AMERICAN JOURNAL OF DIAGNOSTIC IMAGING
2024; 11(1): 9-13.
10.5455/ajdi.20240930083147

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CASE REPORT

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Bilateral optic perineuritis as the initial presentation of giant cell arteritis

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Abstract

Giant cell arteritis (GCA), also known as Horton's disease, is a granulomatous vasculitis affecting large and medium-sized arteries, frequently leading to visual complications due to ophthalmic artery involvement. We present a case of a 72-year-old woman with bilateral optic perineuritis, marking the first manifestation of GCA. The patient experienced severe headaches and progressive vision loss, with brain Magnetic Resonance Imaging (MRI) revealing thickening of the temporal artery walls and optic nerve sheath enhancement. Optic perineuritis, while rare, can be an initial sign of GCA, underscoring the importance of early recognition and treatment to prevent irreversible visual impairment.

ARTICLE HISTORY

Received September 30, 2024
Accepted October 23, 2024
Published November 04, 2024

KEYWORDS

Vasculitis, Giant cell Arteritis, Horton, Perineuritis

INTRODUCTION

Giant cell arteritis (GCA), also known as Horton's disease, is the most prevalent primary systemic vasculitis in adults, typically affecting individuals over the age of 50. Despite its frequency in this age group, GCA remains a rare condition overall, with an incidence ranging from 5 to 7.5 cases per 100,000 people [1]. Ocular involvement occurs in 25–50% of cases, primarily due to inflammation obstructing the ophthalmic arteries or the short posterior ciliary arteries, which supply blood to the optic nerve and optic disc. The most frequent complication is arteritic anterior ischemic optic neuropathy (AAION), caused by ischemia of the optic disc resulting from vasculitis affecting the short posterior ciliary artery. Arteritic posterior ischemic optic neuropathy, which involves infarction of the intraorbital segment of the optic nerve and manifests as retrobulbar optic neuropathy, is another potential outcome. Less commonly, central retinal artery or cilioretinal artery occlusion may occur.

However, visual impairment due to optic perineuritis is extremely rare in the context of GCA, with only a few reported cases in the literature [1–5]. Studies suggest that the incidence of optic perineuritis as a presenting symptom in GCA is significantly lower compared to other ocular manifestations, such as AAION, which accounts for a substantial proportion of visual complications [6]. This rarity emphasizes the importance of recognizing atypical symptoms like perineuritis, as timely diagnosis and treatment can prevent irreversible visual loss.

CASE REPORT

We report the case of a 72-year-old woman, with a history of diabetes managed with oral hypoglycemic agents, who presented with bilateral optic perineuritis as the initial sign of GCA. The patient experienced severe headaches for 10 days that were resistant to standard medical therapy, followed by a progressive decline in visual acuity in both eyes.

Upon ophthalmological examination, the patient's corrected visual acuity was 2/10 in both eyes, and a relative afferent pupillary defect was observed bilaterally. Fundoscopy revealed papilledema in both eyes. Temporal artery pulses were absent, while the rest of the physical examination was unremarkable.

Laboratory tests showed elevated C-reactive protein (CRP) levels at 20 mg/L (normal < 3 mg/L) and an erythrocyte sedimentation rate (ESR) of 88 mm in the first hour (normal < 10 mm) and 116 mm in the second hour (normal < 20 mm). Serological tests for syphilis were negative, indicating that this infectious etiology was unlikely. The rheumatoid factor was also

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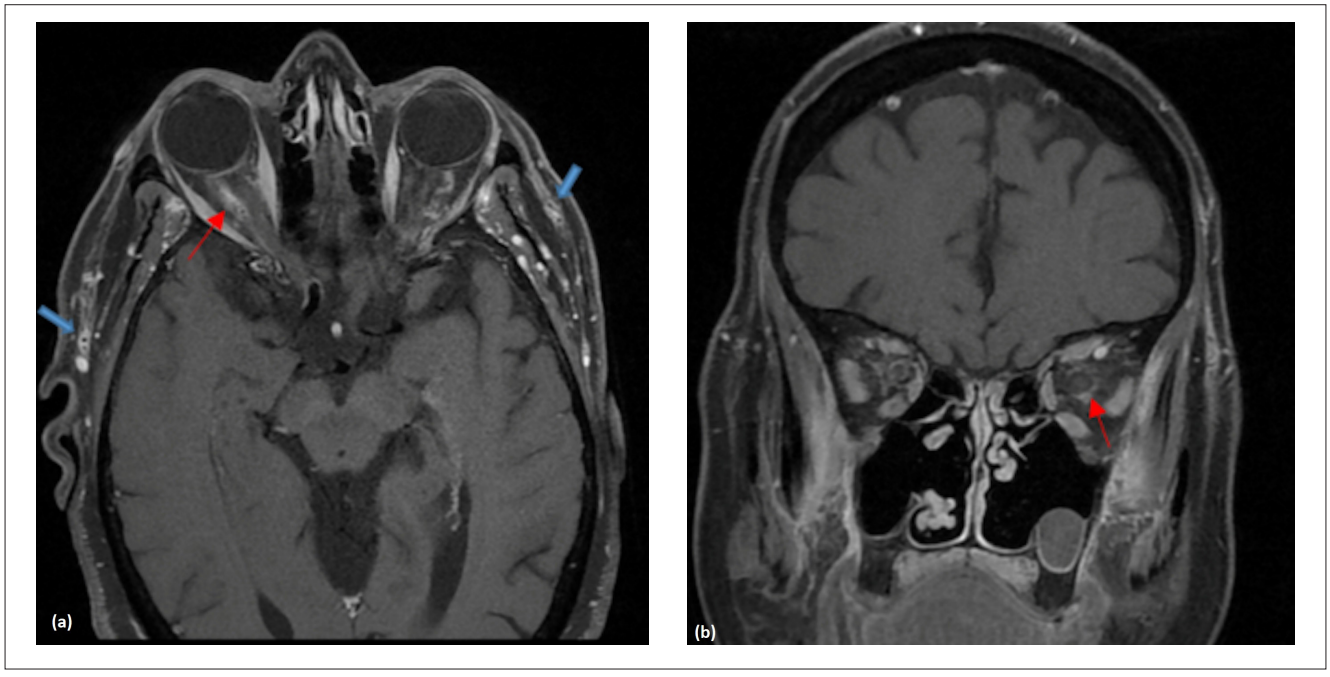


Figure 1. Brain MRI with T1-weighted Dixon sequences after gadolinium injection, in axial (a) and coronal (b) slices, shows bilateral perineural enhancement of the optic nerve sheaths (red arrows). Note the contrast enhancement of the superficial temporal artery walls bilaterally (blue arrows).

within normal limits, further supporting the exclusion of rheumatoid arthritis as a contributing factor. Additionally, Anti-Neutrophil Cytoplasmic Antibodies (ANCA) testing was negative, which helped rule out vasculitis associated with autoimmune conditions. Visual field testing showed enlargement of the blind spots and bilateral central scotomas.

Brain Magnetic Resonance Imaging (MRI) revealed thickening and intense contrast enhancement of the walls of the temporal arteries, along with bilateral enhancement of the optic nerve sheaths. Temporal artery ultrasound confirmed the presence of a hypoechoic halo and reduced lumen patency, though blood flow velocities were preserved (Figure 1 and Figure 2).

Given the clinical presentation, laboratory findings, temporal artery ultrasound, and MRI results, a diagnosis of bilateral optic perineuritis secondary to GCA was established. The patient was treated with intravenous methylprednisolone for five days, followed by a gradual tapering of oral prednisone, resulting in significant clinical improvement.

DISCUSSION

Optic perineuritis is an inflammatory orbital disease affecting the sheath of the optic nerve within the orbit. It can arise idiopathically or as a secondary effect of systemic inflammatory disorders, including primary vasculitis, Behçet's disease, syphilis, sarcoidosis, Crohn's disease, IgG4-related disease, and idiopathic orbital inflammatory syndrome (IOIS) [1]. Additionally, myelin oligodendrocyte glycoprotein (MOG)-associated disorders (MOGAD) are increasingly recognized as significant causes of optic perineuritis, particularly in younger patients. MOGAD has emerged as a distinct category of demyelinating diseases, often presenting with optic nerve involvement and necessitating careful diagnostic consideration [2]. In our case, a thorough diagnostic workup was performed to exclude these conditions, which included imaging studies and serological tests. The presence of classic clinical features and imaging findings consistent with GCA helped to confirm the diagnosis of optic perineuritis secondary to this systemic condition.

The most prevalent pathological feature is lymphocytic or granulomatous infiltration of the optic nerve sheath. In rare instances, inflammatory involvement around the blood vessels in the peripheral region of the optic nerve may also occur. This can lead to extrinsic compression or localized infarcts resulting in perineural effects [3]. The average age of onset is 40 years, with a female predom-

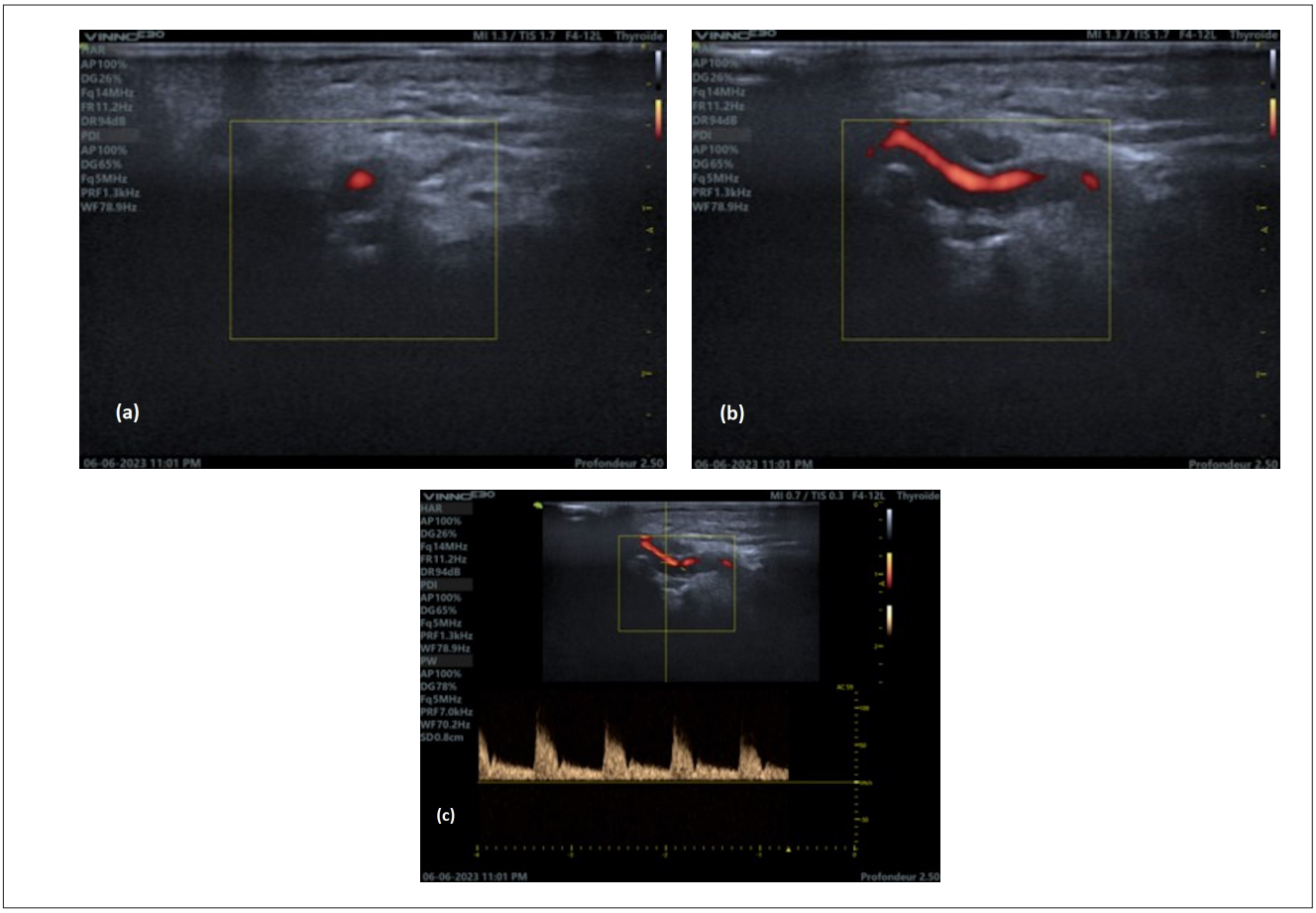


Figure 2. Doppler ultrasound of the temporal arteries in axial (a) and longitudinal (b) views shows significant hypoechoic wall thickening of the proximal portions of both the right and left temporal arteries, measuring up to 5 mm in maximum thickness. This thickening significantly reduces their lumens, creating a characteristic bilateral halo appearance while maintaining patency and preserved blood flow velocity (c).

inance. Patients often report unilateral or bilateral ocular pain that may intensify with eye movements. Alterations in the visual field are the most frequently noted symptom, potentially presenting as peripheral, paracentral, or cecocentral scotomas. Visual acuity is affected in approximately half of the cases, particularly if the macula is involved. Diplopia, exophthalmos, and oculomotor nerve palsies may occur but are infrequent [1–3].

The preferred diagnostic approach is MRI, utilizing gadolinium injection and fat suppression techniques. This imaging typically reveals circumferential enhancement of the intraorbital portion of the optic nerve sheath [3]. Less frequently, enhancement may also be observed at the periphery of the optic nerve, in the surrounding orbital fat, or in the extraocular muscles [3]. The challenges in diagnosing optic perineuritis in the context of GCA arise from the overlap of symptoms with other conditions, particularly IOIS and optic neuritis. Recent literature emphasizes the need for careful clinical assessment and a high index of suspicion, especially when atypical symptoms are present [3].

Optic neuritis is the primary differential diagnosis; however, there are significant differences in initial clinical presentation, underlying causes, management, and outcomes between the two conditions [4]. Optic neuritis generally affects younger individuals (ages 25–35) and is closely linked to the formation of demyelinating lesions in the central nervous system. Conversely, optic perineuritis typically presents at an average age of 41 years without a heightened risk of developing demyelinating diseases. Visual field loss in optic neuritis is primarily central, leading to more frequent impairment of visual acuity compared to optic perineuritis. While visual loss in optic neuritis often resolves without corticosteroid therapy, it may continue to progress

in optic perineuritis without appropriate treatment. Consequently, timely medical intervention is vital for optic perineuritis, whereas, it does not enhance visual outcomes in optic neuritis [3–4].

Moreover, patients with optic perineuritis frequently experience relapses when treatment is ceased, contrasting with classic optic neuritis, where episodes are usually self-limiting. The presence of extraocular symptoms should raise suspicion for optic perineuritis rather than optic neuritis [3].

To rule out secondary causes, laboratory tests should include syphilis serology, antinuclear antibodies, rheumatoid factor, ANCA, and serum IgG4 levels. The ESR and CRP are less informative in differentiating between primary and secondary causes, as they can be elevated in both conditions. A lumbar puncture is also advised to exclude possible underlying infections or neoplastic processes. Biopsy of the optic nerve sheath is recommended only in cases demonstrating clinical progression despite adequate treatment [5].

MRI of the brain typically shows contrast enhancement around the optic nerve sheath in its intraorbital segment, with or without the involvement of other orbital structures [6]. The role of color Doppler ultrasound of the temporal artery is significant in diagnosing GCA and may serve as a substitute for temporal artery biopsy. This non-invasive test can detect vascular wall edema, which appears as a halo when performed by skilled practitioners [7] (Figure 2).

Initial treatment consists of intravenous infusions of methylprednisolone, followed by a gradual tapering of oral corticosteroids to reduce the risk of relapse. The choice of intravenous methylprednisolone over immediate oral corticosteroids is supported by current guidelines, which recommend intravenous administration for severe or progressive cases of optic perineuritis to achieve faster therapeutic levels and better control of inflammation [8, 9]. Other immunomodulatory medications, such as azathioprine, may also be utilized for maintenance therapy. Relapses are common and generally respond to additional corticosteroid courses [8, 9].

Long-term management of patients with optic perineuritis requires careful follow-up to monitor for potential relapses and assess visual outcomes. Regular ophthalmological evaluations, including visual field testing, are recommended. In cases of recurrent episodes, alternative treatments such as mycophenolate mofetil or rituximab may be considered, depending on the patient's response to corticosteroid therapy and the severity of symptoms [10].

The visual prognosis for optic perineuritis is generally favorable, with notable improvements in vision and pain reduction when treatment is initiated promptly. Factors that can negatively affect prognosis include delays in treatment and the presence of pre-existing ocular conditions [4–6].

CONCLUSION

In conclusion, optic perineuritis, while rare, is a critical initial manifestation of GCA that requires special attention. Early recognition is vital, as it enables prompt therapeutic interventions, reducing the risk of irreversible visual complications. Clinicians must maintain a high index of suspicion, particularly in older patients with atypical visual symptoms.

A thorough evaluation, including imaging and laboratory tests, is essential to exclude other potential causes, such as IOIS. Current treatment guidelines recommend intravenous methylprednisolone, emphasizing a proactive approach tailored to disease severity.

Regular long-term follow-up is crucial to monitor for relapses and assess visual outcomes. Therefore, incorporating optic perineuritis into the diagnostic framework for GCA is essential for optimal patient management and preserving visual quality of life.

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