ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXVII | No. 1 March 2023

Secondary optic neuropathy due to sphenoid-ethmoidal sinus mucocele

Khabibullo Khasanov,
Gulnarakhon Alikhodjayeva,
Jakhongir Yakubov,
Ilkhom Khujanazarov

DOI: 10.33962/roneuro-2023-013

Romanian Neurosurgery (2023) XXXVII (1): pp. 80-84
DOI: 10.33962/roneuro-2023-013
www.journals.lapub.co.uk/index.php/roneurosurgery

Secondary optic neuropathy due to sphenoid-ethmoidal sinus mucocele

Khabibullo Khasanov[1], Gulnarakhon Alikhodjayeva[1], Jakhongir Yakubov[2], Ilkhom Khujanazarov[1]

[1] Department of traumatology, orthopaedics, military-field surgery, and neurosurgery, Tashkent Medical Academy. Tashkent, UZBEKISTAN
[2] Department of Skull base surgery, Republican Specialized Scientific-Practical Medical Center of Neurosurgery. Tashkent, UZBEKISTAN

ABSTRACT

Secondary visual impairment induced by sinusitis is a rare condition that cannot be recognized in all cases. A steady decline in visual acuity and visual field together or alone is the main symptom patients may complain of on admission. This might be hard for general practitioners in Uzbekistan, as possible causes are either intracranial or ophthalmic abnormalities. Hence, it is frequently misdiagnosed or leads to late diagnosis once visual impairment becomes severe. In this paper, we discuss the case of a 9-year-old boy with impaired vision on the left side that was detected almost too late and could have led to complete vision loss. Ineffective conservative therapy was provided for four months. CT and MRI confirmed a lesion in the left sphenoethmoidal sinus. The patient then underwent endoscopic sphenoiethmodotomy with drainage of the left sphenoethmoidal sinus. In the early postoperative phase, as early as the next day after the surgical procedure, the patient experienced visual improvement. Forty days following surgery, in combination with postoperative conservative care in an eye hospital, there was a noticeable improvement in vision. In conclusion, it is crucial for ophthalmologists, neurologists, and ENT surgeons to focus on inflammation in the sphenoethmoidal sinus in children even with mild vision impairment.

Keywords
optic neuropathy
endonasal surgery,
sphenoid sinus mucocele

Corresponding author:
Khabibullo Khasanov

Tashkent Medical Academy.
Tashkent, Uzbekistan

xasanovneuro@gmail.com

ISSN online 2344-4959
© Romanian Society of Neurosurgery

First published
March 2023 by
London Academic Publishing
www.lapub.co.uk

INTRODUCTION

Visual disturbances in optic neuropathies may be produced by numerous etiologic events such as toxic, nutritional, and other ophthalmological as well as intracranial disorders[1,2]. The most prevalent intracranial pathologies that can lead to optic neuropathies are lesions in the sellar and parasellar areas such as pituitary tumors, meningiomas, craniopharyngiomas, and intracerebral lesions affecting the visual pathways, as well as primary or secondary hydrocephalus. In addition to numerous main ophthalmologic illnesses [3], rhinogenic optic neuritis may also be induced by sphenoid sinus mucocele with sinusitis, ethmoiditis, and onodi cell inflammation [4-7].

In some selected circumstances, it is difficult to make a differential diagnosis. When children with good musculoskeletal development

show evidence of gigantism and complain of vision difficulties, they contact multiple physicians such as endocrinologists, neurologists, and even neurosurgeons for probable pituitary tumors. In such particular conditions, MRI and CT can be valuable diagnostic tools to detect lesions in the pituitary region that compress the optic nerve and produce vision abnormalities. MRI is the best choice to evaluate optic nerves except for its intraocular portion and it is fairly difficult to visualize the retina with high-resolution MR imaging. If a lesion is diagnosed in the sphenoidal and ethmoidal sinuses, notably inflammation of the Onodi cells, the physician should think of secondary optic neuritis resulting from this inflammatory process.

CASE REPORT

A 9-year-old kid first visited the outpatient clinic of the Republican Specialized Scientific Practical Medical Center of Neurosurgery in Tashkent, Uzbekistan. His major complaints were acute progressive headache and moderate visual problems when his mother initially consulted his brother, a young neurosurgeon. The patient was tall and had strong musculoskeletal development. He complained of headache and slight visual disruption in the left eye that had continued for a week.

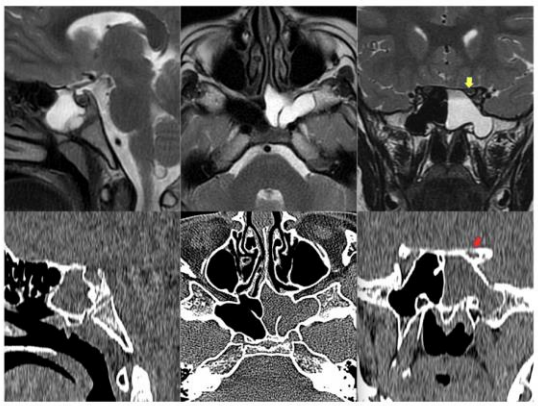

Figure 1. Preoperative MRI and CT show a lesion within the left sphenoethmoidal sinus. On CT, there is no bone erosion around the optic canal. Yellow arrow optic nerve, Red arrow- optic canal.

The patient had no fever or other symptoms; yet, he experienced rhinogenous sinusitis and tonsillitis multiple times in early childhood. In addition, the boy's family history was negative for neurologic or autoimmune disorders. Because of his appearance, which was suggestive of a pituitary lesion, he was advised for an MRI scan. An ophthalmologic examination yielded normal results, but MRI showed a lesion in the left sphenoethmoidal sinus but did not discover any abnormalities in the sellar region (Figure 1). On visual evoked potential (VEP) (table 1), there was extending latency of the P100mc component by 104,2 mc on the left eye and 100,1 mc on the right eye. Ophthalmological examination: visual field: OD-mild stage of myopia and OS-myopic astigmatism, left temporal hemianopsia, visual acuity was as follows: OD-0.9; 0S-0,1. The patient was then advised to consult an ophthalmologist and an otolaryngologist (ear, nose, and throat specialist) in the region where he lived for additional assessment and treatment. After 4 months of conservative outpatient treatment by an ophthalmologist and an otolaryngologist with antibiotics and steroids, the patient's mother called his brother, who was a neurosurgeon, complaining of his son's progressive worsening of visual acuity in his left eye. Ophthalmologic and fundus investigations revealed alterations in the vascular anatomy and optic nerve. CT indicated a left sphenoethmoidal lesion (sinusitis) with no mass effect on the left optic canal; no bone erosion was seen with an inferior-medial wall of the left optic canal (figure 1). Collegial consultation with experts in endoscopic neurosurgery, and several ENT Surgeons and ophthalmologists, as well as literature searches, led to the conclusion that optic neuropathy was diagnosed to be induced by sphenoethmoidal sinusitis.

Table 1. Neuroophthalmological examination

	Visual acuity		Visual field		Visual evoked potential P100mc component	
	Right	Left	Right	Left	Right	Left
Preoperative	O,9	0,1	normal	Hemianopsia	100,1	104,2
Postoperative	1,0	0.9	normal	normal	100,1	101,7

As a result, in case of probable bone involvement and any intraoperative intracranial findings, along with neurosurgeons, endoscopic sphenoethmoidotomy with the excision of the lesion and draining of the left sphenoid sinus and ethmoidal sinus was performed. During surgery, no direct intracranial invasion and no direct

compression of the optic nerve were identified. In the early postoperative phase, as early as the next day after the surgical treatment, the patient reported visual improvement. Then the patient was taken to the Eye hospital where he got postoperative conservative outpatient treatment with antibiotics and steroids. Forty days after surgical therapy in combination with postoperative conservative treatment in an eye hospital, there was a considerable improvement in vision. 40 days follow-up ophthalmologic examination (table 1) showed significant positive changes such as P100mc component latency decreased and was almost similar in both eyes and, On CT (Figure 2) near total removal of mucocele in the left sphenoid sinus and there was a small residual one within the left ethmoidal sinus respectively.

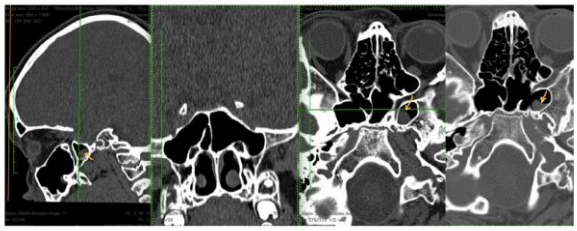

Figure 2. postoperative CT. 40 days post-operative CT shows near total drainage of the left sphenoethmoidal sinus with small remaining mucocele (arrow) within the left ethmoidal sinus.

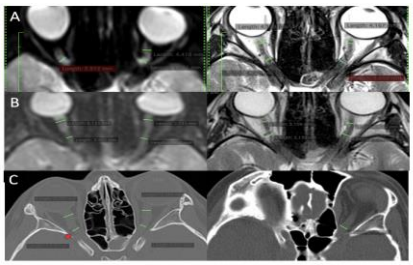

Figure 3
MRI images of a 9-year-old boy: thickness is 3 mm.
A. MRI imaging 4 months before surgery shows the diameter of 3,86 mm retrobulbar left optic nerve and 3,92 mm right optic nerve. On the T2 axial cut MRI image, the diameters of left and right optic nerves on retrobulbar space are 3,6mm and 4,2mm respectively. And on this T2 axial cut image, we can see a lesion that is very close to the left optic nerve and the optic chiazm.
B. Follow UP MRI just 5 days before surgery. On axial cut diffuse MRI shows diameters of left and right optic nerves on the retrobulbar space are 3,4mm and 4,01mm respectively. On the T2 axial cut MR image, the diameter of retrobulbar left and right optic nerves are 4,4mm and 5,1mm respectively.
C. Preoperative CT (left to right): diameters of right and left optic nerves on retrobulbar space are 4,9mm and 5,6mm respectively. Postoperative 40 days follow-up CT shows 5,4 mm diameter of left optic nerve on retrobulbar space.

Discussion

Mucoceles are encapsulated fluid-filled lesions, that can represent in paranasal sinus cavities and there is rarely involvement of sphenoid sinus (up to 3% of all cases) as they are often present in individuals aged over 40 [7]. Since the optic canal and the optic nerve have a closer relationship with the posterior part of the sphenoethmoidal sinus, the Onodi cells which are usually found in that part can cause serious retrobulbar optic neuropathy and in association with cholesterol granuloma, it causes bone destruction and compression [8,10,11]. The most typical symptoms are headache or facial pain, and occasionally, even if this happens seldom, there are the optic and other III, IV, and VI cranial nerves become involved. Cheng et al. described the case in the pediatric population, where they documented a big sphenoid sinus mucocele of a 10-year-old boy who exhibited right eyelid droop and double vision. Even though there was compression and cerebral involvement, following surgery, the patient reported full clearance of mucocele and excellent visual outcome and they argue that the literature reveals cranial neuropathy due to sphenoid sinus mucocele is highly unusual. Prompt diagnosis and care may lead to improved clinical results and the avoidance of permanent neuropathy.

However, postoperative care also plays a significant role as there is usually secondary optic neuritis that needs to be handled carefully. Otsuka et.al. [9] concur with their perspective on the importance of preoperative visual acuity as the prognostic factor for postoperative vision in patients with rhinogenic optic nerve neuropathy, however, poor preoperative vision may be improved by surgical therapy. In addition to this, younger patients are likely to have comparably better results than older people. Steroid pulse treatment occasionally might be utilized for visual acuity improvement following surgery. Even in their research, there was not any significant difference in the prognosis of vision based on the presence or absence of steroid pulse therapy, in our experience, we consider that both preventative preoperative and postoperative use of steroids may enhance surgical results.

Numbers of studies have shown that the origin of visual impairment related to mucoceles involves direct mechanical compression, local ischemia, and inflammation. They indicate distinct prognoses. While the slow manifestation of clinical symptoms and a better prognosis typically occur in mechanic compression, the latter two, ischemia and inflammation generate an early start of symptoms that may have a bad prognosis [12-15]. Carefully preoperative screening may indicate various causes of optic neuropathy. However, it is difficult to identify a normal optic nerve and an optic nerve lesion. The disc component may be analyzed by utilizing fundoscopy and high-resolution optical coherence tomography, whereas the other three parts are less accessible and require imaging procedures like MR imaging, sonography, and CT. MR imaging is preferred to the other tests described above thanks to several features such as exceptional soft tissue resolution, the absence of ionizing radiation, and high diagnostic accuracy. MR imaging has a

sensitivity of 60-75% and a specificity of up to 90% for identifying abnormalities in the optic nerve [16,17]. In typical circumstances, the intraorbital portion of the optic nerve has a mean diameter ranging from 2.2 to 5.2 mm. The diameter of the optic nerves normally grows throughout the first two years of life. However, for children aged 6 to 12, whereas the diameter of the optic nerves on axial cut MR images on the retrobulbar level is 2.26 0.38 mm, it is 2.27 0.41 mm on coronal plane MRI on the retrobulbar space. As a consequence, on both axial and coronal incisions, the midaspect part of the optic nerve should be thinner than the retrobulbar region [18].

In our case, we assume that the visual impairment that the patient experienced was secondary owing to inflammation of the sphenoethmoidal sinus, specifically the posterior region which normally has a tighter relationship with intracanalicular optic nerves. Even though there is no direct compression on the optic nerves, this inflammation and toxins are thought to irritate the optic nerve creating a circulatory disturbance and nerve trophic dysfunction. As a consequence, this might lead to a deterioration in vision. Our findings on radiological exams such as MRI (figure 3A and 3B) and CT (Figure 3C), measuring diameters of the optic nerves in retrobulbar space might identify probable alterations in circulation and optic nerves` an or/and hypotrophy that commonly leads in visual difficulties.

In early observation days, when the patient first came to admission because of headache and mild impaired vision, an early MRI showed a mass lesion within the left sphenoid and ethmoidal sinus. We measured the diameters of both optic nerve and it was almost similar in distal part of the retrobulbar optic nerve in both eyes (Figure 3A), however, in its proximal part, close to mass lesion there was a gap between the diameters of the retrobulbar left and right optic nerves on a T2 axial cut MR scan(3.6 mm and 4.2 mm, respectively). The diameters of both optic nerves should be almost the same in principle, but in the present patient, there was a gap between the diameters of the right and left optic nerves as seen on T2 MRI images, and the difference was around 1.2mm, suggesting hypotrophy of the left optic nerve. The diameters of the right and left optic nerves, as well as their difference, remained stable in early preoperative computed tomography (figure 3C right); however, due to the resolution of CT compared to MR imaging, those numbers were slightly different; 4,9mm and 3,6mm in retrobulbar space, respectively. The gap between the right and left optic nerves was about 1.3mm. Even after 4 months of conservative therapy by otolaryngologists and ophthalmologists, the patient's visual acuity had deteriorated, and they have referred to us again. Because of these alterations and decreasing visual acuity, conservative therapy was proven ineffective, and the patient needed surgical intervention. The patient was checked again 40 days following endonasal surgery and postoperative conservative therapy with antibiotics and steroids. His vision dramatically improved, and he recovered rapidly. There was also no headache. The diameter of the left optic nerve was 5,4 mm on CT, which was approximately identical to the diameters of the right optic nerve we measured before the surgery. As a consequence, the diameter of the left optic nerve expanded from 3,6mm to 5,4mm (figure 3C left). However, owing to a lack of precision and resolution quality, CT may not be a viable technique for assessing the diameter of the optic nerve, which we feel is a drawback of our research.

Conclusion

Secondary optic neuropathy, due to sphenoid sinus mucocele is common in the pediatric population. Pricey exams such as MRI and CT are required for a correct diagnosis and treatment selection. Minimally invasive surgical therapy is the major therapeutic objective to conquer even such basic and infrequent instances. If inflammatory illnesses become chronic in youngsters. Rhinogen sinusitis may occasionally produce visual difficulties since sinusitis is prevalent in both children and adults.

References

1. Margolin E, Blair K, Shemesh A. Toxic and Nutritional Optic Neuropathy. 2022 May 20. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–. PMID: 29763154.
2. Behbehani R. Clinical approach to optic neuropathies. Clin Ophthalmol. 2007 Sep;1(3):233-46. PMID: 19668477; PMCID: PMC2701125.
3. Rodriguez-Beato FY, De Jesus O. Compressive Optic Neuropathy. [Updated 2022 Jul 12]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK560583/.
4. Deb AK, Neena A, Sarkar S, Devraj A, Panicker G, Stephen

AM. Bilateral compressive optic neuropathy secondary to sphenoid sinus mucocele mimicking bilateral retrobulbar neuritis. Saudi J Ophthalmol. 2022 Jun 13;35(4):368-370. doi: 10.4103/1319-4534.347312. PMID: 35814987; PMCID: PMC9266473.

5. Lee SM, Lee KI. Recurred Sphenoethmoidal Mucocele After Treated With Simple Endoscopic Marsupialization. J Craniofac Surg. 2021 Oct 1;32(7):e661-e663. doi: 10.1097/SCS.0000000000007825. PMID: 34191777.
6. Ting MYL, Shan M, Gantz O, Zhang-Nunes S, Wrobel B. Optic Neuropathy due to an Ethmoid Mucocele: A Case Report and Literature Review. Case Rep Ophthalmol. 2019 Aug 6;10(2):227-234. doi: 10.1159/000501898. PMID: 31692625; PMCID: PMC6760354.
7. Chang H, Silva M, Weng J, Reilly E, Levine C, McCrea HJ. Large sphenoid mucocele presenting with cranial neuropathies in a 10-year-old boy: case report and literature review. Childs Nerv Syst. 2022 May;38(5):1035-1039. doi: 10.1007/s00381-021-05314-5. Epub 2021 Aug 4. PMID: 34347143.
8. Tzamalis A, Diafas A, Riga P, Konstantinidis I, Ziakas N. Onodi Cell Mucocele-Associated Optic Neuropathy: A Rare Case Report and Review of the Literature. J Curr Ophthalmol. 2020 Mar 23;32(1):107-113. doi: 10.1016/j.joco.2019.08.006. PMID: 32510023; PMCID: PMC7265265.
9. Otsuka M, Yunoki T, Ozaki H, Tachino H, Takakura H, Shojaku H, Hayashi A. Prognostic Factors for Postoperative Visual Acuity in Patients with Rhinogenic Optic Neuropathy. J Ophthalmol. 2019 Oct 3;2019:3417425. doi: 10.1155/2019/3417425. PMID: 31687196; PMCID: PMC6794975.
10. Geng C, Tong Q, Wang Y, Huang S, Wang M, Xing Z, Sun K. Onodi cell mucocele with cholesterol granuloma is more likely to cause serious optic neuropathy, compared with simple Onodi cell mucocele. Ear Nose Throat J. 2022 Feb 25:1455613221080069. doi: 10.1177/0145561322 1080069. Epub ahead of print. PMID: 35213237.
11. Morino T, Yanagihara T, Kuroyanagi H, Ohto H. Optic neuropathy caused by expanded Onodi cell mucocele. BMJ Case Rep. 2020 Jun 2;13(6):e235311. doi: 10.1136/bcr-2020-235311. PMID: 32487523; PMCID: PMC7269546.
12. Nickerson JP, Lane AP, Subramanian PS, Izbudak I. Onodi cell mucocele causing acute vision loss: radiological and surgical correlation. Clin Neuroradiol. 2011;21:245-248.
13. Tseng CC, Ho CY, Kao SC. Ophthalmic manifestations of paranasal sinus mucoceles. J Chin Med Assoc. 2005;68: 260-264.
14. Fukuda Y, Chikamatsu K, Ninomiya H, Yasuoka Y, Miyashita M, Furuya N. Mucocele in an Onodi cell with simultaneous bilateral visual disturbance. Auris Nasus Larynx. 2006;33: 199-202.
15. Kim YS, Kim K, Lee JG, Yoon JH, Kim CH. Paranasal sinus mucoceles with ophthalmologic manifestations: A 17-year review of 96 cases. Am J Rhinol Allergy. 2011;25: 272-275.
16. Chawla B, Sharma S, Sen S, et al. Correlation between clinical features, magnetic resonance imaging, and histopathologic findings in retinoblastoma: a prospective study. Ophthalmology 2012; 119:850 –56 CrossRef Medline
17. Schueler A, Hosten N, Bechrakis N, et al. High resolution magnetic resonance imaging of retinoblastoma. Br J Ophthalmol 2003;87: 330 –35 CrossRef Medline
18. Al-Haddad CE, Sebaaly MG, Tutunji RN, Mehanna CJ, Saaybi SR, Khamis AM, Hourani RG. Optic Nerve Measurement on MRI in the Pediatric Population: Normative Values and Correlations. AJNR Am J Neuroradiol. 2018 Feb;39(2):369-374. doi: 10.3174/ajnr.A5456. Epub 2017 Dec 7. PMID: 29217740; PMCID: PMC7410600.