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CASE REPORT

Recurrent Optic Neuropathy Caused by a Mucocele of the Anterior Clinoid Process after a 5-Year Remission: A Case Report and Literature Review

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ABSTRACT

A 32-year-old male presented with acute left vision loss during a second recurrence of optic neuropathy. Steroid pulse therapy had been effective in both the first episode 9 years previously and the first recurrence 5 years previously. Magnetic resonance imaging demonstrated an anterior clinoid process mucocele compressing the optic nerve. Although surgical treatment was performed, improvement was limited. This report indicates that steroid pulse therapy could be an alternative treatment to obtain temporary remission, but surgical treatment should be considered to prevent irreversible neurological deficits. This paper also presents a review of the literature on anterior clinoid process mucoceles.

Keywords: Anterior clinoid process, craniotomy, high-dose methylprednisolone, mucocele, optic neuropathy

INTRODUCTION

Mucocele formation in a pneumatised anterior clinoid process is a rare condition.[1,2] Expansion of the mucocele may cause headache, vision loss, visual field deficits, ophthalmoplegia, diplopia, ptosis, or color blindness secondary to multiple cranial nerve paralysis.[2–7] The optic nerve is most commonly affected,[7] resulting in visual impairment due to compression or inflammation of the nerve.[8,9] Because this condition may have a poor prognosis,[3,4,6] urgent surgical treatment including craniotomy or endoscopic drainage has been proposed.[2,5,7,9,10] The long-term course of this disease without surgical treatment has not been well described, especially when alternative treatment resulted in improvement and remission for several years. We herein present a case involving a patient with optic neuropathy due to mucocele formation in a pneumatised anterior clinoid process treated with steroid pulse therapy. Improvement and remission were achieved for several years, but recurrent optic neuropathy then developed. We also review the literature on anterior clinoid process mucoceles causing visual dysfunction or oculomotor deficits.

CASE REPORT

A 32-year-old male visited our hospital for evaluation of a 6-day history of headache and acute visual loss and dull pain in his left eye. The initial neuro-ophthalmological evaluation revealed a corrected visual acuity of 20/20 in his right eye and counting fingers at 10 cm in his left eye. A positive relative afferent pupillary defect was observed in his left eye. The patient's ocular movements were normal. A reduced face sensation was not detected. Slit-lamp examination revealed a normal anterior segment. Dilated fundus examination revealed left optic nerve atrophy. His laboratory data were unremarkable and showed no signs of infection. He had no history of chronic sinusitis or nasal or sinus surgery. He had a similar ocular history of sudden visual loss in his left eye that had occurred twice (9 and 5 years previously). He reported that he had received steroid pulse

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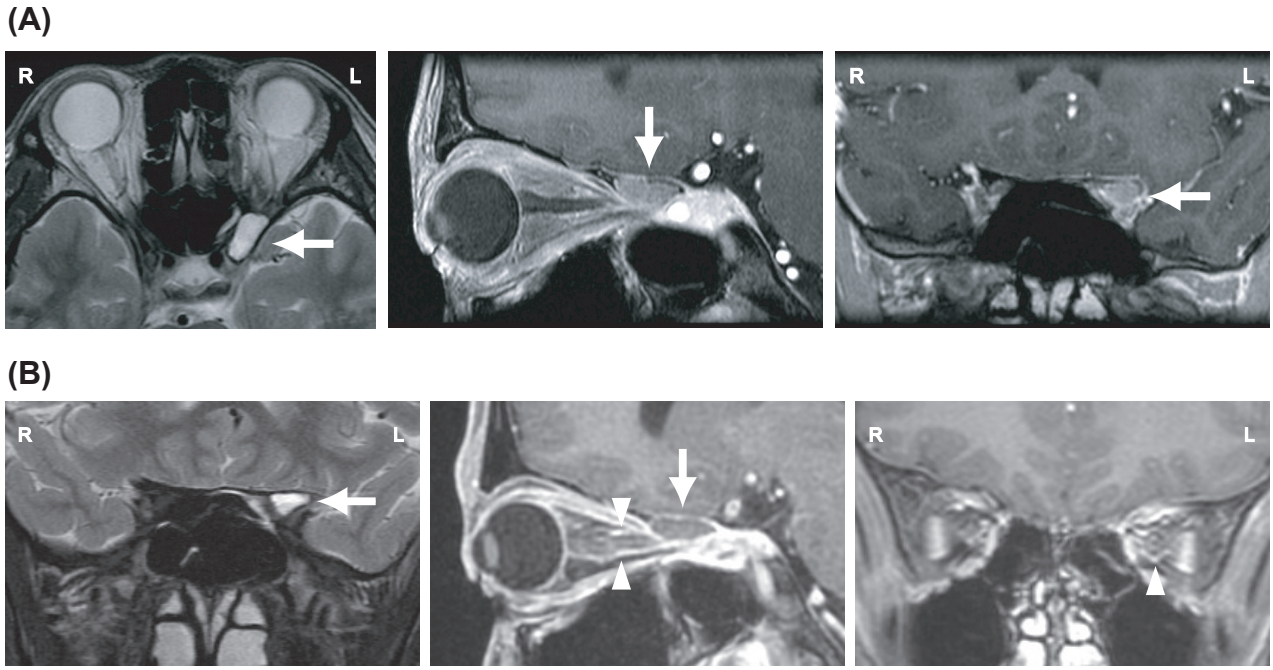

FIGURE 1 (A) T2-weighted MR image (left panel: axial image) and gadolinium-enhanced T1-weighted MR image (middle and right panels: parasagittal image and coronal image, respectively) of the brain at the first visit. A cystic lesion of high signal intensity without contrast enhancement on T1- and T2-weighted images was present in the left optic canal, compressing the optic nerve (white arrows). (B) T2-weighted MR image (left panel: coronal image) and gadolinium-enhanced T1-weighted MR image (middle and right panels: parasagittal image and coronal image, respectively) at the first recurrence 5 years previously. T2-weighted (left panel) and parasagittal T1-weighted (middle panel) images show a cystic lesion compressing the optic nerve (white arrows). Arrowheads in the middle panel indicate a slight enhancement effect in the optic nerve sheath. The enhancement is also observed in the coronal section (right panel, arrowhead) taken at the level indicated by the arrowheads in the middle panel.

therapy for each episode and that his visual acuity had recovered each time. Computed tomography demonstrated a cystic lesion between the anterior clinoid process and the sphenoid sinus, and both T1- and T2-weighted magnetic resonance (MR) images demonstrated high signal intensity without gadolinium contrast enhancement in the same region compressing the left optic nerve (Figure 1A). Because the patient declined surgical treatment, high-dose methylprednisolone (1000 mg/day, 3 days, two courses) was intravenously administered followed by an oral taper, as previously prescribed. However, the improvement in his visual acuity was poor (20/400), and a residual central scotoma was revealed by Goldmann perimetry (Figure 2A). The cystic lesion remained on follow-up MR images. According to the patient's medical records, the MR images during the first recurrence (5 years previously) demonstrated that the cystic lesion was already present in the same region and was compressing the optic nerve (Figure 1B, left and middle panels); slight enhancement at the optic nerve sheath was also evident (Figure 1B, middle and right panels). High-dose methylprednisolone (1000 mg/day, 5 days) was effective during that episode. The patient's visual acuity improved from counting fingers at 20 cm to 20/28. Goldmann perimetry revealed remarkable recovery from only a small visual field in the upper nasal region to almost a full visual field with a residual small inferior paracentral relative scotoma (Figure 2C, D).

For improvement in visual function and to prevent another recurrence of optic neuropathy, the patient underwent craniotomy for resection of the cyst, which was diagnosed as a mucocele by pathological examination. After the operation, his visual acuity slightly improved to 20/300, but the central scotoma remained during the 6-month follow-up period (Figure 2B).

DISCUSSION

A pneumatised anterior clinoid process is reportedly observed with an incidence of 4% to 29%.[11] The formation of an anterior clinoid process mucocele is considered to occur secondary to closure of the passage from the paranasal sinus that aerates the pneumatised anterior clinoid process.[1,2,7,9] Because expansion of the mucocele may cause severe visual impairment, prompt surgical treatment is recommended.[7,9,10]

In the present case, steroid pulse therapy was effective in two previous ocular episodes. Acute retrobulbar neuritis could have been a differential diagnosis;[12] however, we failed to identify obvious inflammation in the optic nerve itself on the brain MR

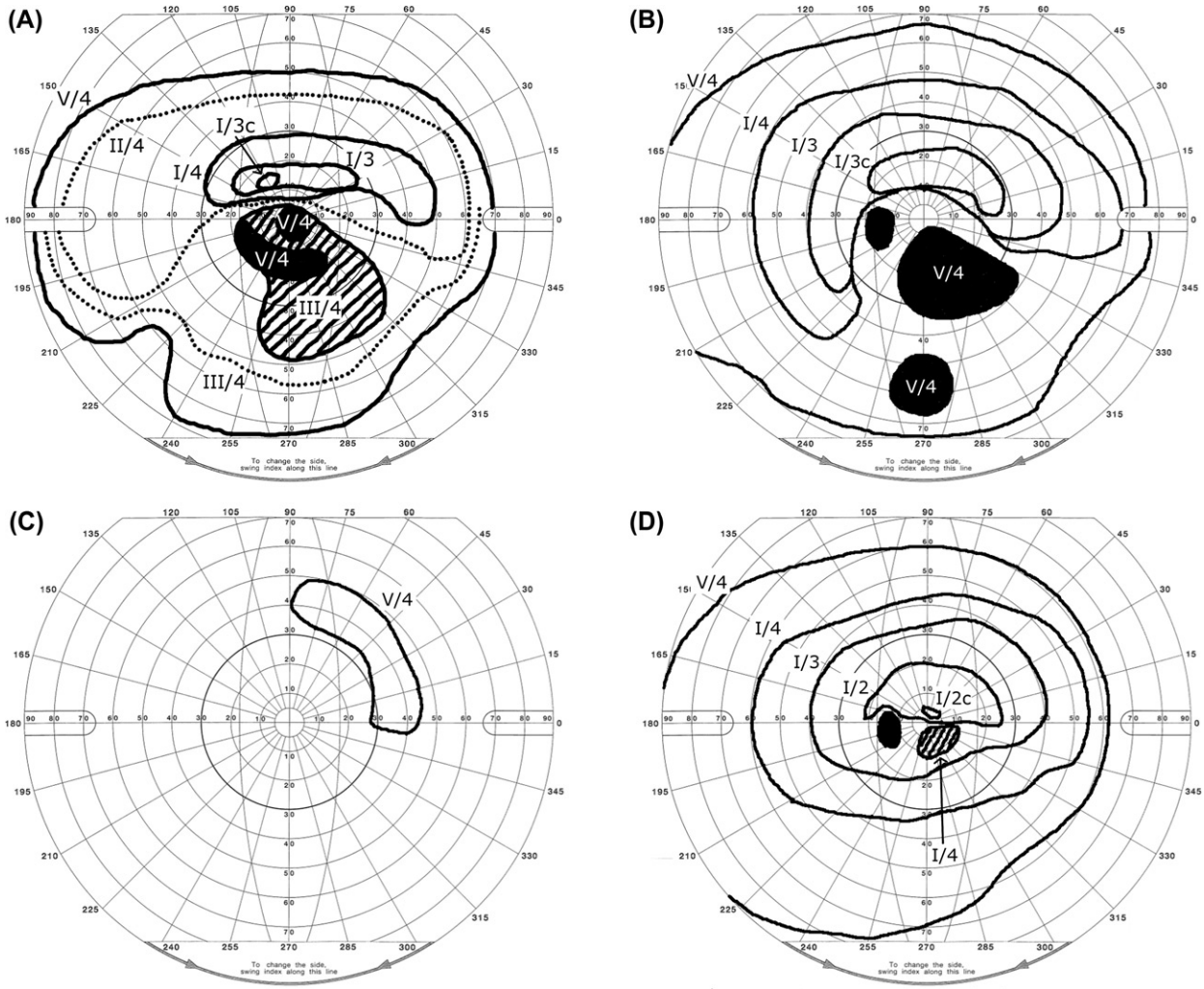

FIGURE 2 Visual field of the left eye plotted with Goldmann perimetry (A) after steroid pulse therapy and (B) after mucocele resection. The improvement was limited. The results of the first recurrence 5 years previously are displayed in (C) (before steroid pulse therapy) and (D) (after steroid pulse therapy). The visual filed remarkably improved with steroid pulse therapy. The painted-out and hatched regions indicate an absolute and a relative scotoma, respectively.

images during the first recurrence. Instead, slight enhancement of the optic nerve sheath on the parasagittal and coronal slices was observed in addition to the cystic lesion (Figure 1B, middle and right panels). It is plausible that the inflammation secondary to the mucocele spread into the optic nerve sheath and that the corticosteroid reduced the inflammation in the optic nerve sheath, resulting in the improvement and 5-year remission. The effectiveness of systemic steroids for optic neuropathy caused by sphenoid sinus mucoceles has been described; in those cases, the chief pathogenesis of the optic neuropathy was considered to be inflammation rather than compression.[13] On the other hand, in the current episode (second recurrence), visual recovery was more limited than that after the first recurrence. Because of the obvious compression of the optic nerve by the mucocele without obvious enhancement in either the optic nerve or optic nerve sheath on MR images, the chief pathogenesis in the current episode was considered to be optic nerve compression.[9] It is possible that the size of the mucocele increased to some extent during the 5 years, although it was difficult to distinguish a difference in size based on the MR images.

Recurrence of optic neuropathy caused by a cyst in the anterior clinoid process was described in a previous report.[3] Although the pathology was uncertain, the cyst in that case was considered to be a mucocele; the patient's visual acuity improved after oral prednisolone, but optic nerve atrophy occurred. Both that case and ours indicate that steroid pulse therapy may be an alternative, temporarily effective treatment; however, it may be insufficient to prevent deterioration of optic nerve atrophy. Surgical treatment should be considered for a better visual prognosis because of reduction of the operative risk with advances in surgical techniques and instruments.[2]

We reviewed the literature of anterior clinoid process mucoceles causing visual dysfunction or oculomotor deficits (Table 1). Among all identified

TABLE 1. Reported cases of anterior clinoid process mucoceles.

Article	Age (year)	Sex	Visual impairments	Diplopia or ocular movement disorder	Pain	Hypesthesia	Treatment	Outcome
Johnson et al.[3] (1986)	56	M	Light perception	+	Supraorbital		Craniotomy	Slight improvement in vision (from light perception to hand motion)
Johnson et al.[3] (1986)	59	M	20/40	–	None	–	Oral prednisone	Unchanged
Schwaighofer[1] et al. (1989)	34	F	Sudden loss of vision	–	Retro-orbital	–	Craniotomy	Full recovery
Dunya et al.[12] (1996)	32	M	20/400	+			Endoscopic surgery	Full recovery
Chou et al.[4] (1999)	68	M	No light perception	+	Headache	+	Craniotomy	Substantial improvement in visual acuity (from no light perception to hand motion)
Chung et al.[14] (1999)	48	F	—	+	Headache		Craniotomy	Full recovery in visual acuity
Lim et al.[5] (1999)	61	M	—	+	Retro-orbital	+	Craniotomy	Full recovery
Deshmukh and DeMonte[8] (2007)	20	F	Visual field defect				Antibiotics	Full recovery
Thurtell et al.[6] (2007)	50	F	No light perception	–	Orbital	–	Intravenous corticosteroid followed by craniotomy	No recovery
Vaphiades et al.[15] (2007)	36	M	20/80	–	None	–	Endoscopic surgery	Substantial improvement in visual acuity (from 20/80 to 20/25)
Kwon et al.[16] (2009)	52	M	0.01	–			Endoscopic surgery	Substantial improvement in visual acuity (from 0.01 to 0.8)
Chagla et al.[9] (2010)	38	F	Light perception	–	Headache	–	Craniotomy	Substantial improvement in visual acuity (from light perception to 6/36)
Forer et al.[2] (2010)	50	M	—	+	None		Endoscopic surgery	Full recovery
Nundkumar et al.[10] (2012)	32	M	Hand motion	–	None	–	Endoscopic surgery	Full recovery
Wang et al.[7] (2013)	44	M	Nonspecific visual fields deficits	+	Retro-orbital	+	Craniotomy and high-dose dexamethasone	Almost full recovery
Hopf-Jensen et al.[17] (2014)	66	M	—	+	None	–	Craniotomy	Full recovery

M = Male; F = Female.

cases, only two involved nonsurgical treatment.[3,8] In another case, a patient with severe sinusitis and ear infection presented with visual field loss and received oral antibiotics, resulting in full visual recovery.[8] Because our patient did not have sinusitis or any symptoms of infection, antibiotics were not considered. Cystic lesions may mimic neoplasms such as meningiomas or schwannomas, which should be considered as differential diagnoses. However, their possibility is unlikely because of the lack of contrast enhancement on MR images, lack of calcification on computed tomography, and hyperintensity on both T1- and T2-weighted MR images.

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REFERENCES

[1] Schwaighofer BW, Sobel DF, Klein MV, Zyroff J, Hesselink JR. Mucocele of the anterior clinoid process: CT and MR findings. *J Comput Assist Tomogr* 1989;13:501–503.

[2] Forer B, Hui NY, Sethi DS. Unilateral ophthalmoplegia secondary to anterior clinoid process mucocele. *J Neuroophthalmol* 2010;30:321–324.

[3] Johnson LN, Hepler RS, Yee RD, Batzdorf U. Sphenoid sinus mucocele (anterior clinoid variant) mimicking diabetic ophthalmoplegia and retrobulbar neuritis. *Am J Ophthalmol* 1986;102:111–115.

[4] Chou PI, Chang YS, Feldon SE, Chen JT. Optic canal mucocele from anterior clinoid pneumatisation. *Br J Ophthalmol* 1999;83:1306–1307.

[5] Lim CC, Dillon WP, McDermott MW. Mucocele involving the anterior clinoid process: MR and CT findings. *AJNR Am J Neuroradiol* 1999;20:287–290.

[6] Thurtell MJ, Besser M, Halmagyi GM. Anterior clinoid mucocele causing acute monocular blindness. *Clin Exp Ophthalmol* 2007;35:675–676.

[7] Wang AC, Than KD, Ramnath S, Pandey AS. Anterior clinoid mucocele presenting with orbital apex syndrome. *Surg Neurol Int* 2013;4:63.

[8] Deshmukh S, DeMonte F. Anterior clinoidal mucocele causing optic neuropathy: resolution with nonsurgical therapy. Case report. *J Neurosurg* 2007;106:1091–1093.

[9] Chagla AS, Bhaganagare A, Kansal R, Tyagi D. Complete recovery of visual loss following surgical treatment of mucopyocele of the anterior clinoid process. *J Clin Neurosci* 2010;17:670–672.

[10] Nundkumar N, Mittal M, Kupsky WJ, Folbe A, Mittal S. Complete recovery of acute monocular visual loss following endoscopic resection of anterior clinoid mucocele: case report and review of the literature. *J Neurol Sci* 2012;312:184–190.

[11] Mikami T, Minamida Y, Koyanagi I, Baba T, Houkin K. Anatomical variations in pneumatization of the anterior clinoid process. *J Neurosurg* 2007;106:170–174.

[12] Dunya IM, Frangieh GT, Heilman CB, Miranda MR, Rand LI, Hedges TR. Anterior clinoid mucocele masquerading as retrobulbar neuritis. *Ophthal Plast Reconstr Surg* 1996;12:171–173.

[13] Yasuda Y, Morita T, Akiguchi I, Kimura J, Kameyama M. Sphenoid sinus mucocele with recurrent visual disturbance. *Eur Neurol* 1992;32:225–227.

[14] Chung DS, Park YS, Lee JH, Kang JK. Mucocele of the anterior clinoid process: case report. *Neurosurgery* 1999;45:376–378.

[15] Vaphiades MS, Yunker JJ, Roberson GH, Meyer DR, Mills DM. Optic neuritis is nothing to sneeze at. *Surv Ophthalmol* 2007;52:106–110.

[16] Kwon SH, Kim SH, Yoon JH. Anterior clinoid mucocele coexisting with sphenoid sinus mucocele. *Auris Nasus Larynx* 2009;36:598–600.

[17] Hopf-Jensen S, Rubarth O, von DAI, Riis P, Preuss H, Preiss M, Borm W, Muller-Hulsbeck S. Isolated oculomotor nerve palsy caused by a mucocele of an aerated anterior clinoid process. *Clin Neuroradiol* 2014;24:161–164.