Surgical management for IgG4-related ophthalmic disease by a transcranial biopsy combined with extraorbital decompression: illustrative case

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BACKGROUND  Immunoglobulin G4–related ophthalmic disease (IgG4-ROD) accounts for 22% of Japanese lymphoproliferative orbital disease and occurs in 4%–34% of patients with IgG4-related disease, according to the largest case series published to date. Because the optic nerve mass often appears as a tumor-like lesion, it is important, although difficult, to differentiate IgG4-ROD from other orbital tumors and diseases, and biopsy is essential for diagnosis. Here, the authors describe the surgical management of an IgG4-ROD case.

OBSERVATIONS  A 63-year-old man presented to the authors' hospital with proptosis and visual impairment. Ophthalmic examination revealed intraocular hypertension. IgG4-related disease with an ophthalmic lesion was suspected on the basis of a blood test and imaging studies. Transcranial biopsy with extraorbital decompression was performed. The patient's symptoms, including visual impairment, improved 3 days after operation, and his IgG4-related disease resolved after corticosteroid treatment.

LESSONS  The standard treatment for IgG4-related disease is systemic corticosteroid therapy. However, this treatment should not be administered to patients with IgG4-ROD who have a high risk of blindness. In this case, the authors completed a diagnostic and symptom-relieving transcranial biopsy without affecting the patient's aesthetic characteristics. This is the first study, to our knowledge, to report extraorbital decompression via a transcranial approach as a surgical option for IgG4-ROD.

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KEYWORDS  IgG4-related disease; IgG4-related ophthalmic diseases; transcranial orbital approach; orbital tumor; extraorbital decompression
He had recently noticed a relatively rapid aggravation of proptosis and conjunctival injection. Magnetic resonance imaging (MRI) showed a mass in the right orbit. Physical examination revealed proptosis, ptosis, and abduction restriction of the right eye. Ophthalmic examination revealed right visual acuity of 0.7 LogMAR (logarithm of the minimum angle of resolution) in the right eye compared with ~0.1 LogMAR in the left eye. The patient’s intraocular pressures were 31 mm Hg and 15 mm Hg in the right and left eyes, respectively.

Contrast-enhanced MRI revealed a contrast-enhanced mass lesion around the right optic nerve sheath (Fig. 1). Contrast-enhanced computed tomography of the whole body showed systemic lymphadenopathy that manifested in the right lacrimal gland, bilateral parotid gland, mediastinal lymph nodes, and right inguinal lymph nodes. Blood test results revealed elevated IgG4 concentrations up to 1255 mg/dL. On the basis of the above findings, IgG4-ROD was suspected. For a definitive diagnosis and to alleviate symptoms, we performed biopsy via the transcranial approach (Video 1).

The surgery was performed with the patient under general anesthesia. Craniotomy was performed using Yaşargil’s pterional method. The patient was placed supine. After head fixation in a Mayfield clamp, the head was rotated 30° contralaterally. The skin incision began at the level of the zygomatic arch at the preauricular region (1 cm anterior to the tragus) and curved along the hairline until 1–2 cm from the midline. The curve was slightly (approximately 1 fingerbreadth) augmented posteriorly at the supra-auricular level to create space for the temporal muscle to retract posteriorly (Fig. 2A). The temporal muscle was incised through the plane between the laminae of the temporal fascia to preserve the frontal branch of the facial nerve. The cutaneous flap was inferiorly reflected. The muscle flap was incised along the linear temporalis, except for the posterior 3-fingerbreadth region, and pushed posteriorly to expose the base of the temporal bone. Craniotomy was designed using 3 burr holes on the cranium (MacCarty’s keyhole, 1 hole anterior to the squamous suture at the level of the zygomatic root, and 1 hole inferior to the linear temporalis line; Fig. 2B). The sphenoid ridge was drilled off, and the lateral wall of the orbit was flattened. After elevating the dura propria, with minimal retraction of the dura, we performed clinoidectomy of the anterior clinoid and drilled out the optic canal. The superior and lateral walls of the orbit were sufficiently removed (Figs. 2C and 3A). The periorbital sheath was incised, and biopsy was performed between the lateral rectus and superior rectus muscles (lateral approach). The orbital sheath was reconstructed with temporal fascia to decrease the intraocular pressure. The histopathological results were consistent with IgG4-related disease. The superior and lateral walls were left open for decompression (Fig. 3B). The bone flap was put back in its original position and fixed to the skull with titanium plates and screws. Calcium phosphate bone paste was used to cover the bone defects. The edges of the temporalis fascia and those of the temporal muscle were sutured to the original position, and the skin was also sutured.

Ophthalmological examination performed 3 days after the operation showed dramatic improvement in visual acuity (~0.1 LogMAR in the right eye). Intraocular pressure had decreased to 15 mm Hg. Postoperative MRI showed significant improvement in proptosis (Fig. 4). After confirming the IgG4-ROD diagnosis on the basis of histopathological results, oral administration of corticosteroids was initiated 7 days postoperatively and continued after discharge. The lesion showed gradual resolution after treatment. During the 6-month follow-up, complete remission was attained, and no recurrence was shown.

**Discussion**

**Observations**

Although the treatment of IgG4-ROD is hampered by a lack of prospective randomized clinical trials, corticosteroid therapy is
reported to be a treatment option.\textsuperscript{8,9} To commence this treatment, a definitive diagnosis must be achieved on the basis of histopathological examination. Thus, biopsy from the most accessible lesion and in the least invasive manner is usually considered. In addition, although corticosteroid treatment is reported to result in a prompt response in IgG4-ROD cases, several weeks are required for remission,\textsuperscript{7,10} and there are concerns about the relatively high recurrence rate (18\%–58\%).\textsuperscript{11} Moreover, in IgG4-ROD with ocular hypertension, corticosteroid treatment could aggravate symptoms by increasing the intraocular pressure.\textsuperscript{12} We could not risk exacerbating the ocular hypertension and causing blindness as a result. Therefore, in cases such as ours, it is important to relieve the optic nerve from intraocular pressure as soon as possible to improve symptoms and prevent permanent nerve damage.

Ominato et al.\textsuperscript{13} reported a case series of patients with IgG-ROD treated solely with surgical debulking. The results showed that debulking surgery could be a treatment option for IgG4-ROD. However, because of the lack of accumulated investigations and properly designed studies, the efficacy of removing or debulking the tumor remains questionable.\textsuperscript{14} In our case, mass reduction was not necessary. Considering the risk of complications, we decided to avoid debulking surgery. In contrast, an extraorbital decompression was considered effective and feasible. In fact, it has already been reported that medial

\textbf{FIG. 2.} A: Scheme of skin incision and temporal muscle incision. The skin incision began from the preauricular region at the level of the zygomatic arch. The curve of the incision was slightly (approximately 1 fingerbreadth) augmented posteriorly at the supra-auricular level to create space for the posterior retraction of the temporal muscle, and it was ended at 1–2 cm from the midline (\textit{red line}). B: Scheme indicating the direction (arrows) of retraction of the skin flap and that of the temporal muscle. The cutaneous flap was reflected inferiorly. The muscle flap was incised along the linear temporals, except for the posterior 3-fingerbreadth region (\textit{green line}), and pushed posteriorly to expose the base of the temporal bone. Craniotomy was designed by creating 3 burr holes on the cranium (MacCarty’s keyhole, 1 hole posterior to the squamous suture on the level of the zygomatic root, and 1 hole inferior to the linear temporals line, \textit{blue line}). C: Scheme of the extraorbital decompression. The lateral and superior orbital walls were drilled off (\textit{blue highlighted area}).

\textbf{FIG. 3.} Postoperative bone computed tomography (CT). Comparison between pre- and postoperative axial CT images of bone (A). Lateral wall of the orbit is drilled off. Posterior view of three-dimensional bone image showing the lateral wall and orbital roof were removed (B).
The invasiveness, especially in terms of cosmesis, was our concern. The transcranial approach for orbital tumors was initially developed by Dandy in 1941 and has been modified ever since. The current concept of the transcranial orbital approach is a frontotemporal (FT) craniotomy with a combination of orbitectomy and zygomectomy. The reconstruction of pterional, temporal, and zygomatic walls of the orbit was performed via a simple FT craniotomy; orbitectomy and zygomectomy were avoided. The patient had no objective aesthetic deterioration, and the results were also subjectively satisfactory.

Lessons

In conclusion, we could complete a diagnostic and symptom-relieving transcranial biopsy without affecting the patient’s aesthetic characteristics. In patients with IgG4-ROD in critical condition, extraorbital decompression might be an option before administering systemic corticosteroid to prevent blindness. Furthermore, this method could be a possible strategy for any kind of orbital tumors with elevated intraocular pressure.

To the best of our knowledge, this is the first study to report extraorbital decompression and biopsy via the transcranial approach as a surgical technique for IgG4-ROD. This method can be applied in patients with severe visual impairment and elevated ocular pressure.

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Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Supplemental Information
Videos
Video 1. https://vimeo.com/498368462.

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