Title: Recurrent optic perineuritis after intranasal cocaine abuse

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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. MRI demonstrated optic perineuritis adjacent to a soft tissue mass which intruded the orbit from the nasal cavity. 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: optic perineuritis, cocaine, osteolytic sinusitis, optic neuritis, MRI, CT, histology.

Introduction

Marcus Gunn can be credited with the first systematic classification of optic neuritis in the post-ophthalmoscope area: (i) due to infection (orbit, paranasal sinuses, meninges), (ii) infectious disease (e.g. Syphilis), (iii) multiple sclerosis.[1] At the time, syphilis was a frequent differential diagnosis and among others Buzzard noted that compared to multiple sclerosis, visual loss was more severe, recovered poorly and was associated with paler discs.[2] The osetolytical changes caused by syphilis in the oro-nasal cavities and the anatomical proximity of the sinuses with the optic nerve made a casual relationship likely. At the time there was good autopsy data to support the argument, but case numbers decreased over the years, probably most reliable reflected by a single centre experience (John 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 two 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 methylprednisolon (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 one 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 (RAPD). The BCVA was 1.0 on the right and 0.02 on the left. There was no papil edema, but the left optic disc was pale compared to 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 (Fig.1A). The soft tissue was in direct contact with the left optic nerve and showed a perineural enhancement suspicious of optic perineuritis (Fig. 1B, 1C). MRI showed extensive (reactive) dural enhancement at the anterior skull-base. Computed tomography (CT) showed an extensive destruction of the left medial orbital wall (Fig.1D). There was also some anterior bony destruction on the right.

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

Laboratory testing showed growth of *Staphylococcus Aureus* from the nasal cavity. Red and white cell counts were normal as was the *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 (amoxicilline/clavulanate 1200 mg qds) followed by IVMP (1g per day for three 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.

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 mucoceles gaining access to the orbit. Potentially erosion of the medial orbital wall is 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.[4,5,6,7] Newman was first to describe a case of bilateral optic neuropathy and osteolytic sinusitis in 1989 in a patient with intranasal cocaine abuse.[4] One year later, Goldberg 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 tumor was suspected, but several tissue biopsies only revealed non-specific 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 which 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 non specific 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 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 3rd, 4th and 6th 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 five 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 artifact. In addtion, he 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 which may lead to bone destruction. There is a literature on Wegener granulomatosis, 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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Captions for Figures

Figure 1: Axial (A) and Coronal (B and C) contrast enhanced T1-weighted MR images with fat-suppression shows 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), with a There is ring of contrast enhancement surrounding the left optic nerve borders without contrast enhancement of the optic nerve itself, 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 (A) the Coronal CT 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 Fig.1D). Contrast enhancement of a comparable mass-lesion invading the orbit through the destructed medial orbital wall was seen, without the particular signs of peri-neuritis on the left side (B). Profound enhancement is present in the right orbital apex (C), combined with an extensive dural enhancement at the anterior skull-base, particularly on the right side (arrow). (D) A small degree of swelling of the right optic nerve is present.

Figure 3: Histology of the biopsy taken from the orbital infiltrate demonstrated inflammatory changes and granulation tissue (HE x100).