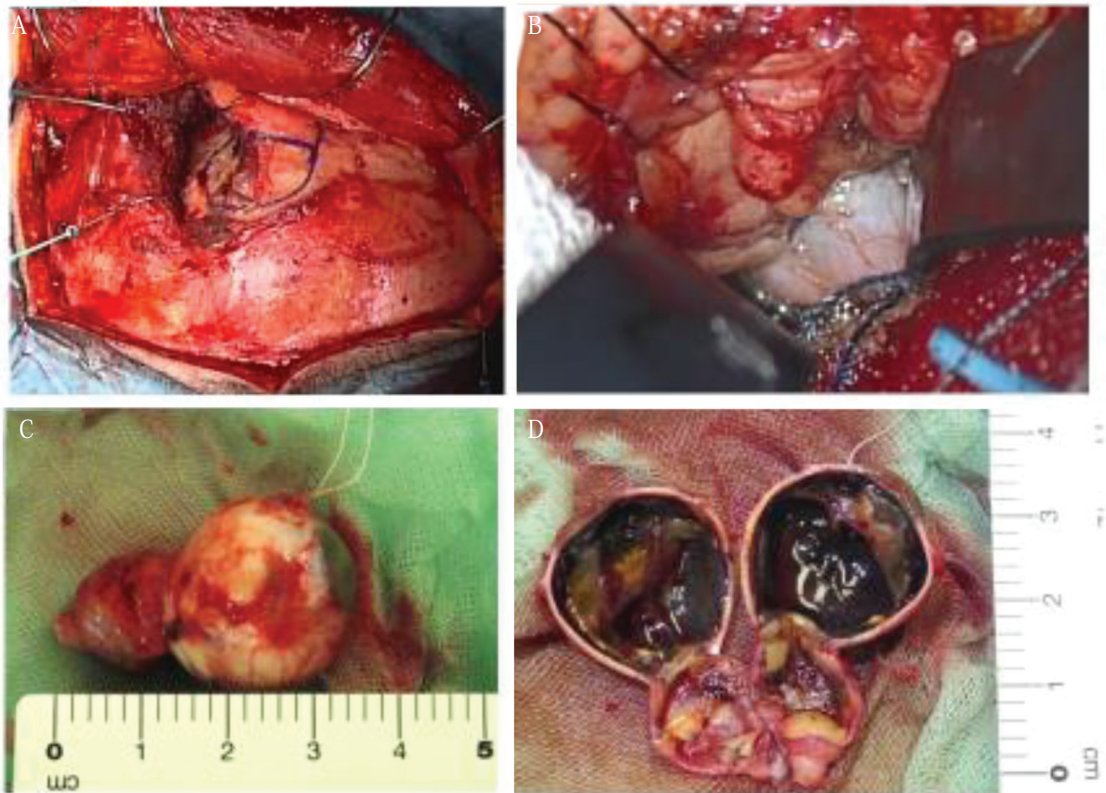


Fig. 2 Surgical procedures. (A) Small craniotomy with one burr hole. (B) Dissection around the structure revealed enlargement of the optic nerve sheath. (C) Gross appearance of the removed left eyeball and the colobomatous cyst. (D) Section along the long axis.

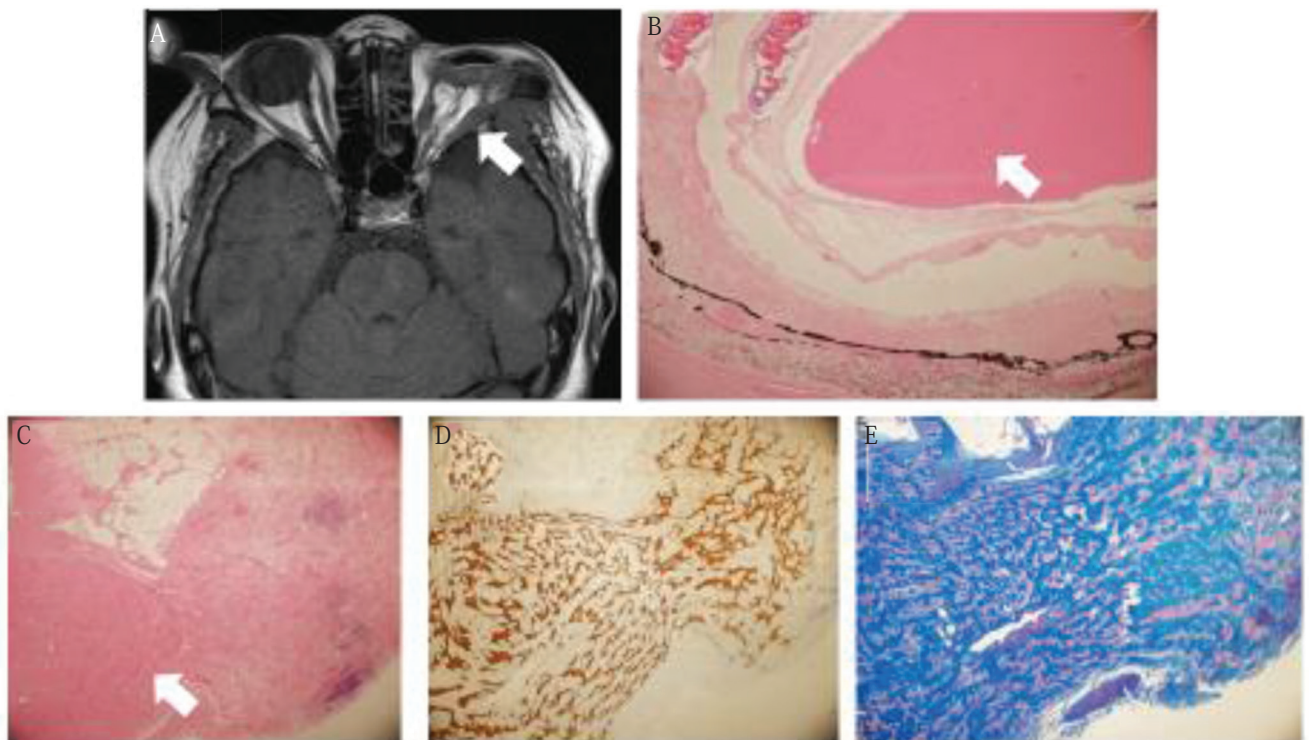


Fig. 3 Postoperative findings and pathology. (A) Axial T1-weighted image. The arrow points to the site of the coloboma. (B) Hematoxylin-eosin (H&E)-stained tissue (40 \times). The coloboma cyst contains blood components (arrow). (C)–(E) Cystic lesion attached to the denatured optic nerve (arrow) and the proliferation of collagen. (F) H&E stain (20 \times). (G) Immunohistochemical stain for Glial fibrillary acidic protein (20 \times). This is positive for astroglia. (H) Immunohistochemical stain for Azan (20 \times). This stains collagen blue.

that can result in severe disability and even blindness are keratoconjunctivitis sicca, episcleritis, scleritic corneal changes, and retinal vasculitis; the most common is rheumatoid scleritis.⁶⁻¹⁰⁾

As she experienced gradually worsening visual disturbance since childhood, we think that a slight retrobulbar structure defect present since birth elicited the formation of a colobomatous cyst. Her RA diagnosed in adulthood brought on scleritis, retinal detachment, and keratitis and severe pain. Because visual acuity depends on the degree of retinal and choroidal involvement, we suspect that her total visual loss was attributable to the detachment of the retinal layer due to the cystic mass and that RA was a contributing factor. Retinal detachment associated with optic nerve colobomas is typically extensive and bullous. A defect in anomalous papillary tissue allows fluid to enter the subretinal space, resulting in retinal detachment.^{2,3)} Because her left vision had been compromised since childhood, the retention of retinal attachments by repairing the defect in the presence of a coloboma was not a good treatment option. Therefore, we aimed at eliminating the cause of her pain, at stopping further damage, and at obtaining a cosmetically satisfactory outcome. According to Moshfeghi et al.,⁴⁾ surgical removal of the affected eye and the attached cyst is the preferred treatment in patients such as ours.

Ocular enucleation is the last resort in patients with end-stage ocular diseases, e.g. ocular malignancies or other pathologies leading to a painful blind eye.¹¹⁾ However, the psychological sequelae of ocular enucleation must be considered. The procedure involves the surgical removal of the entire eyeball with preservation of the conjunctiva, extraocular muscles, orbital fat, and optic nerve. Painful blind eye is one of the most common indications for enucleation after trauma and in patients with ocular tumors.¹²⁾ Neurosurgeons and ophthalmologists participated in our patient's surgery. When lesions beyond a certain size are located retrobulbarly, their *en bloc* resection via the transconjunctival approach is difficult unless the cyst volume is first reduced.

Dandy¹³⁾ reported a neurosurgical approach to the orbit. Depending on the tumor location, surgical-, including medial-, central-, lateral-, and fronto-orbital approaches are used to remove orbital tumors.^{3,14)} Sacks and McLennan¹⁵⁾ selected the transcranial approach for eye removal in patients with optic nerve tumors.

We applied a combined transcranial-supraorbital and transconjunctival approach to remove the orbital structure *en bloc* because our patient refused a facial skin incision. Ophthalmologist favor *en bloc* removal because it facilitates the creation of an optimal tissue bed for the ocular prosthesis. Hernesniemi et al.¹⁶⁾ suggested a modified lateral supraorbital approach as an alternative to the classical pterional approach. Our transcranial-supraorbital approach involved a small

craniotomy with one burr hole. It yielded adequate retrobulbar exposure and rendered neurosurgical maneuvers safe. The appropriate approach is dictated by craniometrics or anatomical characteristics. We encountered no intra- or post-operative complications. At the last follow-up, our patient reported to be free of pain and satisfied with the cosmetic result of our treatment.

Informed Consent

The patient consented to the submission of this case to the journal.

Conflicts of Interest Disclosure

None of the author declares a conflict of interest.

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