Sphenoidal pneumosinus dilatans with bilateral optic nerve meningiomas

Case report

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The authors report a case of bilateral progressive visual loss in a patient with a radiological diagnosis of sphenoidal pneumosinus dilatans. At craniotomy, bilaterally symmetrical meningiomas of the optic nerve sheaths were found at the intracranial optic foramina. Reported cases of sphenoidal pneumosinus dilatans as well as bilateral meningiomas of the optic nerve sheaths are reviewed. The possible mechanisms of visual loss in pneumosinus dilatans and the management of nerve sheath and planum sphenoidale meningiomas are discussed.

KEY WORDS • pneumosinus dilatans • optic nerve meningioma • planum sphenoidale • optic foramen • sphenoid sinus

Sphenoidal pneumosinus dilatans is a rare condition that is felt to be capable of causing progressive optic atrophy and bitemporal field defects. A young man with a radiologically confirmed diagnosis of sphenoidal pneumosinus dilatans presented with progressive loss of visual acuity and optic fields. During the operative procedure to decompress the optic nerves, occult, symmetrical, optic nerve sheath meningiomas were discovered at the intracranial optic foramina.

Case Report

This 18-year-old boy was referred with a 3-month history of decreasing vision in both eyes. He was in excellent general health, and both general and neurological examinations were normal. On neuro-ophthalmological evaluation, his visual acuity was correctable to 20/200 in the right eye and 20/80 in the left eye. He was unable to read any Jaeger lines or recognize any colors on the H-R-R test plates. Tangent screen examination revealed a large ring scotoma breaking into fixation in the right eye, and a smaller ring scotoma in the left eye. In addition, the left peripheral field was slightly constricted. The rest of the examination was normal, except that the left optic disc appeared slightly edematous. Skull films and optic canal polytomography showed diffuse pneumatization of the sphenoidal sinus and upward displacement of the tuberculum sellae (Fig. 1). A computerized tomography (CT) scan was normal. Electroretinography was also normal. A diagnosis of optic nerve compression from pneumosinus dilatans was made.

One month later, the patient returned for follow-up examination. Visual acuity had dropped to 2/200 in the right eye and 20/200 in the left eye. The patient now had a pronounced afferent pupillary defect on the right. The visual fields showed a large central scotoma on the right and a large cecocentral scotoma on the left with slight constriction of both peripheral fields. Ophthalmoscopy revealed bilaterally pale optic discs with diffuse nerve fiber layer loss. There were several drusen of the left optic disc. A diagnosis of Leber’s optic neuropathy was considered.

Examination. The patient was admitted to The Johns Hopkins Hospital where blood studies including serum folate, and B12 levels were normal. The patient and his parents refused further radiological investigation; however, 2 months later, when the patient’s visual acuity had further declined, he was readmitted for further evaluation. At this time, a repeat CT scan...
Bilateral optic nerve meningiomas

Fig. 1. Skull films, anteroposterior (left) and lateral (right) views. Diffuse pneumatization of the sphenoid bone and distortion and displacement of the tuberculum sellae are seen (arrows).

was normal. Cerebral angiography was also normal. A pneumoencephalogram suggested possible enlargement of the intracranial portion of one or both optic nerves.

Operation. The patient underwent a right frontotemporal craniotomy. There was marked prominence of the tuberculum sellae with complete encirclement of the right optic canal and extensive pneumatization of the sphenoid. In addition, a mass contiguous with the optic nerve sheath at the intracranial end of the optic canal was compressing the optic nerve at the foramen and within the canal itself. Biopsy of this mass was read as meningioma. The optic canal was unroofed, and there was no significant extension of the tumor into the canal. It was clear that the tumor was localized to the optic foramen only. The patient was turned, and a left frontotemporal craniotomy was performed. A meningioma, a mirror image of the first, and compressing the left optic nerve, was found at the intracranial end of the left optic canal (Fig. 2). The left optic canal was unroofed. Because both tumors were contiguous with the optic nerve sheaths, no attempt was made to remove them.

Postoperative Course. Postoperatively, the patient’s visual acuity and visual fields were unchanged from preoperative evaluation. They have not changed over a 2-year follow-up period.

Discussion

Pneumosinus dilatans is a rare condition characterized by enlargement of the paranasal sinuses without evidence of localized bone or mucus-membrane changes. Although it may involve the maxillary, ethmoidal, or sphenoidal sinuses, the extent to which it represents a true pathological entity, rather than an anatomical variant of normal sinus architecture, is unclear. To date, seven patients with sphenoidal pneumosinus dilatans have been reported (Table I).1,2,8,9,12,15 All patients had dysfunction of the anterior visual system manifest as slowly progressive visual acuity loss, optic atrophy, and either bitemporal field defects, or grossly constricted fields in those patients with advanced atrophy.

In our patient, polytomography of the optic canals revealed no specific abnormality. Although polytomography of the optic canal was performed in only

Fig. 2. Artist’s representation of bilateral meningiomas of the optic nerve sheaths at craniotomy.
TABLE 1

Clinical course of eight patients with sphenoidal pneumosinus dilatans*

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Sex, Age (yrs)</th>
<th>Visual Acuity</th>
<th>Visual Fields</th>
<th>Fundus</th>
<th>Radiological Findings</th>
<th>Operation</th>
<th>Progress</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bendescu, 1932</td>
<td>F, 24</td>
<td>rt: 1/20; lt: CF</td>
<td>rt: constricted to 10%</td>
<td>optic atrophy bilat</td>
<td>rt: within normal limits; lt: slight constriction of optic canal, distortion of clinoids &amp; sella</td>
<td>transmaxillary sinus sphenoidal decompression on rt</td>
<td>rt improved, 1/20 to 1/10; lt unchanged</td>
</tr>
<tr>
<td>Agati, 1946</td>
<td>M, 54</td>
<td>bitemporal hemianopia</td>
<td>optic atrophy bilat</td>
<td>normal optic canals &amp; upward displacement of sella</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Lombardi, et al., 1968</td>
<td>M, 38</td>
<td>decreased bilat</td>
<td>bitemporal hemianopia</td>
<td>normal bilat</td>
<td>pneumatized sphenoid &amp; ethmoid cells</td>
<td>craniotomy; noncommunicating sphenoid sinus found</td>
<td>—</td>
</tr>
<tr>
<td>Macialowicz, 1969</td>
<td>M, 76</td>
<td>rt: decreased; lt: NLP</td>
<td>optic pallor bilat</td>
<td>normal optic canals; ethmoid &amp; sphenoid enlargement, elevation of planum sphenoidale &amp; sella</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Williams, et al., 1975</td>
<td>M, 73</td>
<td>rt: 10/20; lt: 10/70</td>
<td>bitemporal constriction</td>
<td>lt: optic atrophy</td>
<td>enlarged sphenoid sinus, absence of pituitary fossa, upward displacement of rt anterior cerebral artery</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>Sugita, et al., 1977</td>
<td>M, 26</td>
<td>rt: CF; lt: 20/70</td>
<td>bitemporal field constrictions</td>
<td>rt: optic pallor; lt: normal</td>
<td>enlarged sphenoid sinus, ? defects around canals; small deformed sella</td>
<td>transmaxillary sphenoidal decompression</td>
<td>2 mos postop visual acuity: rt: 20/70; lt: 20/20; field improvement unchanged</td>
</tr>
<tr>
<td>Hirst, et al., 1979</td>
<td>M, 18</td>
<td>rt: 20/200; lt: 20/80</td>
<td>rt: ring scotoma; lt: paracentral scotoma</td>
<td>optic atrophy bilat</td>
<td>sphenoid sinus enlargement, planum sphenoidale elevation; normal optic canals</td>
<td>craniotomy with unroofing of both canals; bilat optic nerve-sheath meningiomas &amp; pneumatized optic canals found</td>
<td>—</td>
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</tbody>
</table>

*CF = counting fingers; NPL = no perception of light.

two of the other seven cases, routine canal and foraminal x-ray films revealed no abnormality apart from sinus enlargement and upward displacement of the planum sphenoidale with distortion of the sella turcica. In view of these findings, the pathogenesis of the anterior visual system dysfunction and bitemporal field loss in these patients is unclear. Gradual constriction of the optic canals could result in direct compression or ischemia of the optic nerves, but this does not, in itself, explain a characteristic presentation of bitemporal field loss. However, the upward displacement of both the pituitary fossa and planum sphenoidale, and distortion of the tuberculum sellae suggest the possibility that there may be either direct compression of the optic chiasm, or interference with chiasmal vasculature. Apart from our patient, only one other case of presumed pneumosinus dilatans has undergone intracranial exploration. Although that patient demonstrated pituitary endocrinological dysfunction, no abnormality was found at craniotomy apart from an enlarged sphenoidal sinus. The optic canals were not unroofed in this patient.

In our patient, bilateral meningiomas at the optic foramina compressed both optic nerves leading to progressive loss of vision and bilateral optic atrophy that was initially thought to be due to pneumosinus dilatans. A total of seven patients with histologically-proven bilateral optic nerve sheath meningiomas have been reported, and another two patients with bilateral signs suggestive of such tumors have been explored on one side only (Table 2). All patients have illustrated clinical features similar to those seen in our
**TABLE 2**

Clinical course of 10 patients with bilateral optic nerve meningiomas*

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Sex, Age (yrs)</th>
<th>Visual Acuity</th>
<th>Visual Fields</th>
<th>Fundus</th>
<th>Radiological Findings</th>
<th>Operation</th>
<th>Progress</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schott, 1877</td>
<td>F, 55</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dandy, 1922</td>
<td>F, 13</td>
<td>rt: NLP; lt: 8/200</td>
<td>lt: constricted</td>
<td>optic nerve pallor bilat</td>
<td>pineal calcification &amp; slight sellar enlargement</td>
<td>bilat symmetrical 1.5-cm cuff tumors at optic foramen with extension into orbit on rt</td>
<td>lt: visual acuity 20/200 &amp; nasal field 2 weeks postop</td>
</tr>
<tr>
<td>Craig &amp; Gogela, 1950</td>
<td>M, 46</td>
<td>rt: 6/15; lt: NLP</td>
<td>rt: nasal field remnant</td>
<td>bilat pale optic nervehead, elevated on rt</td>
<td>normal</td>
<td>3 small meningiomas, lt cuff meningioma at optic foramen removed; similar cuff tumor on rt partially removed</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>NLP bilat</td>
</tr>
<tr>
<td>F, 30</td>
<td>rt: hand motions; lt: NLP</td>
<td>rt: temporal field remnant</td>
<td>bilat pale optic nervehead, elevated on rt</td>
<td>normal</td>
<td>optic foramen cuff tumors; rt canal decompression &amp; removal</td>
<td>rt: subjective improvement</td>
<td></td>
</tr>
<tr>
<td>James &amp; Lawton Smith, 1971</td>
<td>M, 9</td>
<td>rt: 20/40; lt: NLP</td>
<td>rt: nasal field remnant</td>
<td>optic atrophy bilat</td>
<td>possible rounding off of anterior clinoids</td>
<td>symmetrical tumors compressing from lateral aspect of optic nerve at foramen</td>
<td>unchanged</td>
</tr>
<tr>
<td>F, 24†</td>
<td>rt: NLP; lt: 20/50</td>
<td>lt: superior nasal remnant</td>
<td>rt: transient optic nervehead edema progressing to atrophy; optociliary shunt</td>
<td>delta scan, bilat optic nerve involvement</td>
<td>rt canal unroofed with tumor extending under nerve to orbit; lt not explored</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>Hollenhorst, et al., 1978</td>
<td>M, 26</td>
<td>rt: 20/30;†</td>
<td>rt: contracted</td>
<td>bilat pale optic nervehead; optociliary shunt on rt</td>
<td>normal</td>
<td>rt tumor lateral &amp; inferior position in optic canal with later re-exploration; lt not explored</td>
<td>lt: decreasing vision</td>
</tr>
<tr>
<td>F, 18</td>
<td>rt: NLP; lt: 20/70</td>
<td>lt: arcuate scotoma</td>
<td>bilat pale optic nervehead; optociliary shunt on rt</td>
<td>normal</td>
<td>symmetrical cuff tumors on dorsum of optic canal bilat; bilat removal</td>
<td>unchanged</td>
<td></td>
</tr>
<tr>
<td>Trobe, et al., 1978</td>
<td>F, 33</td>
<td>rt: CF; lt: NLP</td>
<td>rt: central scotoma with inferior &amp; superior peripheral constriction</td>
<td>optic pallor bilat</td>
<td>planum sphenoidale hyperostosis rt optic canal with erosion planum sphenoidale on CT scan</td>
<td>bilat cuff meningiomas at intracranial optic foramen with planum sphenoidale involvement; rt canal unroofed</td>
<td>rt: slight improvement</td>
</tr>
<tr>
<td>Hirst, et al., 1979</td>
<td>M, 18</td>
<td>rt: 20/200; lt: 20/80</td>
<td>rt: ring scotoma; lt: paracentral scotoma</td>
<td>optic atrophy bilat</td>
<td>upward displacement of planum sphenoidale, enlargement of sphenoid sinus</td>
<td>craniotomy: both canals unroofed, bilat cuff meningiomas at intracranial optic foramen with pneumatization of optic canals</td>
<td>stable</td>
</tr>
</tbody>
</table>

*CF = counting fingers; NLP = no perception of light.  
†Bilaterality of meningiomas unproven.
patient, including slowly progressive visual acuity loss associated with the development of bilateral optic atrophy. Three patients had transient optic disc edema, and three patients had optic-ciliary shunt vessels. All nine patients reported have shown irregular constriction of their peripheral fields. Although "bitemporal field defects" have been described in two cases, the patients both underwent visual field examination at a stage of advanced atrophy with no light perception in one eye and only a nasal field remnant in the other.

Although the association of bone changes such as blistering, and dilatation of the sphenoidal sinus, in association with planum sphenoidale meningiomas has been well described, small meningiomas at the optic foramen are generally radiologically silent. A variety of examinations including pneumoencephalography, cerebral angiography, CT, and polytomography of the optic canals were performed on eight of the previously reported cases. In only one of these cases was any abnormality noted by the radiological investigations.

Progressive optic atrophy in spite of normal neuroradiological investigations has long been recognized as an indication for surgical exploration of the optic canals. In most cases, occult meningiomas have been discovered. Because it is hard to explain the pathogenesis of visual acuity and field loss in patients with sphenoidal pneuunosinus dilatans and because of the possibility of concomitant compressive pathology as in our patient, progression of visual loss in these patients places them in the same category as those with no radiological findings at all.

Meningiomas that arise at the intracranial end of the optic canal may originate from the dural covering of the optic nerve (as in our patient) and are therefore contiguous with the optic nerve sheath. Attempts at removal or resection of nerve-sheath meningiomas in this location have not resulted in significant improvement in visual acuity or visual field in any of the previously described seven cases. The success of surgical treatment of foraminial meningiomas is related to the ability of the neurosurgeon to remove the tumor without injury to the optic nerve or its blood supply. Because of the intimate relationship of the nerve-sheath meningiomas with the actual optic nerve dura, attempts at removal have resulted in interruption of optic nerve vasculature and pronounced visual loss. However, a similar tumor may arise from the dura of the planum sphenoidale or tuberculum sellae and compress the optic nerve. Such tumors are not contiguous with the optic nerve sheath. Careful microscopic dissection of planum sphenoidale meningiomas that compress the optic nerve at the intracranial end of the optic canal, but are anatomically separate from the optic nerve sheath, may result not only in preservation of visual function but also recovery of visual acuity and field. There is a surgical plane of dissection between these tumors and the optic nerve sheaths so that optic nerve vasculature is maintained.

Although no patients, including our own, with optic nerve-sheath meningiomas at the foramen or in the canal, have improved after unroofing the optic canal, this procedure has generally been undertaken only in those cases with advanced optic atrophy. It is possible that earlier intervention may result in preservation of visual function. Apart from our patient, two other cases with moderate visual function depression only, have had decompression of the pneuunosinus dilatans by a transmaxillary approach. The optic nerve involvement in these cases may have been due to increased pressure within the sphenoidal sinus transmitted to the optic nerve through defects in the pneumatized optic canal.

The differential diagnosis of painless, slowly progressive optic atrophy in the absence of other neurological signs or radiological abnormalities, remains between progressive compressing lesions, and the rare optic neuritides such as Leber's optic neuropathy. Unless there is absolute evidence of such a neuropathy, surgical exploration remains the only method of establishing a diagnosis.

Although sphenoidal pneuunosinus dilatans is rare, it appears to remain an unexplained cause of progressive visual acuity loss and bitemporal visual field defects. Whether the radiological appearance of pneuunosinus dilatans is in fact a sign of meningioma is unclear. Nevertheless, because of the difficulty in explaining the visual field loss simply on the basis of pneumatization of the sphenoidal bone and in view of our patient with presumed pneuunosinus dilatans who was found at surgery to have bilateral optic nerve-sheath meningiomas, we believe that patients with neuroradiological signs of pneuunosinus dilatans warrant exploration, not only to establish a diagnosis but also to attempt decompression of the optic nerve by unroofing the optic canal.

Addendum

Since this paper was reported in part at the Tenth Annual Neuro-Ophthalmic Pathology Conference in Baltimore, Maryland, in 1979, two other patients with progressive optic neuropathy and radiological evidence of sphenoidal pneuunosinus dilatans have undergone intracranial exploration (H. Stanley Thompson, M.D., Iowa City, Iowa, personal communication, 1979; Ronald M. Burde, M.D., St. Louis, Missouri, personal communication, 1979). Both patients were discovered at operation to have occult meningiomas compressing the intracranial and intracanalicular optic nerves. We believe that these two case reports further support the theory that sphenoidal pneuunosinus dilatans associated with progressive optic neuropathy may represent a sign of occult meningioma.
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References


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