CASE REPORT

Successful surgical optic nerve decompression in a patient with hypertrophic pachymeningitis due to granulomatous polyangiitis

Joeri de Hoog,¹ Victor Volovici,² Ruben Dammers²

¹Department of Ophthalmology, Erasmus Medical Center, Rotterdam, The Netherlands
²Department of Neurosurgery, Erasmus Medical Center, Rotterdam, The Netherlands

Correspondence to

Joeri de Hoog,
 j.dehoog@erasmusmc.nl

Accepted 8 January 2015

SUMMARY

A 57-year-old woman presented with subacute vision loss of first the left, and later the right eye. She was diagnosed with granulomatous polyangiitis with hypertrophic pachymeningitis and optic nerve compression. Her visual acuity could not be permanently restored with immune suppressants alone, so a surgical decompression of the right optic nerve, via a modified cranio-orbitozygomatic pretemporal approach, was performed. Her right eye regained 20/20 vision and has remained stable during 8 months of follow-up.

BACKGROUND

The central nervous system (CNS) is involved in 7–11% of patients with granulomatous polyangiitis (GPA).¹ According to Seror *et al.*,² CNS involvement may either develop as a consequence of the invasion of the granulomatous tissue from the sinuses and the orbit or the inflammation can develop in CNS tissue itself (eg, hypertrophic pachymeningitis). Although decreased visual acuity (VA) as a result of optic nerve compression appears in 17% of all orbital and other cases,³ decreased acuity as a result of hypertrophic pachymeningitis is extremely rare and mainly described in case reports.^{4–5} By writing this report we would like to provide evidence that surgical decompression can be an effective therapy in this rare complication of GPA.

CASE PRESENTATION

A 57-year-old Caucasian woman from Croatia was referred with total blindness of the left eye (LE) occurring within 2 weeks.

Six months earlier she had been examined by a neurologist because of headaches, nausea, transient palsy of cranial nerves III and VIII, and gait imbalance. Two weeks before referral she noticed loss of the lower visual field of the LE, progressing to barely light perception over several days. She was admitted to the neurology department of a district hospital, where the diagnosis of pituitaritis of unknown cause was made, based on MRI findings and exclusion of giant cell arteritis. A high dosage of intravenous methylprednisolone (1000 mg daily, 3 days) was given, with no effect on VA and the patient was referred to our centre.

On referral, VA was 20/20 in the right eye (RE) and no light perception in the LE. Slit lamp and fundus examinations were unremarkable as were optic coherence tomography, fluorescein

angiography and electroretinography. Eye movements were normal and painless. Visually evoked potential testing showed increased latency on the right side, and no response on the left. A prechiasmatic lesion of the optic nerves was considered. Despite the initially present full VA and a normal visual field of the RE, impaired colour vision was documented with Ishihara pseudoisochromatic plates. In the following 2 days, VA of the RE decreased to counting fingers and the patient developed an altitudinal scotoma (figure 1).

Laboratory testing revealed elevated C reactive protein and positive antiproteinase-3 antibodies (cytoplasmic antineutrophil cytoplasmic antibodies), but was otherwise unremarkable. Infectious causes were ruled out. Chest X-ray (figure 2) revealed multiple nodular abnormalities. Metastatic disease as well as GPA were considered. Biopsy of the thoracic lesions revealed reactive changes without necrosis and with no signs of malignancy or granulomas. MRI of the brain was repeated and revealed a hypertrophic dura mater with compression of both optic nerves at the entry point of the optic canals (figures 3 and 4). The presumptive diagnosis of GPA with pachymeningitis and optic nerve compression was made.

TREATMENT

The patient received 150 mg of dexamethasone daily intravenously. This temporarily improved her vision in the RE, which unfortunately decreased after cessation of the high dosage dexamethasone therapy. The intravenous steroids were readministered. Nevertheless, additional cyclophosphamide, plasmapheresis and rituximab could not stop visual decrease in her better RE.

Therefore, the decision to perform a surgical decompression of the right optic nerve was made (figure 5). An epidural anterior clinoidectomy as described by Krisht and Kadri,⁶ via a modified cranio-orbitozygomatic (COZ) pretemporal approach, was performed. The hypertrophic dura mater was sent in for histological analysis and showed a granulomatous vasculitis with neutrophilic and eosinophilic granulocytes and necrosis, confirming the diagnosis of GPA. We chose to operate the optic canal of the eye that was most likely to benefit from surgery, that is, the RE. After the second surgery the patient chose to not undergo a second procedure on the left side, considering the risk of surgery and the small chance of full visual recovery. Owing to little experience with this approach in our

CrossMark

To cite: de Hoog J, Volovici V, Dammers R. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208110

Figure 1 Goldmann kinetic perimetry of the right eye, showing an enlarged blind spot, along with an altitudinal visual field defect.

clinic, a simultaneous decompression of both sides was not performed, to minimise possible complications.

OUTCOME AND FOLLOW-UP

The patient made a swift recovery initially, but the third day after surgery she suffered from a subdural and intraparenchymal haemorrhage in the surgical area, which was evacuated by performing a craniotomy. The haemorrhage was probably due to acetylsalicylic acid use, and developed despite thorough

haemostasis during the first procedure. She made a good recovery, and her VA in the RE reached 20/20 and remained stable during the follow-up of 8 months. Her postoperative medication consisted of dexamethasone 8 mg thrice daily, which was tapered to 0.5 mg twice daily over the course of 6 months. Cyclophosphamide was given monthly during the postoperative 6 months and was stopped thereafter. The patient has continued to use azathioprine 150 mg daily since her surgery. Her systemic condition at follow-up was unremarkable, apart from fatigue possibly due to her immunosuppressive medication. MRI performed 3 months after surgery revealed an adequate decompression of the optic nerve (figure 6).

DISCUSSION

Compression of the optic nerves due to hypertrophic pachymeningitis in GPA is a rare disorder. We have no concise, experience based, let alone evidence based, treatment indications. We suggest that, whenever immunosuppressive therapy fails or very high dosage of steroids and additional suppressants, which cannot be sustained on the long term, are needed to regain VA, surgery is indicated.

A literature review performed in order to prepare the surgical intervention in our case revealed only scarce information. Some reviews on the ocular manifestations of GPA and its treatment do not mention the possibility of surgical intervention at all.⁷⁻⁹ Thorne and Jabs¹⁰ indicate that orbital decompression might become necessary in certain cases with rapid progression but do not elaborate on the subject. Two cases described by Takazawa *et al*⁵ were radiologically almost identical to our patient, but again no surgery was performed, since immune suppression had

Figure 2 Chest X-ray, showing multiple nodular abnormalities.

Figure 3 T2-weighted coronal together with a T2-weighted axial image of the orbit shows an increased subarachnoid space in the intraorbital portion of the optic nerves (white arrows). Notice the sleeve of cerebrospinal fluid surrounding the nerves as the result of their entrapment in the intraorbital subarachnoid space, a consequence of the hypertrophic dura, which appeared to compress the optic nerve at the point where the nerve entered the canal. Transitory inflammation of the dura with acute compression, ischaemia of the optic nerves and subsequent loss of vision was the working diagnosis. This diagnosis was supported by the clinical evidence, the imaging and the fact that the patient's sight would improve after the administration of extremely large doses of dexamethasone.

a sufficient effect. The current recommended medical treatment includes high doses of glucocorticoids and/or cyclophosphamide and, more recently, rituximab.¹¹ Li *et al* describe a retrospective study of extracranial transtentorial decompression of the optic nerve performed on 92 patients, 4 of whom presented with inflammatory disease and 2 of whom had a confirmed diagnosis of GPA with an associated loss of vision. One of the patients improved from no light perception to counting fingers after surgery, while the other showed no improvement.¹²

We deliberated on the approach to be used in order to adequately decompress the optic nerve. We needed to take into account that due to the nature of the disease, that is, hypertrophy of the dura, the dura covering the optic nerve in its canal needed to be opened completely. We felt, therefore, that a purely endonasal approach would not meet this requirement, since it only gives decompression of the lateral wall of the optic canal. A supraorbital or intraorbital approach, for example, as described by Li *et al*,¹² would only result in an unroofing of the optic canal. The approach we used, the COZ approach with extradural anterior clinoidectomy, resulted in the most adequate decompression of the optic nerve over the complete length of the optic canal with resection of dura covering the nerve. Because there was only scarce information in the literature,

however, we only agreed to this approach when complete blindness was imminent. The excellent recovery of the patient suggests that the COZ optic nerve decompression with opening of the dura along its entire course is an effective method of treatment even when compared with the extracranial decompression and might be considered as a treatment option for cases of optic nerve compression in GPA that are refractory to immunosuppressive treatments.

Figure 5 Anatomy of the anterior and middle skull base, sellar and parasellar regions. Observe the entrance to the optic canal. It is bounded on the lateral and inferior sides by the anterior clinoid process, and the optic strut, respectively. After removal of a bone flap in the pterional region (classic pterional approach), the greater wing of the sphenoid (Sph.) bone was drilled and the dural attachments were cut. The lateral wall of the orbit and the roof of the optic canal were opened (modified crani-orbitozygomatic approach). Subsequently, the anterior clinoid could be drilled and eventually removed, as well as the optic strut. The presence of the intracranial part of the internal carotid artery creates the need for extremely precise drilling. Blood loss was more than expected, most likely due to the patient's anticoagulant therapy. With the optic canal decompressed on its lateral side, the dura could be opened, whereby a large quantity of cerebrospinal fluid surrounding the nerve in its intraorbital trajectory was spontaneously evacuated, confirming adequate nerve decompression. The ophthalmic artery was also followed in its intracanalicular portion as it crossed over the optic nerve. ACP, anterior clinoid process; For. Rotund., foramen rotundum; PCP: posterior clinoid process; SOF, superior orbital fissure.

Figure 4 T1-weighted axial image of the orbit shows the left optic nerve being compressed in its canal by the hypertrophic dura (arrowhead). Surgery was performed on the other side, but this image was most illustrative.

Figure 6 T2-weighted coronal together with a T2-weighted axial image of the orbit shows marked reduction of the sleeves of cerebrospinal fluid around the right optic nerve.

Learning points

- ▶ Optic nerve compression is a rare but serious complication of GPA.
- ▶ Optic nerve compression must be considered as a diagnosis in patients presenting with visual loss and hypertrophic pachymeningitis.
- ▶ Surgical decompression can restore full visual function, when treatment with immune suppressants fails.

Contributors JdH has written the article together with VV and RD. JdH is the patient's ophthalmologist, whereas the surgery was performed by RD. All three authors have substantially contributed to the writing of this case report, either in writing or in reviewing and correcting.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Hoffman GS, Kerr GS, Leavitt RY, et al. Wegener granulomatosis: an analysis of 158 patients. *Ann Intern Med* 1992;116:488–98.
- 2 Seror R, Mahr A, Ramanoelina J, et al. Central nervous system involvement in Wegener's granulomatosis. *Medicine* 2006;85:54–65.
- 3 Woo TL, Francis IC, Wilcsek GA, et al. Australasian orbital and adnexal Wegener's granulomatosis. *Ophthalmology* 2001;108:1535–43.
- 4 Liewluck T, Schatz NJ, Flotter PF, et al. Compressive retrobulbar optic neuropathy due to hypertrophic pachymeningitis. *Intern Med* 2008;47:1761–2.
- 5 Takazawa T, Ikeda K, Nagaoka T, et al. Wegener granulomatosis-associated optic perineuritis. *Orbit* 2014;33:13–16.
- 6 Krisht AF, Kadri PA. Surgical clipping of complex basilar apex aneurysms: a strategy for successful outcome using the pretemporal transzygomatic transcavernous approach. *Neurosurgery* 2005;56(2 Suppl):261–73; discussion 261–73.
- 7 Pakrou N, Selva D, Leibovitch I. Wegener's granulomatosis: ophthalmic manifestations and management. *Semin Arthritis Rheum* 2006;35:284–92.
- 8 Holle JU, Laudien M, Gross WL. Clinical manifestations and treatment of Wegener's granulomatosis. *Rheum Dis Clin North Am* 2010;36:507–26.
- 9 Holle JU, Gross WL. Neurological involvement in Wegener's granulomatosis. *Curr Opin Rheumatol* 2011;23:7–11.
- 10 Thorne JE, Jabs DA. Ocular manifestations of vasculitis. *Rheum Dis Clin North Am* 2001;27:761–79.
- 11 Sharma A, Kunar S, Wanchu A, et al. Successful treatment of hypertrophic pachymeningitis in refractory Wegener's granulomatosis with rituximab. *Clin Rheumatol* 2010;29:107–10.
- 12 Li KK, Teknos TN, Lai A, et al. Extracranial optic nerve decompression: a 10-year review of 92 patients. *J Craniofac Surg* 1999;10:454–9.

Copyright 2015 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow