Spontaneous Resolution of Optic Perineuritis in a Patient with Crohn’s Disease

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Optic perineuritis · Crohn’s disease · Spontaneous resolution · Untreated optic perineuritis

Abstract
Optic perineuritis (OPN) is a rare condition characterized by inflammation of the optic nerve sheath. The underlying etiologies of OPN are varied and include systemic inflammatory conditions such as Crohn’s disease. It has been asserted that OPN requires prompt treatment in all cases. However, we report a case of OPN in a patient with Crohn’s disease that spontaneously resolved without treatment, underscoring that observation is a reasonable approach in some cases of OPN.

Introduction
Optic perineuritis (OPN) is a rare orbital inflammatory disease affecting the optic nerve sheath. The condition is heterogeneous in its etiology and presentation. Patients with OPN commonly present with ocular pain or discomfort exacerbated by movement, accompanied by variable reduction in visual acuity and peripheral vision defects [1, 2].

While many cases of OPN are idiopathic (primary OPN), most are secondary to an identifiable systemic disease [3], for which OPN is sometimes the initial presenting feature. Systemic associations for secondary OPN include inflammatory conditions such as Crohn’s disease [4], sarcoidosis [3, 5, 6], Behçet disease [7], granulomatosis with polyangiitis [8], giant
Definitive diagnosis of OPN is made radiographically by post-contrast fat-suppressed T1-weighted magnetic resonance imaging (MRI) images that show isolated or predominant enhancement of the optic nerve sheath [2, 15]. It is important to differentiate OPN from optic neuritis as their investigation and treatment differ. Whereas optic neuritis is often self-limiting in typical cases, there are limited data on the clinical course of untreated OPN [1]. Poor visual outcomes have been reported in cases of OPN where steroid treatment was delayed [2, 16].

It has been asserted that every case of OPN requires prompt treatment with immunosuppressive or anti-inflammatory medications [17]. However, this assertion is challenged by cases in which OPN has self-resolved. Here, we report a case of OPN in a patient with Crohn’s disease that spontaneously resolved without treatment, indicating that observation may be a reasonable strategy in some patients.

Case Presentation

A 28-year-old woman was referred to ophthalmology for a 1-week history of right eye pain with eye movement without vision loss. She had an 8-year history of Crohn’s disease treated with azathioprine. Ophthalmological exam revealed a visual acuity of 20/20 in both eyes, Humphrey 24-2 SITA-Fast visual field testing was normal, and there was no relative afferent pupillary defect. Color vision as measured by Ishihara color plates was 10/14 OD and 14/14 OS. Dilated fundus examination showed mild optic disk edema in the right eye and a normal left optic nerve (Fig. 1). Optical coherence tomography of the retinal nerve fiber layer showed an average thickness of 296 μm OD and 87 μm OS. Optical coherence tomography of the macula was normal in both eyes. The patient was suspected of having OPN given the prominent pain and preserved visual function, and MRI of the orbits and brain was performed. This showed enhancement of the right optic nerve sheath, consistent with the diagnosis of OPN, which may have been related to her underlying diagnosis of Crohn’s disease (Fig. 2). A workup was initiated that included a normal complete blood count and normal or negative erythrocyte sedimentation rate, C-reactive protein, antinuclear antibody, antineutrophil cytoplasmic antibodies, angiotensin-converting enzyme, VDRL, Bartonella serology, Lyme serology,
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aquaporin-4 antibodies, and myelin oligodendrocyte glycoprotein antibodies. Chest X-ray was also normal. The treatment options for OPN were discussed with the patient, including observation or treatment with corticosteroids. Since the pain was manageable and her visual function was preserved, she opted to observe. The patient reported that her eye pain resolved after 2 weeks. A follow-up appointment 1 month later showed normal visual function with 20/20 visual acuity and normal 24-2 SITA-Fast visual fields. There was only minimal optic disk edema at this time. At the 3-month follow-up, visual function remained normal, and she had no optic disk edema.

Discussion

OPN is a heterogeneous condition causing inflammation of the optic nerve sheath for which standard treatment is anti-inflammatory or immunosuppressive therapy, usually high-dose corticosteroids. The overwhelming majority of OPN cases have been treated with steroids [1, 3, 15] or, somewhat less commonly, with NSAIDS alone [2]. It has been suggested that prompt treatment of OPN is essential to avoid poor visual outcomes [2, 16, 17]. Our case challenges this, demonstrating that OPN can resolve spontaneously and completely without treatment, and adds additional evidence that OPN may be associated with Crohn’s disease.

Our literature review revealed 6 cases of OPN that spontaneously resolved. In 1 case, resolution of OPN was confirmed by imaging in addition to clinical resolution [18]. Tung et al. [18] described a 60-year-old woman who presented with visual loss, severe retro-orbital pain bilaterally, and headache worsened by lying down. Her eye pain resolved spontaneously at 6 weeks, and follow-up MRI confirmed resolution of perineural enhancement. Wals et al. [19] described a 52-year-old woman with combined idiopathic neuroretinitis and OPN whose visual function and fundus exam normalized at 11 months without intervention. Bergman et al. [20] described 2 cases of untreated OPN. The first was a 58-year-old man who initially presented with dimmed vision, headache, and retro-orbital swelling. He received an MRI

![Fig. 2. a MRI of the orbit T1 post-contrast with fat suppression demonstrating mild right optic nerve sheath enhancement (red arrow). b MRI orbit T2 shows loss of the cerebrospinal fluid signal around the right optic nerve (yellow arrow) without increased signal in the optic nerve itself.](image-url)
1 week later that confirmed slight perineural edema, but vision had normalized; therefore, no treatment was initiated. The second case was a 60-year-old man who presented with eye pain and slight proptosis. He received only lubricating eye drops, and pain subsided at 2-month follow-up. Finally, in a recent Canadian case series, 2 patients were identified as untreated, 1 with idiopathic OPN and normal visual function, and the other with OPN secondary to myelin oligodendrocyte glycoprotein antibody disease and a 2-year history of visual symptoms [3]. No further details of their workup and clinical course were provided. We were unable to retrieve any cases of OPN attributable to Crohn’s disease that spontaneously resolved without treatment.

The diagnosis of OPN is this young patient may be related to her underlying Crohn’s disease. Crohn’s disease and ulcerative colitis are the two primary causes of inflammatory bowel disease (IBD) and are both considered systemic inflammatory conditions, with extraintestinal manifestations (EIMs) most commonly affecting the joints, eyes, skin, and hepatobiliary system [21]. Female sex and age under 40 are risk factors for the development of EIM in IBD [22]. Inflammatory ophthalmic manifestations are estimated to occur in 3.5–6.8% of patients with Crohn’s disease [22], with uveitis and episcleritis being most frequent [23]. Previous cases of optic neuritis in IBD have been reported [4, 24, 25], as has one previous case of OPN in a patient with Crohn’s disease [4]. While the pathogenesis of EIM in IBD is incompletely understood, proposed mechanisms include systemic upregulation of proinflammatory cytokines and cross-reactivity of antigens [26].

In conclusion, some authors have written that OPN requires prompt treatment in all cases. Our case reveals that some patients may have spontaneous improvement without treatment. Previous cases of OPN in the literature that similarly resolved without treatment support that this was not a chance event. Treatment should not be delayed where a clear treatable cause of OPN is identified (eg. infection such as syphilis) or when pain is severe. However, in patients with relatively preserved visual function and manageable pain, observation is a reasonable initial strategy. Patients should have close follow-up to ensure that there is no concerning worsening. Our case also adds additional evidence that OPN may be associated with Crohn’s disease. A review of systems regarding symptoms of IBD should be considered in a new case of OPN.

**Statement of Ethics**

Written informed consent was obtained from the patient for publication of the details of their medical case and accompanying images. Research Ethics Approval was not required for this paper as per University of Toronto Research Ethics Board. The study complies with the guidelines for human studies and was conducted in accordance with the World Health Organization Declaration of Helsinki.

**Conflict of Interest Statement**

The authors have no conflicts of interest to declare.

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Author Contributions

Lauren Pickel contributed to conception and design, preparation of the manuscript, and final approval of the manuscript. Jonathan A. Micieli contributed to conception and design, data collection, critical revisions, and final approval of the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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