

EP-018

불응성 면역글로블린 G4 관련 질환에 동반된 중증 안구돌출의 삼벽 안와감압술 치료

(Severe Exophthalmos in Refractory Immunoglobulin G4-Related Disease Treated with Three-Wall Orbital Decompression)



인제대학교 해운대백병원

김동균, 윤성호, 선욱, 이경아*

Purpose: Immunoglobulin G4-related disease (IgG4-RD) is a systemic fibro-inflammatory disorder that frequently involves orbital tissues, leading to progressive exophthalmos. Although systemic corticosteroid therapy is the primary treatment, surgical intervention may be required for patients who remain refractory to medical management. We report a case of severe, medically refractory exophthalmos in IgG4-RD that was successfully managed with three-wall orbital decompression following a relapse during the transition from steroids to immunosuppressants.

Methods: A 62-year-old male presenting with proptosis underwent excisional biopsies of suspected xanthelasma, which histopathologically confirmed IgG4-RD. While initial steroid therapy provided clinical improvement, the subsequent tapering of steroids and initiation of steroid-sparing immunosuppressants resulted in disease flare-up. Preoperative exophthalmometry revealed measurements of 21 mm (right) and 18 mm (left), with orbital computed tomography confirming significant tissue expansion (Fig. 1). Due to the progressive nature of the deformity, a bilateral three-wall orbital decompression was performed (Fig. 2).

Results: The postoperative course was uneventful without any neuro-ophthalmic complications. At the 3-week follow-up, exophthalmometry demonstrated a marked reduction to 14 mm bilaterally. During the 3-month outpatient follow-up period, the patient maintained stable orbital volume and expressed high satisfaction with the aesthetic and functional outcomes (Fig. 3).

Conclusion: While systemic medical therapy remains the gold standard for IgG4-RD, three-wall orbital decompression should be considered a viable and effective therapeutic adjunct for patients with severe exophthalmos that is refractory to conventional immunosuppressive regimens.

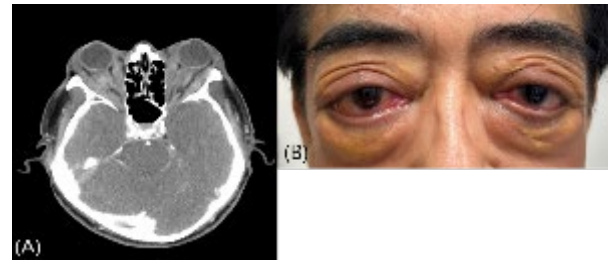


Figure 1. Preoperative clinical and radiological assessment of medically refractory IgG4-related orbital disease. (A) Axial view of preoperative orbital computed tomography (CT) showing significant retrobulbar tissue expansion and resultant marked exophthalmos. (B) Preoperative clinical photograph illustrating prominent bilateral proptosis, measured at 21 mm on the right and 18 mm on the left.



Figure 2. Intraoperative findings during bilateral three-wall orbital decompression. Intraoperative photograph demonstrating the surgical exposure and decompression process. The procedure involved extensive removal of the orbital walls and incision of the periorbita to allow for the prolapse of orbital fat and reduction of intraorbital pressure.



Figure 3. Postoperative clinical outcome at 3 weeks following orbital decompression. Follow-up clinical photograph demonstrating a significant reduction in proptosis (14 mm bilaterally) and a stable postoperative appearance. The patient showed high aesthetic and functional satisfaction without neuro-ophthalmic complications.