Exophthalmos

Exophthalmos (also called exophthalmus, exophthalmia, proptosis, or exorbitism) is a bulging of the eye anteriorly out of the orbit.

Classification

Exophthalmos can be either bilateral (as is often seen in Graves' disease) or unilateral (as is often seen in an orbital tumor). Complete or partial dislocation from the orbit is also possible from trauma or swelling of surrounding tissue resulting from trauma.

Non-Pulsatile Exophthalmos.

Pulsatile Exophthalmos.

Differential diagnosis

In the case of Graves' disease, the displacement of the eye is due to abnormal connective tissue deposition in the orbit and extraocular muscles which can be visualized by CT or MRI.

Complications

If left untreated, exophthalmos can cause the eyelids to fail to close during sleep leading to corneal dryness and damage. Another possible complication would be a form of redness or irritation called "Superior limbic keratoconjunctivitis", where the area above the cornea becomes inflamed as a result of increased friction when blinking. The process that is causing the displacement of the eye may also compress the optic nerve or ophthalmic artery, leading to blindness.

Meningioma-associated proptosis (MAP) can be cosmetically and functionally debilitating for patients with sphenoorbital meningioma and other skull base meningiomas, and there is limited information on the quantitative improvement in proptosis after surgery. Because less extensive removals of tumor involving the orbit fail to reduce proptosis, Bowers et al., has adopted an aggressive surgical approach to the removal of tumor involving the periorbita and orbit. The authors of this study retrospectively reviewed outcomes of this surgical approach.

All surgeries for MAP performed by a single surgeon between January 1, 2002, and May 1, 2015, were reviewed. Age, sex, visual symptoms, number and types of surgical treatments, cavernous sinus involvement, complications, duration of follow-up, residual tumor, use of adjuvant radiation therapy, and extent of proptosis resolution as measured by the exophthalmos index (EI) pre- and postoperatively and at the final follow-up were recorded.

Thirty-three patients (24 female [73%]) with an average age of 51.6 years were treated for MAP. Of

the 22 patients with additional visual symptoms (for example, loss of visual acuity, field cut, or diplopia), 15 had improved vision and 7 had stable vision. No patients had worse proptosis after treatment. The average preoperative El was 1.39, the average immediate postoperative El was 1.23, and the average final El at the most recent follow-up was 1.13. Thus, average overall El improvement was 0.26, but the average immediate El reduction was 0.16, demonstrating that proptosis progressively improved during the postoperative period. Residual cavernous sinus involvement was present in 17 of 18 patients who had had preoperative cavernous sinus meningioma involvement. Only 2 patients in the series had recurrent tumor at the orbital region, and their proptosis improved again after reoperation. One case of delayed vasospasm and 2 cases of postoperative trigeminal numbness (V2) were recorded. The average follow-up was 4.5 years (53.8 months).

In this series, all patients experienced proptosis improvement and none had worse visual symptoms at the final follow-up, although proptosis resolution occurred over time. Only 2 patients had tumor recurrence at the orbit that required surgery. Surgical complications were uncommon. Study results suggest that aggressive resection of MAP is well tolerated and offers superior proptosis elimination with infrequent recurrence at the orbit. Importantly, no cases of enophthalmos were noted despite the lack of formal reconstruction of the orbit ¹⁾.

Treatment

Heller et al., sought to quantitatively assess the effect of orbital volume before and after reconstruction to determine the optimal strategy to achieve proptosis correction.

All surgeries involving orbital wall reconstruction for orbital or intracranial pathology that preoperatively resulted in proptosis between 2007 and 2017 were reviewed. Proptosis was measured by the exophthalmos index (EI): the ratio of the distance of the anterior limit of each globe to a line drawn between the anterior limit of the frontal processes of the zygomas, comparing the pathological eye to the normal eye. Postoperative radiographic measurements were taken at least 60 days after surgery to allow surgical swelling to abate. The orbit contralateral to the pathology was used as an internal control for normal anatomical orbital volume. Cases with preoperative EI < 1.10, orbital exenteration, or enucleation were excluded.

Twenty-three patients (16 females and 7 males, with a mean age of 43.6 \pm 22.8 years) were treated surgically for tumor-associated proptosis. Nineteen patients harbored meningiomas (11 en-plaque; 8 sphenoid wing), and one patient each harbored an orbital schwannoma, glomangioma, arteriovenous malformation, or cavernous hemangioma. Preoperative El averaged 1.28 \pm 0.10 (range 1.12-1.53). Median time to postoperative imaging was 19 months. Postoperatively, the El decreased to a mean of 1.07 \pm 0.09. Greater increases in size of the reconstructed orbit were positively correlated with greater quantitative reductions in proptosis (p < 0.01). Larger volume of soft tissue pathology was also associated with achieving greater proptosis correction (p < 0.01). Residual exophthalmos (defined as El > 1.10) was present in 8 patients, while reconstruction in 2 patients resulted in clinically asymptomatic enophthalmos (defined as El < 0.95). Tumor invasion into the superior orbital fissure sinus was associated with residual proptosis (p = 0.04).

Proptosis associated with intracranial and orbital pathology represents a surgical challenge. The EI is a reliable and quantitative assessment of proptosis. For orbital reconstruction in cases of superior orbital fissure involvement, surgeons should consider rebuilding the orbit at slightly larger than anatomical volume ²⁾.

Case reports

Acquired proptosis and progressive abducens nerve palsy due to overpacked coiling material: rare sequelae of carotid cavernous fistula embolization ³⁾.

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Bowers CA, Sorour M, Patel BC, Couldwell WT. Outcomes after surgical treatment of meningiomaassociated proptosis. J Neurosurg. 2016 Sep;125(3):544-50. doi: 10.3171/2015.9.JNS15761. Epub 2016 Jan 22. PubMed PMID: 26799293.

Heller RS, David CA, Heilman CB. Orbital reconstruction for tumor-associated proptosis: quantitative analysis of postoperative orbital volume and final eye position. J Neurosurg. 2019 Mar 8:1-6. doi: 10.3171/2018.12.JNS181385. [Epub ahead of print] PubMed PMID: 30849755.

Teoh RJJ, Ain Masnon N, Bahari NA, Ch'ng LS. Acquired proptosis and progressive abducens nerve palsy due to overpacked coiling material: rare sequelae of endovascular treatment for carotid cavernous fistula. BMJ Case Rep. 2023 Oct 10;16(10):e255406. doi: 10.1136/bcr-2023-255406. PMID: 37816571.

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