MEGA JOURNAL OF CASE REPORTS

OPEN ACCESS

Case Report

Compiled Date: May 15, 2024

Orbital Apex Syndrome without Facial Fractures: A Case Report and Review of Literature

Amin Hasheminia[1]*, Michael A Malik[2], Majid Rezaei[3], Tirbod Fattahi[4], Navid Ahmady Roozbahany[5]

[1]Department of Biomedical and Molecular Sciences, Queen's University, Kingston, Canada

[2]Department of Oral & Maxillofacial Surgery, College of Medicine, University of Florida Health, Jacksonville, Florida, USA

[3]Department of Oral & Maxillofacial Surgery, College of Medicine, University of Florida Health, Jacksonville, Florida, USA

[4]Department of Oral & Maxillofacial Surgery, College of Medicine, University of Florida Health, Jacksonville, Florida, USA

[5]Private Practice, Toronto, Canada

Corresponding author: Amin Hasheminia, Department of Biomedical and Molecular Sciences, Faculty of Health Sciences, Queen's University, 18 Stuart Street Kingston, Canada, Tel: +1 (416) 676-4543

Abstract

Orbital Apex Syndrome (OAS) is an extremely rare yet significant complication following craniomaxillofacial trauma. While its pathogenesis is still being explored, the syndrome's clinical features predominantly involve a combination of vision loss and ophthalmoplegia. Despite the severity of its implications, there is scant literature addressing traumatic OAS without associated facial bones fracture. A comprehensive ISI Web of Knowledge, PubMed, Google Scholar, Scopus, Embase, and the Cochrane library search was performed along with presenting a case of isolated OAS. The main focus was on studies presenting traumatic OAS with no facial bone fracture or severe traumatic brain injuries. Two studies (two patients total) met the inclusion criteria. Treatments ranged from observation only to high-dose corticosteroids and surgical optic canal decompression. However, visual acuity remained impaired regardless of type of treatment. A standardized protocol on a case-by-case basis is yet to be defined.

Keywords: Traumatic Orbital Apex Syndrome (OAS); Traumatic Optic Neuropathy (TON); Ophthalmoplegia; Traumatic Superior Orbital Fissure Syndrome; Isolated Traumatic OAS

Introduction

Orbital Apex Syndrome (OAS) is a complex condition characterized by a combination of vision loss and ophthalmoplegia due to damage to the structures within the orbital apex-a region rich with critical nerves and vessels supplying the eye and periorbital tissues. The cases of OAS with craniofacial bone fractures are well documented; however, the literature is particularly scant on traumatic OAS in the absence of severe traumatic brain injuries or craniofacial fractures. As a result, there is a lack of consensus on the underlying etiology, pathophysiology and optimal therapeutic approach for this subgroup of OAS. This paper seeks to bridge this gap by detailing the clinical course and management of a case of isolated traumatic OAS. Furthermore, a review of literature sheds light on the nature of isolated traumatic OAS and evaluates the implications of current treatment strategies. Aiming to enhance the clinical understanding of OAS, this study sets out to: 1) characterize the clinical presentation and progression of traumatic OAS without facial bone fractures; 2) evaluate the treatment options employed and their outcomes; and 3) propose recommendations for a more unified treatment protocol to improve patient prognosis.

Case Presentation

A young adult male, otherwise healthy, presented to the emergency department at University of Florida Health in Jacksonville, Florida, after experiencing a head injury secondary to assault. Patient was reported to have a blow to the head with a metal hanger. Patient was found to have altered mental status, complaining of headache and visual disturbance. On physical examination, the patient had a small scalp laceration anterior to the left preauricular area, and a small left earlobe laceration, and dried blood at both nostrils. There were no signs or symptoms of facial bone fractures. Right eye examination was within the normal limit. However, the left eye was found to have ptosis with complete ophthalmoplegia (cranial nerves III, IV, VI palsy) **(Figure 1)**. Left pupil was dilated (8 mm), round, not reactive to light. Right eye vision was intact, though the patient denied light perception on the left eye, with assessed visual acuity of 20/400. Additionally, a Relative Afferent Pupillary Defect (RAPD) was observed in the left eye. On dilated fundoscopy, vitreous was clear bilaterally, Maculae were flat and optic nerves were sharp, pink with no elevation. Remainder of the head and neck examination was

not remarkable. A head Computed Tomography (CT) revealed comminuted fractures of the anterior sphenoid bone, along with small air and bone fragments in the superior orbital fissure. There was also slight narrowing of the optic canal without involvement in the intraconal region. A small patchy Subarachnoid Hemorrhage (SAH) and pneumocephalus were identified. There was no evidence of swelling in the optic nerve or ocular muscles **(Figure 2)**. No other craniofacial injury was noted. The neck CT was normal, and no aneurysm was detected in the brain CT angiography.

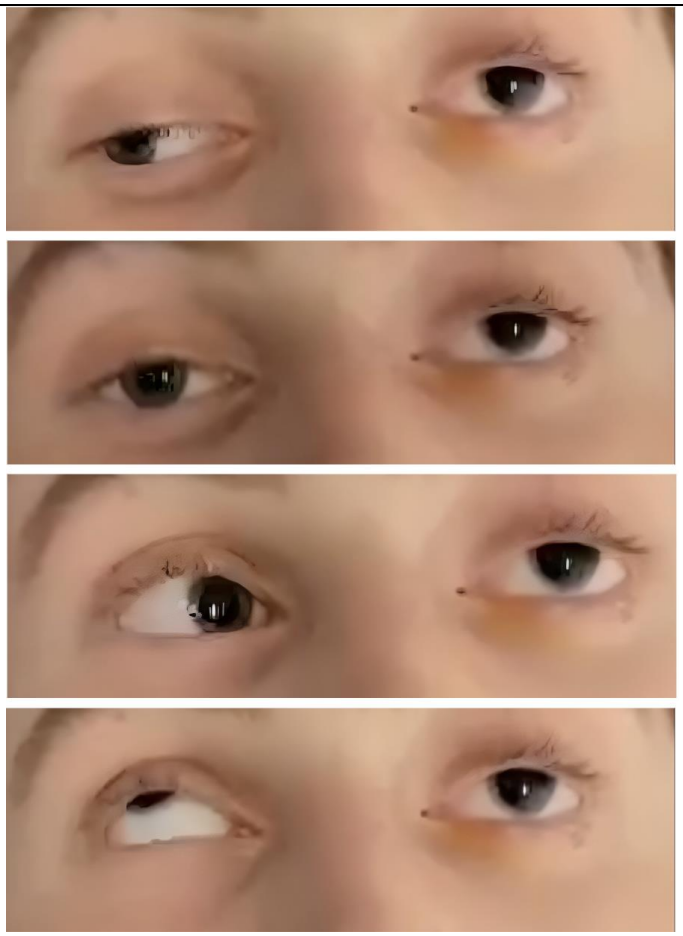

Figure 1: Initial clinical presentation of left eye ophthalmoplegia and ptosis in the patient.

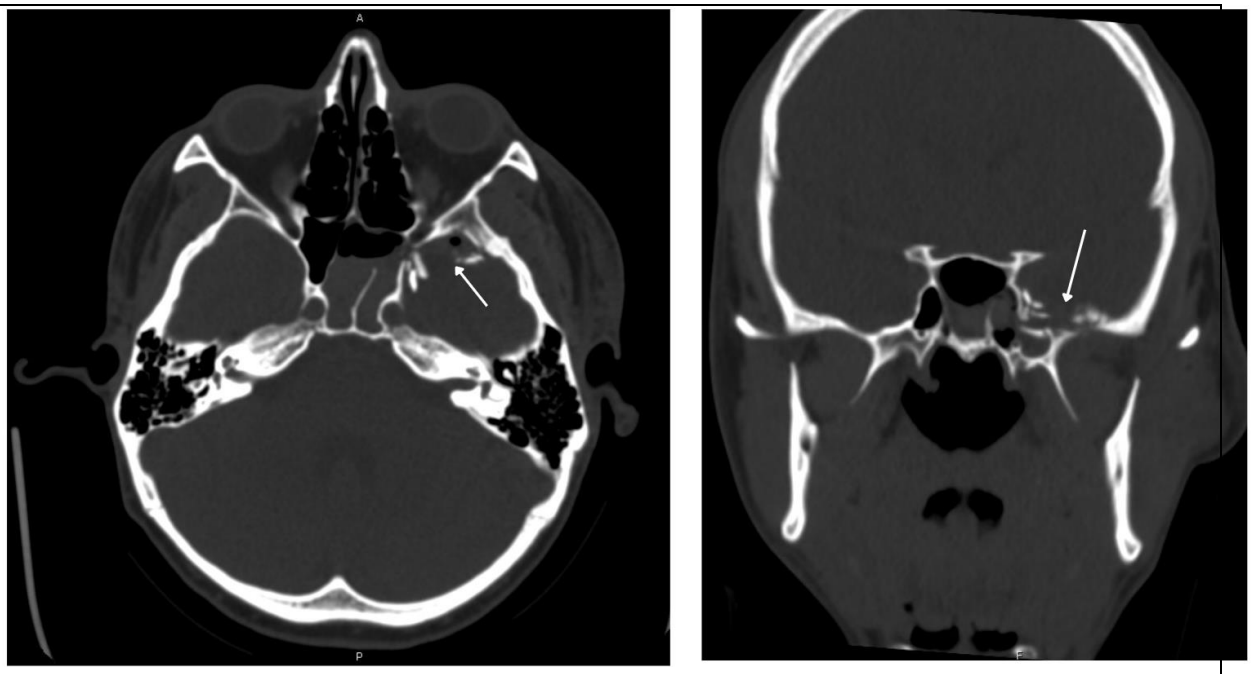

Figure 2: Axial and coronal CT images of the patient's head showing small air and comminuted fractures of the anterior sphenoid bone. The white arrows show bone fragments in the superior orbital fissure.

A diagnosis of traumatic orbital apex syndrome without facial bone fractures was made. No immediate medical or surgical intervention was deemed necessary for the patient, who was closely monitored for 48 hours. Patient regained full mental capacity during the admission. Repeat head CT showed the SAH and pneumocephalus to be stable in size. The second ophthalmologic examination at 48 hours remained unchanged except a mild temporal pallor in the left optic nerve. Patient was eventually discharged. At one month follow-up, the patient displayed a slight improvement in upward gaze, with no changes in the pupillary condition and visual acuity. Although there was no red desaturation observed, the optic nerve exhibited temporal pallor. Optical Coherence Tomography (OCT) showed normal structure of Retinal Nerve Fiber Layer (RNFL). At 6 months, the patient continued to have no light perception out of the left eye, even though extraocular muscle movements showed mild progression, mainly in upward and downward gazes. Patient was also able to blink, with a good corneal sensation. OCT finding was significant for global thinning of RNFL.

Review of Literature

Materials and Methods

A systematic review of English literature published from 1960 to 2023 was conducted by searching the ISI Web of Knowledge database, PubMed, Scopus, Embase, and the Cochrane library using keywords “orbital apex syndrome”, "superior orbital fissure syndrome", “traumatic optic neuropathy”, “TON”, and “ophthalmoplegia”. Our goal was to identify articles presenting patients with traumatic orbital apex syndrome without any associated facial bone fracture and moderate to severe traumatic brain injuries.

The PRISMA Flow Diagram provides a visual representation of our systematic selection process **(Figure 3)**. Initially, a total of 81 records from the databases were identified. Four duplicate records were removed, and an additional set of records was marked as ineligible by automation tools, leaving 32 records included at the

screening stage. These 32 studies were carefully reviewed. Reports of OAS associated with facial fractures (n=29) and those related to severe brain head injuries (n=20) were excluded. Ultimately, two studies met the inclusion criteria and were included in the comprehensive review.

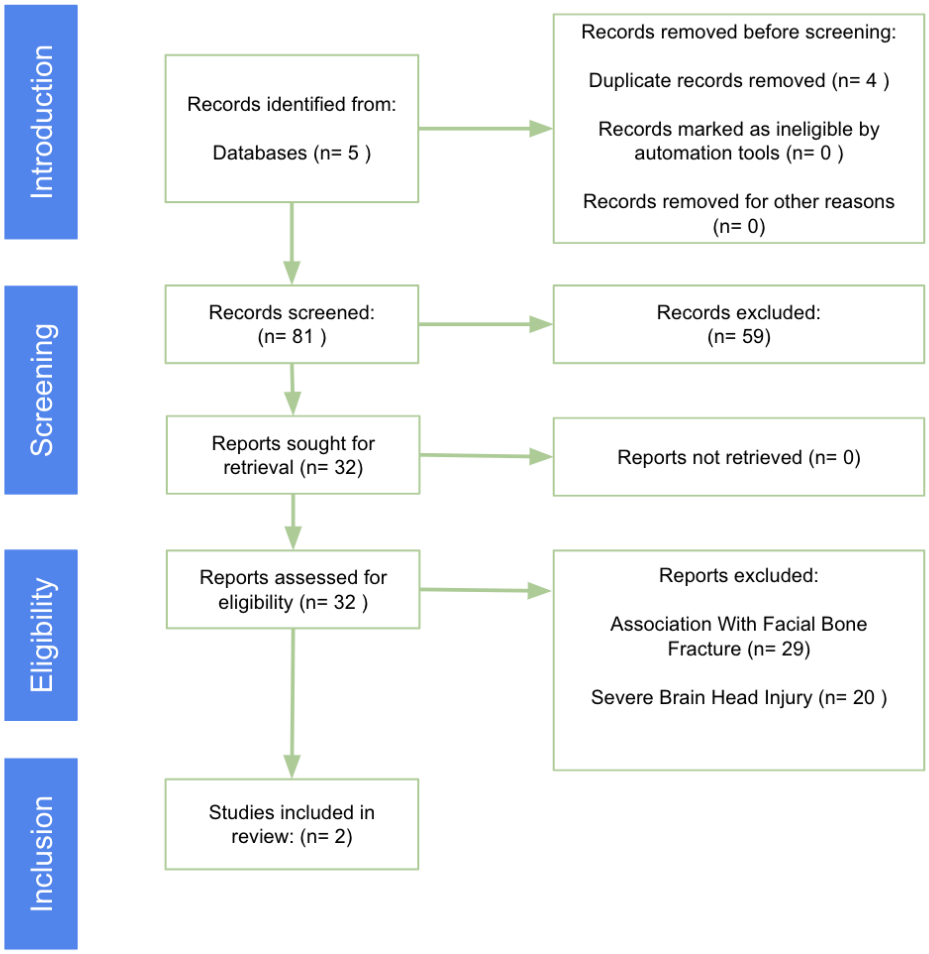

Figure 3: PRISMA Flow Diagram for Study Selection on Isolated Traumatic Orbital Apex Syndrome.

Discussion and Results

The orbital apex is a complex region where neural, vascular, and bony structures converge. This confined space houses the Optic, Oculomotor, Abducens, and Nasociliary nerves. The oculomotor, Abducens, and Nasociliary nerves access the orbit through the superior orbital fissure, though, the optic nerve enters the orbit via the optic canal, alongside the ophthalmic artery [1]. Importantly, the optic nerve is enveloped by meningeal coverings, which firmly adhere to the surrounding periosteum [2]. Understanding the relationship between these structures is essential for diagnosis and management of various orbital and neuro-ophthalmological injuries. The orbit functions as a closed compartment, and rapid intra orbital pressure increase following trauma can potentially cause permanent vision loss due to ischemic damage to the retina and optic nerve. Additionally, associated retro-bulbar hemorrhage, edema, and orbital emphysema are often present in such cases [2-4]. Traumatic Optic Neuropathy (TON) could be categorized into primary and secondary injuries. Primary TON involves mechanical shearing of optic nerve axons and contusion necrosis due to immediate ischemia from damage to the optic nerve microcirculation. In contrast, secondary TON results from apoptosis of both injured and initially

intact adjacent neurons. Notably, many TON patients exhibit a combination of both primary and secondary mechanisms to varying degrees [5]. OAS is an uncommon complication of craniomaxillofacial trauma that endangers vision. Despite the low incidence rate, craniomaxillofacial trauma stands out as the primary contributor to OAS. Given the reported incidence of superior orbital fissure syndrome at only 0.3-0.8% [6] and traumatic optic neuropathy at 0.7–2.5% [5], it is logical to infer that the prevalence of traumatic OAS (where both conditions coexist) would be notably scarcer than either condition in isolation. The literature contains only a limited number of documented cases of this rare syndrome. This syndrome's clinical features result from the involvement of cranial nerves II, III, IV, VI, and the ophthalmic division of cranial nerve V [7]. Clinical signs such as anesthesia, external ophthalmoplegia, ptosis, fixed dilated pupil, lacrimal hyposecretion, diplopia, and decreased visual acuity are highly indicative of OAS [8]. These symptoms result from various nerve and muscle dysfunctions in the orbital apex region. Anesthesia, especially in the cornea and upper eyelid, is due to trigeminal nerve damage. External ophthalmoplegia arises from impaired oculomotor, trochlear, and Abducens nerves, leading to double vision (diplopia). Ptosis occurs due oculomotor or sympathetic nerve dysfunction [9]. Decreased visual acuity is associated with optic neuropathy. Imaging techniques such as high-resolution MRI and CT scans aid in diagnosis [10]. Two other syndromes, superior orbital fissure syndrome and cavernous sinus syndrome, present similar symptoms to OAS [11]. In addition to the cranial nerves affected in OAS, Cavernous Sinus Syndrome (CSS) involves sympathetic nerves and the maxillary division of the trigeminal nerve. Optic nerve neuropathy distinguishes true OAS from Superior Orbital Fissure Syndrome (SOFS) [8]. OAS may result from both direct and indirect injuries. Direct injuries typically involve penetrating trauma or impingement by displaced bony fragments in the orbital apex region. These injuries can cause compression and damage to the cranial nerves, leading to the clinical features of OAS [8,11]. Indirect injuries occur when high-energy impact to the face transmits shear forces to the superior orbital fissure or optic canal. Such forces can disrupt the delicate structures in the orbital apex, including the cranial nerves [12,13]. In their study using holographic interferometry on dried human skulls, Anderson et al. [14] forces applied to certain facial structures like the supraorbital ridge and malar eminence could result in stress concentrations in areas of the orbital roof and floor with subsequent traction, shearing, and shock waves on optic nerve. Heike Huempfner-Hier and colleagues [5] used finite element analysis to simulate the impact forces on the forehead. Their findings showed that low-force impacts created stress that spread towards the optic foramen and chiasm, while higher impacts led to fracture patterns in the anterior skull base, including the optic canal. They suggested that even minor stresses in the optic foramen could cause unnoticed microscopic damage and vision loss, while more significant impacts could result in comminuted fractures, potentially affecting the optic canal's integrity.

In our systematic review, we only found 2 case reports, each reported one patient sustaining OAS without associated facial fracture [15,16]. Both patients, as well as the patient in our case report experienced blunt head trauma around the orbital area. A summary of these 2 patients is shown in **Table 1**.

Table 1: Summary of 2 reported cases of Orbital Apex Syndrome following non-penetrating trauma.

Case #	Loss of consciousness	ptosis	proptosis	Pupil diameter and RAPD	Extra ocular movement	soft tissue trauma	Visual acuity	Ophthalmologic examination findings	Sensory nerve involvement	MRI	CT
1 [15]	One minute	Yes	Yes	Not mentioned	Limited except for lateral gaze	Laceration over the left superior orbital rim	NLP OS	edematous optic nerve, cherry red spot	No	Was not ordered	Normal
2 [16]	No	Yes	No	Large pupil and mild RAPD	complete left ophthalmoplegia, with some relative sparing of abduction only	mild ecchymosis and abrasion of the eyelids	6/12 OS	Normal	No	very mild high signal at the orbital apex	Normal

Clinical and Paraclinical Findings Per Case.

Peter and Pearson in 2010 presented a case of non-penetrating orbital trauma to the left eye secondary to assault with delayed presentation to emergency department. Patient had complete left ptosis associated with a large pupil, blurred vision and diplopia. Fundoscopy findings were normal. The CT of brain and orbits was normal and no evidence of bony or penetrating injury was detectable. Oral steroid, tapering over 6 weeks was started. Visual acuity and sensation returned to normal in 3 weeks, though ptosis and ocular motility continued to improve more gradually over 6 months; Also, the pupil remained dilated with an impaired reaction to light and accommodation **Table 2**.

Table 2: Quality Assessment of Reviewed Case Studies on Isolated Traumatic Orbital Apex Syndrome.

Criteria	Gupta & Khan (2015)	Peter & Pearson (2010)
Clear Objective Stated	Yes	Yes
Detailed Patient Demographics	No	Yes
Clinical Presentation Described	Yes	Yes
Diagnostic Methods Described	No	Yes
Treatment Methods Described	No	Yes
Outcome Measures Stated	No	Yes
Follow-up Period Specified	No	Yes
Potential Conflict of Interest Declared	No	Yes
Limitations Discussed	No	Yes

In 2015, Gupta and Khan, reported a case of OAS from blunt ocular trauma to the left eye without bony involvement after a fall accident. Patient had no light perception, complete ptosis, restricted ocular motions except the lateral gaze. Dilated fundus examination revealed central retinal artery occlusion with edematous optic nerve. CT imaging showed no bony involvement, with proptosis of the globe and moderate inflammation of orbital contents. The authors did not discuss their treatment plan for the patient nor the visual acuity in short-term follow up. The appearance of the optic disc in fundoscopy is influenced by the location and timing of optic nerve damage. Damage in the posterior part of the optic nerve, similar to our patient, often results in a normal-looking optic disc, while damage anterior to the entry site of the central retinal vessels may cause swelling and retinal hemorrhage in the optic disc **[10]**. Regardless of the initial appearance, optic nerve atrophy and pallor typically develop about six weeks after the injury **[17]**. The treatment of optic nerve neuropathy in OAS remains a subject of debate, and there is limited specific literature on its treatment. Currently, three main treatment options exist:

- Observation Alone **[18]**: Some cases of OAS are managed through observation without active intervention.
- High-Dose Corticosteroids **[19]**: In certain instances, high-dose corticosteroids are prescribed as a treatment option. However, it's worth noting that the use of steroids has a theoretical basis in reducing inflammatory reactions.
- Surgical Optic Canal Decompression **[9]**: Surgical intervention involving optic canal decompression is another approach to managing OAS.

A Retrospective study by Zhenxing Li et al. **[20]** on 15 patients with a history of Traumatic Orbital Apex Syndrome confirmed that CN IV experienced the lightest injury in Traumatic Orbital Apex Syndrome (TOAS). They stated that it is related to unique anatomical features. It is the thinnest cranial nerve and is positioned above CN III in the Superior Orbital Fissure (SOF). CN IV's location allows it to endure less traction during trauma compared to CNs III and VI, which pass through the common tendinous ring. Based on their study, CN II (optic nerve) tends to suffer severe injury in TOAS cases. This was attributed to its close attachment to surrounding bones in the optic canal, making it vulnerable to external impact forces. Additionally, CN II is highly sensitive to shock pressure and hypoxia, making regeneration difficult. The study suggests that while CN II may not achieve satisfactory functional outcomes, other cranial nerves involved in TOAS show significant

improvement with appropriate medical or surgical intervention. These improvements can alleviate symptoms of ophthalmoplegia and facial disability. The International Optic Nerve Trauma Study **[14]** compared the outcomes of Traumatic Optic Neuropathy (TON) treated with corticosteroids, optic canal decompression surgery, or observation. Surprisingly, this study didn't find a significant difference in visual outcomes between these treatment groups, and no clear advantage was observed for either steroid therapy or optic canal decompression surgery. It's important to highlight that there are distinctions between direct and indirect TON in terms of prognosis. Direct TON typically results in severe and permanent visual impairment with limited chances of recovery **[21]**. On the other hand, indirect TON cases managed conservatively have a better prognosis, with reported visual function retention over the long term, especially if spared three months after the injury **[10]**. The baseline Visual Acuity (VA) after trauma is a crucial predictor of the final outcome, with initially poor VA associated with limited or no visual recovery. Various factors, such as loss of consciousness after trauma and absence of visual recovery after 48 hours can influence visual recovery and final VA **[10]**.

Conclusion

Traumatic Orbital Apex Syndrome (OAS) without facial bone fractures reveals a complex clinical presentation, with a variable progression depending on the severity and nerves affected. Treatment options such as high-dose corticosteroids, surgical decompression, and observation showed mixed outcomes. A more unified treatment protocol is suggested, with emphasis on early and accurate diagnosis, individualized treatment plans, and a multidisciplinary approach involving various specialists. This tailored strategy aims to enhance patient prognosis by addressing the unique aspects of each case, highlighting the need for ongoing research and collaboration in the management of OAS.

References

1. Lieber S, Fernandez-Miranda JC. Anatomy of the Orbit. J Neurol Surg B Skull Base. 2020;81(4):319-32.
2. Ghannam JY, Al Kharazi KA. Neuroanatomy, Cranial Meninges. In: StatPearls. Treasure Island (FL): StatPearls Publishing, 2023.
3. Turgut B, Karanfil FC, Turgut FA. Orbital Compartment Syndrome. Beyoglu Eye J. 2019;4(1):1-4.
4. Lateral Canthotomy and Cantholysis: Overview, Indications, Contraindications. 2023.
5. Karimi S, Arabi A, Ansari I, Shahraki T, Safi S. A Systematic Literature Review on Traumatic Optic Neuropathy. J Ophthalmol. 2021:2021:5553885.
6. Chen CT, Chen YR. Traumatic Superior Orbital Fissure Syndrome: Current Management. Craniomaxillofac Trauma Reconstr. 2010;3(1):9-16.
7. Apex Orbital Fracture Clinical Presentation: History, Physical, Causes. 2023.
8. Mohankumar A, Gurnani B. Orbital Apex Syndrome. In: StatPearls. Treasure Island (FL): StatPearls Publishing, 2023.
9. Anderson RL, Panje WR, Gross CE. Optic Nerve Blindness Following Blunt Forehead Trauma. Ophthalmology. 1982;89(5):445-55.
10. Goyal P, Lee S, Gupta N, Kumar Y, Mangla M, Hooda K, et al. Orbital apex disorders: Imaging findings and management. Neuroradiol J. 2018;31(2):104-125.

11. Badakere A, Patil-Chhablani P. Orbital Apex Syndrome: A Review. Eye Brain. 2019:11:63-72.
12. Shokri T, Zacharia BE, Lighthall JG. Traumatic Orbital Apex Syndrome: An Uncommon Sequela of Facial Trauma. Ear Nose Throat J. 2019;98(10):609-12.
13. Uberti M, Hasan S, Holmes D, Ganau M, Uff C. Clinical Significance of Isolated Third Cranial Nerve Palsy in Traumatic Brain Injury: A Detailed Description of Four Different Mechanisms of Injury through the Analysis of Our Case Series and Review of the Literature. Emerg Med Int. 2021:2021:5550371.
14. Huempfner-Hierl H, Bohne A, Wollny G, Sterker I, Hierl T. Blunt forehead trauma and optic canal involvement: finite element analysis of anterior skull base and orbit on causes of vision impairment. Br J Ophthalmol. 2015;99(10):1430-4.
15. Gupta R, Khan YA. Traumatic orbital apex syndrome. Can J Ophthalmol. 2015;50(1):e8-e11.
16. Peter NM, Pearson AR. Orbital Apex Syndrome from Blunt Ocular Trauma. Orbit. 2010;29(1):42-4.
17. Yu-Wai-Man P. Traumatic optic neuropathy—Clinical features and management issues. Taiwan J Ophthalmol. 2015;5(1):3-8.
18. Optic Atrophy - EyeWiki. 2023.
19. Levin LA, Beck RW, Joseph MP, Seiff S, Kraker R. The treatment of traumatic optic neuropathy: the International Optic Nerve Trauma Study. Ophthalmology. 1999;106(7):1268-77.
20. Miller NR, Tsai RK. Optic Neuropathies: Current and Future Strategies for Optic Nerve Protection and Repair. Int J Mol Sci. 2023;24(8):6977.
21. Li Z, Zhang D, Chen J, Wang J, Lv L, Hou L. Functional Recovery of Cranial Nerves in Patients with Traumatic Orbital Apex Syndrome. Biomed Res Int. 2017:2017:8640908.

Citation of this Article

Hasheminia A, Malik MA, Rezaei M, Fattahi T, Roozbahany NA. Orbital Apex Syndrome without Facial Fractures: A Case Report and Review of Literature. Mega J Case Rep. 2024;7(5):2001-2010.

Copyright