CASE REPORT

Recurrent Optic Perineuritis after Intranasal Cocaine Abuse

S. Coppens, A. Petzold, P. de Graaf, and W. A. E. J. de Vries-Knoppert

VU University Medical Center, Amsterdam, The Netherlands

ABSTRACT

Recurrent optic perineuritis can be related to orbital inflammation. Here we present the case of a 46-year-old male patient in whom recurrent episodes of optic perineuritis were related to chronic osteolytic sinusitis following intranasal cocaine abuse. Magnetic resonance imaging (MRI) demonstrated optic perineuritis adjacent to a soft tissue mass that intruded the orbit from the nasal cavity. Computed tomography (CT) confirmed destruction of the medial orbital wall. *Staphylococcus aureus* was cultured and biopsy showed granulomatous tissue. Visual outcome was poor. We review the literature and discuss the diagnostic pitfalls and management implications in relation to optic (peri)neuritis originating from the nasal sinuses.

Keywords: Cocaine, CT, histology, MRI, optic neuritis, optic perineuritis, osteolytic sinusitis

INTRODUCTION

Marcus Gunn can be credited with the first systematic classification of optic neuritis in the post-ophthalmoscope area: due to (i) infection (orbit, paranasal sinuses, meninges); (ii) infectious disease (e.g. syphilis); and (iii) multiple sclerosis.¹ At the time, syphilis was a frequent differential diagnosis and among others Buzzard noted that compared with multiple sclerosis, visual loss was more severe, recovered poorly and was associated with paler discs.² The osteolytic changes caused by syphilis in the paranasal cavities and the anatomical proximity of the sinuses with the optic nerve made a causal relationship likely. At the time, there were good autopsy data to support the argument, but case numbers decreased over the years, probably most reliably reflected by a single-centre experience (Johns Hopkins), from 8% in 1931 to 0% in 1952.³ In fact, there is very little literature on sinus disease-related optic neuritis in the last couple of decades. The present case illustrates that these early observations have not become obsolete and continue to inform the present differential diagnosis of optic neuritis.

CASE REPORT

A 46-year-old male patient presented to us 2 weeks after onset of severe loss of vision in his left eye. Loss of vision was preceded by sinusitis and perinasal discomfort. There were no other symptoms. He had a past medical history of recurrent sinusitis and two earlier episodes of visual loss. Five years ago, the best-corrected visual acuity (BCVA) in the right eye was reduced within 3 days to perception of light (PL) and BCVA was 1.0 in the left eye. A diagnosis of optic neuritis was made and he was treated with intravenous methylprednisolone (IVMP). The BCVA returned to 1.0 in the right eye and remained unchanged on the left side. The second episode of visual loss occurred 1 year ago during an episode of sinusitis. This time the left eye was affected and BCVA decreased to 0.6, with complete visual recovery (BCVA 1.0) under oral antibiotics.

The family history was non-contributory. He used to smoke 20 cigarettes a day and regularly sniffed cocaine for many years.

On examination we saw a healthy-looking, well-dressed, adequate, and fully alert right-handed man. There was a left relative afferent pupillary defect

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Correspondence: S. Coppens, VU University Medical Center, De Boelelaan 1117, 1081 HV, Amsterdam, The Netherlands. E-mail: s.coppens@vumc.nl

(RAPD). The BCVA was 1.0 on the right and 0.02 on the left. There was no papilloedema, but the left optic disc was pale compared with the right. There were no other ophthalmological or neurological abnormalities, including eye motility restrictions or proptosis. The rhinological examination showed a large nasal septum perforation with diffuse mucosal inflammation.

The Goldmann perimetry revealed a large, dense central scotoma on the left and was normal on the right (data not shown).

He underwent imaging to investigate whether the sinusitis extended to the orbit or optic canal. To our surprise, magnetic resonance imaging (MRI) demonstrated a soft-tissue mass in the nasal cavity with extension into the left orbit (Figure 1A). The soft tissue was in direct contact with the left optic nerve and showed a perineural enhancement suspicious of optic perineuritis

(Figure 1B and C). MRI showed extensive (reactive) dural enhancement at the anterior skull base. Computed tomography (CT) showed an extensive destruction of the left medial orbital wall (Figure 1D). There was also some anterior bony destruction on the right.

The CT and MR images of the left orbita were compared with those (retrospectively) obtained 5 years earlier from a different hospital in The Netherlands (Figure 2A–D). The CT showed bony destruction of the medial orbital wall on the left side and a small bone defect co-localising with the larger area of destruction on the right side 5 years later (Figure 2A). Less perineural enhancement was seen in the left orbit (Figure 2B). In contrast, profound enhancement was seen in the right orbital apex (Figure 2C), combined with an extensive dural enhancement and a slide swelling of the right optic nerve (Figure 2D).

FIGURE 1 Axial (A) and coronal (B and C) contrast-enhanced T1-weighted MR images with fat suppression show a large soft-tissue mass invading the orbit from the nasal cavity (arrows) (A). The proximity of this soft-tissue mass to the left optic nerve is better appreciated on the coronal images (B and C). There is ring of contrast enhancement surrounding the left optic nerve borders without contrast enhancement of the optic nerve itself (arrow in B), which is suggestive of optic perineuritis. (D) Coronal computed tomography (CT) demonstrated destruction of the left medial orbital wall and some damage also to the right medial orbital wall. Bilaterally, the wall of the maxillary sinus is thickened and sclerotic due to chronic sinusitis.

FIGURE 2 Five years prior to admission, the patient presented to a different hospital for a similar problem. Scan alignment was not identical to those images shown in Figure 1. The closest approximation for the coronal CT (A) and the coronal contrast-enhanced T1-weighted MR images with fat suppression (B, C, and D) are presented. (A) CT scan shows the same large defect in the medial orbital wall on the left side. On the right side, CT imaging retrospectively suggests a small bone defect (arrow), which co-localises with the larger area of bone destruction seen in 2012 (compared with Figure 1D). Contrast enhancement of a comparable mass lesion invading the orbit through the destroyed medial orbital wall was seen, without the particular signs of perineuritis on the left side (B). Profound enhancement is present in the right orbital apex (arrow in C), combined with an extensive dural enhancement at the anterior skull base, particularly on the right side (arrows in C and D). (D) A small degree of swelling of the right optic nerve is present.

Laboratory Testing

Laboratory testing showed growth of *Staphylococcus aureus* from the nasal cavity. Red and white cell counts were normal, as was C-reactive protein (CRP). A biopsy of the left orbital mass showed granulation tissue, consistent with chronic fibrosing inflammation (Figure 3).

Patient Management

A diagnosis of optic perineuritis was made and a 6-day course of intravenous antibiotic treatment was started (amoxicillin/clavulanate 1200 mg qds) followed by IVMP (1 g per day for 3 days) treatment. At last follow-up 10 weeks after onset, BCVA, MRI, and Goldmann visual fields remained unchanged. The sinusitis had improved and the aspect of the nasal mucosa had returned to normal. He was discharged with the advice to stop cocaine sniffing and of strict naso-oral hygiene with aid of regular nasal lavage.

FIGURE 3 Histology of the biopsy taken from the orbital infiltrate demonstrated inflammatory changes and granulation tissue (HE, $\times 100$).

DISCUSSION

This case report highlights the importance of recalling the association of optic neuritis with pathology originating in the sinuses. Since the original classification of Marcus Gunn in 1904,¹ most recent case reports were on cancer and sinus mucocoeles gaining access to the orbit. Potential erosion of the medial orbital wall creates an open entry zone from the paranasal sinus into the orbit. Indeed, several cases of optic neuropathies have been documented following intranasal cocaine abuse.

Our review of the literature revealed five cases with optic neuritis following cocaine consumption.^{4–7} Newman *et al.* were first to describe a case of bilateral optic neuropathy and osteolytic sinusitis in 1988 in a patient with intranasal cocaine abuse.⁴ One year later, Goldberg *et al.* presented three cases of orbitopathy or optic neuropathy associated with long-standing intranasal cocaine abuse and chronic sinusitis. In two of these cases, there was radiographic evidence for inflammation of the sinuses gaining access to the orbit. Patients were treated with steroids with moderate success.⁵ Two more cases were reported.^{6,7} The first was a 48-year-old woman with chronic intranasal cocaine consumption in whom an infiltrating mass from the maxillary sinus extended into the orbit, leading to proptosis and optic neuropathy. A tumour was suspected, but several tissue biopsies only revealed nonspecific chronic inflammation.⁶ The other case was a 29-year-old male with extensive bony destruction due to nasal cocaine consumption. He developed a subperiosteal abscess and optic neuritis that responded well to treatment with IVMP and antibiotics.⁷ Likewise, this combination therapy can lead to full recovery following an orbital apex syndrome related to intranasal cocaine consumption. As in our case, a tissue biopsy revealed nonspecific granulation without any evidence of granuloma formation, vasculitis or malignancy.⁸

The destructive effect of intranasal cocaine on nasal cartilage and bone structures is well recognised from the ear-nose-throat (ENT) literature. Alexandrakis *et al.* described seven cases in whom chronic intranasal cocaine consumption resulted in extensive bone destruction related sinusitis and orbital cellulitis. These patients partly responded to treatment with steroids and/or antibiotics and did not suffer from an optic neuropathy at the time.⁹ There is also evidence that intra-orbital inflammation following cocaine-induced chronic osteolytic sinusitis may spare the optic nerve and may either cause mechanical restriction of eye movements or possibly affect the third, fourth, and sixth cranial nerves leading to diplopia.¹⁰ Clinical evidence for proptosis in these patients should trigger orbital imaging. This case made a full recovery under combined treatment with antibiotics and IVMP.¹⁰

The present case is unique in that recurrent bilateral optic neuritis and optic perineuritis occurred over a period of 5 years. All episodes occurred in the context of sinus inflammation. It seems plausible to attribute this to the large bone defect as a direct result of a chronic osteolytic sinusitis after cocaine snorting. Recovery of visual acuity was poor after the last episode, characterised by a dense central scotoma. We suspect that this was caused by the perineuritis predominantly affecting those axons projecting from the macula through the outer inferonasal portion of the optic nerve. On the other hand, the first episode of loss of vision occurred on the right side. Retrospectively, there was already a small bone defect, which can easily be mistaken as an artefact. In addition, the profound dural enhancement in the anterior skull base, around the optic nerve in continuity with the orbital apex enhancement, suggests an alternative causes of inflammatory optic (peri-)neuritis.

Of note, the clinical differential diagnosis is broad. One should consider those inflammatory, neoplastic, infectious, and idiopathic disorders that may lead to bone destruction. There is a literature on granulomatosis with polyangiitis, sarcoidosis, relapsing polychondritis, systemic lupus erythematosus, mixed connective tissue disease, lymphomas, bacterial sinus infections, syphilis, and fungal infections (notably *Aspergillus* in diabetic patients).

In conclusion, the present case highlights the relationship between recurrent optic neuritis and optic perineuritis in the context of intranasal cocaine consumption causing an open connection between the orbit and nasal cavities. The exact pathological mechanism remains uncertain, as both intra-orbital spread of inflammation and cocaine-related ischaemia may damage the optic nerve. Combination treatment with antibiotics and IVMP should be initiated as early as possible. Rigorous nasal sinus hygiene is advised in patients with an open orbital wall.

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figure 3 of this article is available in colour online at www.informahealthcare.com/oph

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