Optic Neuropathy, Secondary to Ethmoiditis, and Onodi Cell Inflammation during Childhood: A Case Report and Review of the Literature

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Abstract

Optic neuropathy consists of several etiological events. The primary etiologies of its acute form include optic neuritis, ischemic optic neuropathy, inflammatory (non-demyelinating) disorders, and trauma. Its subacute and chronic forms are most often linked to compressive, toxic, nutritional, or hereditary-genetic causes. Visual loss, dyschromatopsia, and visual field defects are the presenting symptoms. The Onodi cell (sphenoethmoidal air cell) is an anatomic variant located laterally and superior to the sphenoid sinus; it is closely related to the optic nerve. Onodi cell disorders are rare and may be unnoticed in differential diagnoses of patients with ocular and neurological manifestations. Here, we present the case of a 12-year-old boy with headache and acute loss of sight characterized by hemianopsia in the left eye and retrobulbar optic neuropathy caused by left sphenethmoidal sinusitis with the presence of Onodi cell inflammation. The diagnosis was confirmed by multilayered paranasal computed tomography and cerebral magnetic resonance imaging. Therapeutic treatment resulted in gradual improvement: at the 2-week follow-up, the patient no longer had headaches and his visual acuity returned to normal. Inflammation of Onodi cells should be considered in children with headache and abnormal vision.

Keywords
► optic neuropathy
► ethmoiditis
► Onodi cell
► optic neuritis

Introduction

Optic neuropathy refers to damage of the optic nerve, which can be linked to demyelinating, inflammatory, ischemic, traumatic, compressive, toxic/nutritional, or hereditary events.1–3 Optic neuropathies are rare during childhood, but compressive/tumoral and inflammatory events are the most frequent etiologies of childhood optic neuropathies.1

Optic nerve impairment is the cause of more or less gradual visual loss, discoloration, visual field defects and other ophthalmologic and neurologic involvements. The optic nerve is topographically correlated with different brain structures, including the Onodi cell (spheno-ethmoidal air cell).2,3 The Onodi cell was first described by Adolf Onodi in 1904: it is an anatomical variant of the posterior ethmoid cells surrounding the optic nerve and ventilated by the sphenoid sinuses. When the migration of the posterior ethmoid cells is complete, the Onodi cell is located around the optic nerve. Therefore, there is a close anatomical relationship between the Onodi cell and the optic nerve, which can lead to ophthalmologic complications. In the literature, the incidence of Onodi cells has been reported to range from 7 to 14% in radiological findings.2,3 Anatomical studies of Onodi cells have been reported in up to 60% of samples.3

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Patients with sphenoethmoidal mucocele and visual disturbances are commonly seen by ophthalmologists. Very often, the Onodi cell is an incidental finding unrelated to a specific disorder. However, in some cases, it may be linked to infectious or inflammatory sinusitis, fungus ball, inverted papilloma, or a malignant mucocele or sinonasal tumor. Lesions involving the Onodi cell can cause visual abnormalities based on its anatomical correlation with the optic nerve. Here, we present a case report of a 12-year-old boy with optic nerve compression secondary to ethmoiditis and Onodi cell inflammation whose optic neuropathy may be due to ischemic insult to the compromised blood supply to the optic nerve or the mass effect of direct compressive impact or optic neuritis. Medical treatment led to a progressive and rapid resolution of the patient’s symptoms with full recovery of his visual function.

Case Report

A 12-year-old boy was recovered at the Pediatrics Department of Catania University (Catania, Italy) for the acute, progressive appearance of headaches and visual disturbances with acute visual loss characterized by hemianopsia in his left eye that had persisted for 4 days. The patient did not have fever and no treatment was initiated. The boy’s family history was negative for neurological or autoimmune diseases. His medical history referred to frequent specialist visits for the onset of headaches in recent months. For this reason, cerebral magnetic resonance imaging (MRI) and an ophthalmological examination were made; both yielded normal results. A physical examination conducted upon admission did not yield ocular or nasal symptoms or neurological deficits or abnormalities of the heart, chest, or abdominal organs. An ophthalmological examination and visual field confirmed left temporal hemianopsia and hypovoltated homolateral bonds (Fig. 1). The retina, macula, and intraocular pressures in both eyes were within normal ranges. A preliminary diagnosis of optic neuropathy was made, and additional investigations were conducted to determine the cause of the ocular abnormalities. On the third day of healing, the child presented a closed-nose sensation. The patient’s biochemical, hematological, immunological (white blood cell), and viral markers were within the normal range. An analysis of cerebrospinal fluid (was negative for infectious agents and for specific neuronal autoimmunity. An enhanced cerebral MRI (Fig. 2) revealed increased contrast absorption around the right optic nerve, which was considered to be venous congestion secondary to compression or a retro-orbital inflammatory condition. Paranasal computed tomography (CT) images confirmed these hypotheses and also revealed the presence of the sphenoethmoidal Onodi cell with inflammation of sphenoethmoidal cells on the left eye and diffuse thickening of the bilateral maxillary sinuses (Fig. 3). Based on a hypothetical diagnosis of optic neuropathy secondary to inflammation of the paranasal sinuses, the patient was referred to the otorhinolaryngology team. These researchers performed a nasal endoscopy that revealed the presence of mucopurulent rhinorrhea in the left nasal fossa from the middle meatus and the sphenoethmoid recess. Based on the clinical and radiological results and the presence of inflammation of the Onodi cells, a diagnosis of acute sinusitis of the ethmoid and left maxillary sphenoid was made. The patient was treated with intravenous antibiotics and corticosteroid (ceftriaxone and methylprednisolone) therapy. After 10 days of medical treatment, progressive improvement in the patient’s symptoms was noted and confirmed by a visual field test. At the 2-week follow-up, a cerebral MRI (Fig. 4) revealed marked reduction of the parasanal inflammation. At the 3-month follow-up, the patient’s visual acuity had completely resolved.

Discussion

The patient exhibited diminished visual acuity because of optic neuropathy due to left maxillary ethmoid bone and sphenoid sinusitis and inflammation of the Onodi cells. As Onodi cells are anatomically related to the optic nerve, Onodi cell disorders may result in ophthalmological complications such as rhinogenic optic neuropathies and poor vision. Optic neuropathy is caused by mechanical compression of the optic nerve, circulatory disturbances of the nervous vessel because of mechanical compression, and optic neuritis.

Fig. 1 (A) The left image describes a left hemianopsia on the visual field test. (B) The middle visual field shows left hemianopsia reduction after 1 week of antibiotic and steroid therapies. (C) The right visual field test shows a left residual scotoma after 1 month of systemic antibiotic therapy.
due to inflammation. Because of the presence of a macular star, we suspected that an infection of the retrobulbar region had spread across the optic nerve to the right papilla and caused the neuroretinitis. Optic neuropathy is linked to many causes—the common includes compressive optic neuropathy (including orbital and intracranial meningiomas), pituitary adenomas, and intracranial aneurysms. Optic neuropathy is rarely caused by Onodi air cells. The lateral or superior pneumatization of this posterior ethmoidal air cell in the sphenoid sinus may directly affect the optic nerve. This is clinically significant because any disturbance of Onodi air cells may affect this cranial nerve given their close anatomical proximity. In our case, there is no doubt about the role of inflammation of the paranasal sinuses in the optic neuropathy. However, the etiopathogenesis appears to be related to a hematogenic inflammation of the left optic nerve with a concomitant sinusitis without any biological signs of infectious disease. In the proband, the diagnosis was made on CT of the paranasal sinuses and on a cerebral MRI. CT, particularly on the coronal plane, is an excellent diagnostic tool for accurately determining the relationships linking the optic nerve and the paranasal sinuses, the osteomeatal complex, the course of the optic nerve, and the optic foramen. Axial imaging is particularly used to plan functional endoscopic surgery of the sinus in disorders of the posterior ethmoidal and sphenoidal sinuses. Onodi air cell management remains debatable. However, surgical decompression remains the gold-standard treatment, and adjuvant systemic antibiotic therapy and corticosteroids are widely used as observed in our case. Poor visual prognostic factors

Fig. 2  Magnetic resonance imaging scan showed a phlogistic thickening of the left ethmoidal sinus and particularly around the Onodi cell.

Fig. 3  Computed tomography scan confirms a phlogistic thickening of the left ethmoidal sinus and particularly around the Onodi cell.
include severe deficits in visual acuity and the sudden onset of symptoms.\textsuperscript{9} The interval between the onset of symptoms and surgery is also crucial for visual recovery. To the best of our knowledge, the patient described here represents the first report of such a disorder during childhood. As such, this case demonstrates that Onodi cells and ethmoiditis should be included in the etiology of compressive optic neuropathy in pediatric patients. We note that optic neuropathy is rare during childhood, unlike its prevalence during adulthood. Fukuda et al\textsuperscript{10} described a 45-year-old woman who presented a rare case of small, isolated Onodi cell mucocele manifesting as unilateral chronic optic neuropathy. The patient complained of gradual visual disturbance in her left eye without other neurological abnormalities. Neuroimaging examinations revealed a small cystic lesion located in the left posterior ethmoid extending into the anterior clinoid process. Surgery using a pectoral epidural approach revealed a cystic lesion containing isolated mucosal fluid in the anterior clinoid process. The total removal of the lesion associated with a reopening procedure of the left optic canal led to relief of the patient’s symptoms. A histological examination showed mucocele lesions. Based on the radiological and surgical results, together with the histological examination, the final diagnosis was of mucocele of the Onodi cell. Small Onodi cell mucocele may also cause optic neuropathy, but this condition is extremely unusual.\textsuperscript{11} Wu et al\textsuperscript{11} reported a case of a 28-year-old man who was admitted with a fair visual acuity of 20/100 after treatment for an initial diagnosis of optic neuritis. A subsequent examination suggested compressive optic neuropathy, and neuroimaging confirmed the presence of mucocele of Onodi cells that compressed the optic nerve. The patient underwent right endonasal sphenethmoidectomy with decompression 5 weeks after the initial onset of his symptoms. Three weeks after surgery, the patient’s visual acuity was 20/20, and the resolution of the visual field defect was complete. The patient’s condition was stable 1 year after the procedure. Taflan et al\textsuperscript{12} reported a study of a 61-year-old woman with optic neuropathy due to lesions in the Onodi cell. This patient presented, within a few days, progressive loss of vision in her right eye. CT revealed mucosal thickening and inflammatory signs in the right sphenoid sinus and the presence of Onodi cell on that side. MRI confirmed optic nerve compression. Systemic antibiotic therapy and endoscopic sinus surgery were done. After surgery, the patient’s visual acuity and CT scans were normal. A histopathological investigation revealed polyps in the Onodi cell. Wada et al\textsuperscript{13} examined the efficacy of the identification of the Onodi cell and the classification of the sphenoid sinus using sagittal CT for a sphenoidotomy. These authors studied CT images of the patients’ sinuses (n = 261; 522 sides). Wada et al analyzed the relationships between the lateral side of the anterior wall of the sphenoid sinus and the optic nerve and between the middle of the anterior wall of the sphenoid sinus and the skull or the pituitary gland. The images were classified according to the base of the skull (without the Onodi cell), the optic canal, the sella, or infrasella (all with the Onodi cell). The classification of the anterior wall of the sphenoid sinus based on the Onodi cell allowed for a three-dimensional assessment of the shape of the sphenoid sinus: (1) it opened safely via a complete preoperative assessment of the anterior wall; (2) the position of the superior turbinate; and (3) the position of the sphenoid sinus ostium. Park and Oh\textsuperscript{14} described a 27-year-old man who underwent rapid and severe visual loss in both eyes in association with pain behind his eyes. Based on a presumptive diagnosis of retrobulbar optic neuritis, the patient was treated with intravenous corticosteroids, and his vision improved transitorily. His vision then worsened without light perception, and MRI revealed a bilateral improvement of the optic nerve with increased dural and thickening in the base of the anterior skull, sella, and retroclival areas. These characteristics were
initially interpreted as inflammatory. Nasopharyngoscopy disclosed a soft tissue lesion filling the apex of the nasopharynx and the posterior portion of the ethmoid sinus with associated sinusitis. A biopsy revealed a moderately differentiated squamous cell carcinoma that was believed to have originated in the nasopharynx. Severe bilateral optic neuropathy in nasopharyngeal carcinoma that invaded the base of the skull confirmed that rapidly progressive severe bilateral optic neuropathy in a young patient with periorbital pain is not always recognized as having an inflammatory origin. Furthermore, genetic abnormalities can be responsible for optic atrophy during childhood. Mutations in OPA1 are responsible of 32 to 89% cases of autosomal dominant optic atrophy. This disease typically occurs during childhood with progressive bilateral visual loss because of the neurodegeneration of retinal ganglion cells; multifactorial events may influence both its onset and phenotype. 

Pretegiani et al reported a neuro-ophthalmologic assessment of an Italian group of patients on 60 OPA1 mutations carriers (52 symptomatic) belonging to 13 families. The authors discovered four known mutations, the most common being a lack of conscience c1034G > A, and a new missense mutation, c1193A > C. This new missense mutation was found in a 54-year-old woman with the late-onset phenotype. The authors concluded that the neuropathy was caused by compression (mucoceles and a case of infiltrative carcinoma) and required surgery, regardless of the patient’s response to medical treatment.

In conclusion, ophthalmologists, otorhinolaryngologists, pediatricians, child neurologists, and neuroradiologists should be aware of the presence of the Onodi cell and all sinusopathies that can give rise to optic neuropathy. Delays in diagnosis and treatment increase the risk of vision loss, and it is essential to address these cases in a multidisciplinary way while maintaining close contact among medical staff.

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