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Case Reports
Trauma

Orbital apex disorders: a case series

R. E. Warburton, C. C. D. Brookes, B. A. Golden, T. A. Turvey
Department of Oral and Maxillofacial Surgery, School of Dentistry, University of North Carolina at Chapel Hill, Chapel Hill, NC, USA

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Abstract. Orbital apex syndrome is an uncommon disorder characterized by ophthalmoplegia, proptosis, ptosis, hypoesthesia of the forehead, and vision loss. It may be classified as part of a group of orbital apex disorders that includes superior orbital fissure syndrome and cavernous sinus syndrome. Superior orbital fissure syndrome presents similarly to orbital apex syndrome without optic nerve impairment. Cavernous sinus syndrome includes hypoesthesia of the cheek and lower eyelid in addition to the signs seen in orbital apex syndrome. While historically described separately, these three disorders share similar causes, diagnostic course, and management strategies. The purpose of this study was to report three cases of orbital apex disorders treated recently and to review the literature related to these conditions. Inflammatory and vascular disorders, neoplasm, infection, and trauma are potential causes of orbital apex disorders. Management is directed at the causative process. The cases described represent a rare but important group of conditions seen by the maxillofacial surgeon. A review of the clinical presentation, etiology, and management of these conditions may prompt timely recognition and treatment.

Key words: orbital apex syndrome; superior orbital fissure syndrome; cavernous sinus syndrome; orbital compartment syndrome; ptosis; proptosis; ophthalmoplegia; optic neuropathy; maxillofacial surgery.

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Orbital apex syndrome (OAS) is an uncommon disorder characterized by impairment of cranial nerves III, IV, VI, and the ophthalmic branch of cranial nerve V, and optic neuropathy. Patients present with ophthalmoplegia, proptosis, ptosis, visual impairment, a fixed dilated pupil, and hypoesthesia of the ipsilateral forehead, upper eyelid, and cornea.[1] Superior orbital fissure syndrome (SOFS) presents similarly to OAS, without the accompanying optic nerve impairment.[2,3] Cavernous sinus syndrome (CSS) involves palsy of cranial nerves III, IV, and VI, optic neuropathy, oculosympathetic paresis, and impairment of the ophthalmic and maxillary branches of cranial nerve V.[1] CSS presents with ophthalmoplegia, ptosis, proptosis, decreased vision, and loss of sensation in the ipsilateral forehead, eyelids, cornea, and cheek.[4,5] CSS can present bilaterally.[1] Cranial nerve involvement may be complete or incomplete in all three syndromes.

While frequently described separately in the literature, these three orbital apex disorders share a similar etiology, diagnostic course, and treatment.[1,6] These syndromes can be progressive in nature, with SOFS developing into OAS or CSS.[1] For the purpose of discussion OAS, SOFS, and CSS can be grouped together as a single condition, differentiated chiefly by the anatomical position of the causative pathology.

Three new cases of orbital apex disorders are reported to illustrate the three syndromes and highlight several key causative factors. The relevant anatomy, etiology, and available diagnostic and treatment modalities are also described.

This study was reviewed by the Institutional Review Board and deemed exempt.

Case series

Case 1

A 16-year-old Caucasian male with marked midfacial hypoplasia, retrogenia, and right-sided hemihypoplasia presented for a Le Fort III osteotomy modified to maintain nasal position, right split thickness parietal bone harvest with grafting to the bilateral supraorbital rims and midface, and genioplasty. Preoperative ophthalmic evaluation revealed normal vision (20/20 in the right eye (OD) and 20/25 in the left eye (OS)) with no afferent pupillary defect or anisocoria.

The patient was administered 125 mg of methylprednisolone and 1 g of cefazolin intravenously at the start of the procedure and every 4 h subsequently. The midface was mobilized and advanced without difficulty. The inner cortex remained intact during the parietal bone harvest. The orbital osteotomies were located within 1 cm of the orbital rim and carried into the inferior orbital fissure. No fracture or extension of the osteotomies to the orbital apex was observed.

Upon completion of the procedure, the right pupil was noted to be fixed and dilated with a 4-mm discrepancy when compared to the left pupil. Bilateral pupils were non-reactive, which was attributed to the depth of anesthesia. The coronal and intraoral incisions were reopened and the right orbit was explored, with no hematoma or pulsatile bleeding noted. Ophthalmology was consulted to help evaluate for an intraorbital cause of the patient's anisocoria. Intraocular pressures were found to be normal at 15 mmHg OD and 20 mmHg OS by handheld tonometry. The neurosurgery service was consulted to rule out any intracranial pathology. An emergency computed tomography (CT) of the head and face was obtained, which revealed no intracranial hemorrhage, midline shift, or brainstem compression. No fracture or hematoma involving the superior orbital fissure or orbital apex was noted (Fig. 1). The patient's surgical wounds were then closed and he was transferred to the pediatric intensive care unit for close neurological monitoring.

The patient's postoperative ophthalmic examination revealed complete right ophthalmoplegia, mild proptosis, and moderate ptosis. The right pupil was 6 mm and non-reactive to direct light, although the consensual response was intact. The patient initially reported blurred vision OD that improved through a pinhole occluder. Color vision was intact. He had binocular diplopia as well as anticipated cranial nerve V deficits. The remainder of his neurological examination was benign. Magnetic resonance imaging (MRI) of the brain and orbits was performed, which revealed no signs of orbital apex compression or intracranial pathology.

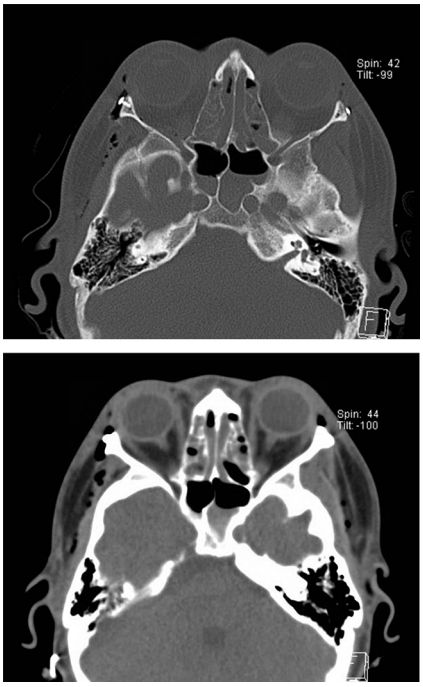

Fig. 1. Intraoperative CT scan revealing no fracture or hematoma involving the right orbital apex or optic canal.

The patient was diagnosed with a presumed SOFS. Intravenous methylprednisolone (125 mg) was continued every 4 h throughout his hospitalization. A formal ophthalmic evaluation at less than 24 h after surgery found visual acuity to be 20/40 OD, 20/20 OS. The patient underwent serial ophthalmic examinations, which showed slight improvement in ptosis and continued ophthalmoplegia. The patient was discharged on postoperative day 2 with a 7-day prednisone taper. At the time of discharge, the patient's vision was 20/30 OD, 20/20 OS with a 2-mm discrepancy in pupillary size and minimal reactivity to light OD.

Three weeks postoperatively, the patient showed improved extraocular movements and ptosis OD. The binocular diplopia and pupillary dilation remained. Five weeks postoperatively, the ptosis and ophthalmoplegia had improved greatly. The right pupil remained dilated with some residual binocular diplopia. At the 10-week follow-up, all symptoms had resolved.

Case 2

A 15-year-old Caucasian male with asthma and chronic rhinosinusitis presented to clinic with a 4-day history of right midfacial pain and swelling without orbital involvement. He was admitted to the hospital with a presumed facial infection of unknown cause for further work-up and intravenous antibiotics. Shortly after admission, the patient developed rapid onset of ophthalmoplegia, proptosis, ptosis, and complete vision loss OD. The right pupil was found to be fixed and dilated. Ophthalmology was consulted and the patient was found to have an intraocular pressure of 57 mmHg OD by handheld tonometry. The patient was diagnosed with an OAS secondary to an orbital compartment syndrome, and an emergency lateral canthotomy with inferior cantholysis was performed bedside. Intraocular pressure improved to 38 mmHg OD after the procedure. Vision loss and ophthalmoplegia were unchanged. Brimonidine and dorzolamide/timolol were administered topically and an emergency CT of the face was obtained. Imaging revealed a large cystic mass in the right maxillary sinus with erosion of the posterior maxillary sinus wall and right orbital cellulitis with posterior globe tenting (Fig. 2).[7]

The patient was taken as an emergency to the operating room for drainage of the right maxillary sinus via a Caldwell–Luc approach and further orbital decompression. Otolaryngology was contacted to determine the benefit of concomitant endoscopic sinus surgery, but the decision was made to defer additional sinus intervention at that time. After drainage of the maxillary sinus, the right intraocular pressure improved to 16 mmHg by handheld tonometry and no additional orbital decompression was pursued. Cultures were obtained and the maxillary sinus contents were sent for histopathological evaluation.

Postoperative visual acuity improved to 20/200 OD. The right pupil was sluggishly reactive. Ophthalmoplegia was unchanged. The patient was started on vancomycin and piperacillin/tazobactam. Maxillary sinus cultures revealed no growth. The intraoperative pathology results returned as sinonasal mucosa with features consistent with acute on chronic rhinosinusitis. Serial ophthalmic examinations revealed improved visual acuity and pupillary reactivity, although extraocular motility deficits persisted.

The patient was discharged to home on postoperative day 5 with a 7-day course of amoxicillin/clavulanic acid. Visual acuity was 20/25 + 2 OD, 20/20 OS at the time of discharge. Subsequent ophthalmology follow-up at 1 month revealed resolution of symptoms except for a persistent right inferior arcuate defect. The patient