Case Report: Optic Disc Edema without Hydrocephalus in Acoustic Neuroma

Joseph Grainger, M.R.C.S.¹ and Palitha S. Dias, F.R.C.S.,¹

ABSTRACT

Traditionally, visual disturbance and optic disc edema are regarded as late manifestations of acoustic neuromas indicating increased intracranial pressure as a result of obstructive hydrocephalus or a sizeable mass lesion. We report the case of a 56-year-old man who presented with visual disturbance and bilateral optic disc edema. Classic features of hydrocephalus were absent. Magnetic resonance imaging showed a large acoustic neuroma. However, there was no ventriculomegaly and at surgery intracranial pressure was normal. We suggest that cerebrospinal fluid protein may have a role in the formation of optic disc edema through a normal pressure communicating type of hydrocephalus. Furthermore, patients with acoustic neuromas and a visual disturbance related to optic disc edema may be inappropriate for treatment with stereotactic radiosurgery and should be offered early surgery to prevent visual deterioration

KEYWORDS: Acoustic neuroma, papilledema, hydrocephalus

Acoustic neuromas, also known as vestibular schwannomas, account for about 8% of intracranial tumors and are the most common lesion of the cerebellopontine angle.¹

The presentation of an acoustic neuroma is related to the size of the lesion and the presence of an intrameatal component. However, hearing loss and tinnitus are by far the most common presenting symptoms, occurring in more than 95% and 63% of patients, respectively.^{1,2} Ophthalmic symptoms are less common, occurring in as many as 16% of patients, and include diplopia, blurring of vision,

obscurations, and tunnel vision.¹ Ophthalmic findings may be related to the effect of the expanding mass on adjacent cranial nerves or to increased intracranial pressure (ICP). They tend to occur with larger lesions. At presentation, optic disc edema may be present in as many as 8% of patients with acoustic neuromas.¹ The edema is usually related to intracranial mass effect or compression of the fourth ventricle resulting in obstructive hydrocephalus. It is well recognized that untreated optic disc edema may cause deterioration of visual function.^{3,4} Therefore, the presence of

Skull Base, volume 15, number 1, 2005. Address for correspondence and reprint requests: Palitha S. Dias, F.R.C.S., Department of Neurosurgery, University Hospital of North Staffordshire, Royal Infirmary, Princes Road, Hartshill, Stoke-on-Trent, ST4 7LN, Great Britain, United Kingdom. E-mail: Palitha.Dias@uhns.nhs.uk. ¹Department of Neurosurgery, University Hospital of North Staffordshire, Royal Infirmary, Stoke-on-Trent, Great Britain, United Kingdom. Copyright © 2005 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2005,15,01,083,088,ftx,en;sbs00433x.

optic disc edema has significant implications for management.

Over the past two decades, the management of acoustic neuromas has changed significantly with the introduction of stereotactic radiosurgical techniques. These procedures may be used in patients with normal ICP and offer high rates of tumor growth control often with regression.⁵ Open surgical techniques are favored in patients with increased ICP to provide rapid decompression.

We present the case of a patient who presented with visual disturbance and optic disc edema with no evidence of obstructive hydrocephalus or increased ICP.

CASE REPORT

A 56-year-old man had a 6-month history of blurred vision. The blurred vision, which was worse in the left eye but affected both eyes, was increasing. Headache had been present initially but had gradually resolved. On further questioning he admitted that 4 months earlier he had become deaf in his left ear and since then had had tinnitus. When he noticed the tinnitus, he had experienced several episodes of dizziness and described symptoms of vertigo. However, these symptoms also had improved by the time he was examined. He had no nausea, vomiting, or incontinence and complained of no memory problems.

On examination he was alert and oriented. His visual acuity was 6/9 bilaterally. Fundoscopy revealed gross bilateral optic disc edema (Fig. 1). His visual field testing demonstrated enlarged blind spots. Sensation in the distribution of the left ophthalmic and maxillary divisions of the trigeminal nerve was reduced to light touch. The left corneal reflex was absent. The facial nerve was intact. Sensorineural deafness in the left ear was confirmed by pure tone audiometry. Hearing in the right ear was normal. His gag reflex was intact and he had no cerebellar signs. His gait was normal.

Figure 1 Fundoscopy showing optic disc edema.

A supratentorial mass lesion was suspected. Computerized tomography revealed no mass lesion. The size of the ventricles was normal, but the fourth ventricle was shifted to the right (Fig. 2). Magnetic resonance imaging showed a large mass lesion at the left cerebellopontine angle entering the internal auditory meatus, consistent with a large acoustic neuroma. The mass had displaced the contents of the posterior fossa to the right, but again the size of the ventricle was normal (Fig. 3).

Management options were discussed with the patient. To prevent further visual deterioration, surgical excision was advised. He underwent posterior fossa craniectomy and excision of the mass lesion. Before the lesion was removed, an external ventricular drain and an ICP monitor were placed. Opening ICP was 14 cm H₂O. Ventricular cerebrospinal fluid (CSF) protein concentration was

Figure 2 Axial computerized tomographic scans showing normal ventricular size and displacement of the fourth ventricle.

Figure 3 Sagittal T2-weighted magnetic resonance imaging showing a large lesion involving the left side of the cerebellopontine angle and normal ventricular size.

0.45 g/dL. CSF protein concentration from the basal cisterns was significantly raised at 4.05 g/dL.

Postoperatively, the patient had a facial nerve palsy. Follow-up at 3 months revealed no optic disc edema and the facial nerve was recovering. The patient was independent in his activities of daily living.

DISCUSSION

Visual disturbance and optic disc edema are unusual presenting features of acoustic neuromas. They usually indicate increased ICP as a result of obstructive hydrocephalus or a sizeable mass lesion. In this case, however, optic disc edema was present even though these other findings were absent. The cause of optic disc edema in this case remains unclear. ICP recorded during surgery was normal. However, this finding does not exclude intermittently increased ICP as is thought to occur in normal pressure hydrocephalus.⁶ Normal pressure hydrocephalus typically causes gait ataxia, memory loss, and incontinence, but these findings were absent in this case. Furthermore, general anesthesia may have reduced the patient's ICP before it was measured.

Sporadic cases of optic disc edema in the absence of increased ICP have been reported in the world literature in patients with leukemia⁷ and iron-deficiency anemia.⁸ Local mechanisms have been proposed for these cases. In this case, a high protein concentration was found in the CSF around the neuroma. We suggest that this high protein concentration may have impaired reabsorption of CSF, albeit intermittently, resulting in a type of communicating hydrocephalus. This mechanism has been suggested to underlie increased ICP and normal pressure hydrocephalus associated with spinal cord schwannomas.⁹ Local protein concentrations were found to be high and symptoms resolved after excision of the lesion. Harada and colleagues¹⁰ reported optic disc edema without hydrocephalus in a patient with neurofibromatosis II and bilateral acoustic neuromas. They also suggested high protein concentration as a possible mechanism. In their case, however, CSF protein concentration was not measured.

In this case, early surgical decompression was favored to prevent optic atrophy from prolonged optic disc edema. We would advocate this approach in preference to stereotactic radiosurgery in patients with optic disc edema, even in the absence of evidence of increased ICP.

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Commentary

This interesting case suggests that papilledema may result from intracranial pathology provoking high cerebrospinal fluid (CSF) protein without the hallmark of increased intracranial pressure (ICP). The authors speculate that the increased CSF protein impaired reabsorption of CSF in their patient, causing communicating hydrocephalus that produced papilledema and visual deterioration.

Traditionally, disc edema with normal ICP has been associated with local ophthalmologic processes such as central vein obstruction, hypertension, optic neuritis, ischemic neuropathy, compressive neuropathy, scleritis, uveitis, diabetes, toxicity, or infection. Bilateral disc edema with increased ICP is characteristic of pathology beyond the optic nerve itself. The level of protein in this patient's CSF was quite high, a condition known to be caused by a variety of intracranial and intraspinal tumors. An increase in the level of protein in the CSF has been postulated to increase ICP by decreasing absorption of CSF by blocking all semi-permeable membranes of the arachnoid granulations. This mechanism was described by Gardner and colleagues¹ in 1954. Given this hypothesis, the absence of hydrocephalus in this patient indicates rather mild cerebral edema from the elevated CSF protein levels. Furthermore, all previous descriptions of the complications related to elevated CSF protein have included papilledema associated with

increased ICP. This report now suggests the undescribed entity of papilledema associated with normal ICP.

The authors concede that the true opening pressure could have been lowered iatrogenically during general anesthesia. They also speculate that ICP might have been elevated intermittently as in normal pressure hydrocephalus. It is important to point out that disc edema may develop in a gradual, stepwise process that may proceed within a time frame of normal ICP. The authors also suggest that intermittent blockage of the fourth ventricle might have served as a mechanism for the stepwise development of papilledema without being associated with an obvious increase in ICP.

This patient's visual complaints, disc appearance, and ophthalmologic details were rather vague. The visual deterioration to 6/9 or 20/30 Snellen acuity is mild. Although patients with papilledema initially may have mild degrees of visual loss, the authors do not exclude simple refractive errors. The authors describe an enlarged blind spot but failed to investigate this visual field change with a more enlightening full-threshold static perimetry. Had this study been performed, it may have shown a defined scotomatous field defect rather than simple enlarged blind spots. Enlarged blind spots also can be associated with simple optic nerve drusen (congenital anomalous disc elevation), which can mimic disc edema. Furthermore, there is no mention that the visual field defects ever resolved after resection of the acoustic neuroma. Finally, the photographs of the optic nerve are not revealing enough to exclude the possibility of simultaneously present disc drusen. Intravenous fluorescein angiography would have been very useful to distinguish the perfusion patterns of drusen, papilledema, and inflammatory or infiltrative processes. Likewise, contrast sensitivity could have been useful to identify diseases that depress nerve conduction as opposed to nonpathologic anatomic variants with normal conduction.

Despite the limited ophthalmologic details, magnetic resonance imaging showed the large acoustic neuroma compressing the fourth ventricle, and the lesion was removed expeditiously.

This case is important because it highlights possible causes of disc edema associated with normal ICP other than local ophthalmologic processes. This is a unique departure from accepted thinking. Therefore, any patient with a swollen disc should be evaluated thoroughly. This evaluation should include comprehensive neuro-ophthalmologic investigations with full-threshold static perimetry. Although the demise of optic neurons with papilledema is a relatively slow process, space-occupying masses should be treated promptly to minimize optic nerve morbidity. When deciding to forego the use of stereotactic neurosurgical technology in lieu of more immediate surgical decompression, however, the physician should consider all variables, including the degree and duration of papilledema, level of surgical expertise, and finally, the availability of stereotactic technology, rather than the mere presence of disc edema itself.

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Peter Aiello, M.D.¹

Commentary

It isn't just roses that raise the issue of semantics. With the advent of the ophthalmoscope in 1851, the appearance of a swollen disc was initially referred to as "optic neuritis." These early descriptions of optic neuritis included many cases undoubtedly related to intracranial mass lesions and therefore to increased intracranial pressure (ICP). It was not until 1908 that Parsons¹ first used the term papilledema to refer to optic disc swelling associated with ICP. Despite this definition, there has been substantial confusion about the implications of

papilledema ever since. It has been a problem, particularly because both inflammation (papillitis) and ischemia (anterior ischemic optic neuropathy) may appear identical to disc swelling from increased ICP. Local ocular problems, including vitreoretinal traction, hypotony, and intermediate uveitis (pars planitis), also may cause disc swelling that cannot immediately be distinguished from disc edema caused by increased ICP.

The authors present a single case report of a patient with an acoustic neuroma presenting with disc edema. As the authors point out, disc edema has potential implications for visual function. Patients with papilledema may have long-term disc swelling without progressive deterioration in central acuity. However, disc swelling from increased ICP will cause progressive optic nerve damage in a substantial portion of patients, as demonstrated by worsening arcuate visual field defects, the eventual loss of central visual function, decreased acuity, and optic atrophy. Such an outcome was demonstrated fairly dramatically in the case of pseudotumor cerebri or "benign increased intracranial pressure" when Corbett and associates² reported a 70% incidence of optic nerve damage. From an ophthalmic perspective, it is therefore imperative that optic nerve function be assessed quantitatively in any patient with disc swelling. The assessment should include best corrected central acuity, quantitative perimetry (usually automated static perimetry), quantitation of afferent pupillary defect, and analysis of the nerve fiber layer. Although past authors have attempted to follow papilledema by measuring the size of the blind spot, this variable does not correlate with optic nerve function and therefore probably is not particularly helpful. Quantitative perimetry must be performed to quantitatively assess the level of optic nerve pathology. Only quantitative assessment can reveal the subtle changes that can indicate progressive damage.

In this particular case, the authors found no evidence of obstructive hydrocephalus, and reported that ICP was normal. Cerebellopontine angle tumors may increase ICP in several ways. They can block ventricular outflow from the fourth ventricle,

resulting in hydrocephalus. They also can produce problems with cerebrospinal fluid (CSF) absorption from protein leaking into the CSF, causing an obstruction at the level of the pacchionian granulations. It is not surprising to find patients without evidence of ventriculomegaly who still have significant increased ICP. The lack of elevated pressure at surgery, however, does not preclude the possibility that patients still had increased ICP previously, even without ventriculomegaly. ICP can be quite variable, particularly in syndromes such as pseudotumor cerebri where intermittent plateau waves can cause disc swelling despite random measurements of normal pressure on spinal taps. As early as 1939, the following statement was made: "One determination of intracranial pressure in a case of disease of the central nervous system is no more instructive than one determination of a patient's temperature during the course of a fever."³

I would agree with the authors that the most likely scenario was that elevated protein in the CSF related to their patient's acoustic neuroma was responsible for problems with CSF outflow and thus papilledema. I would suggest, however, that the only way that the elevated protein is going to cause papilledema is through increased ICP. Therefore, the term "normal pressure hydrocephalus" is inappropriate because pressure, by definition, is not elevated in normal pressure hydrocephalus and there is no evidence of disc edema. I further agree

with the authors that in the setting of advanced optic nerve pathology, especially with evidence of progressive optic nerve damage, surgical excision offers a much more rapid way of normalizing ICP than treatment with the Gamma knife. The presence of papilledema alone, however, is not necessarily an indication for emergent surgery. Only evidence of progressive optic nerve damage (advancing visual field defects) would constitute a neuro-ophthalmic indication for urgent treatment. Clinical decisions on treatment modalities must include as much quantitative information about the clinical status of the patient as possible. Thus neuro-ophthalmological input can be exceedingly helpful in decision-making and management issues in similar cases.

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Steven A. Newman, M.D.¹

Skull Base, volume 15, number 1, 2005. ¹Private practice, Scottsdale, Arizona. Copyright © 2005 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2005,15,01,086,087,ftx,en;sbs00434x.

Skull Base, volume 15, number 1, 2005. ¹Department of Ophthalmology, University of Virginia Health System, Charlottesville, Virginia. Copyright © 2005 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2005,15,01,087,088,ftx,en;sbs00435x.