



CASE REPORT

Optic Perineuritis Secondary to Sjogreen's Syndrome: Case Report

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Keywords

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Optic perineuritis (OPN) is a rare inflammatory disease involving the optic nerve sheath [1], causing pain and disc edema and is often bilateral [2]. The diagnosis of OPN is commonly based on magnetic resonance imaging (MRI) findings and clinical features [3]. In MRI it is possible to visualize a circumferential enhancement around the intraorbital optic nerve with preservation of the nerve itself (doughnut sign) [4]. This is better seen in contrast-enhanced and suppressed coronal MRI sequences with fat. Magnetic resonance imaging may also show enhancement of orbital fat and slight increase in extraocular muscles [1]. The etiologies of OPN include syphilis [5,6], tuberculosis [7,8], herpes zoster [9], sarcoidosis [10,11], Wegener's granulomatosis [12], granulomatosis associated with polyangiitis [13], giant cell arteritis [14,15] Crohn's disease [16], Behçet's disease [1], acute retinal necrosis [17]. However, in most cases, no specific etiology is identified and these cases are categorized as idiopathic OPN [18].

A 68-year-old woman started with a subacute onset of progressive visual acuity worsening associated with bilateral retrobulbar pain about 4 months ago. Fundoscopy was normal and reduced direct and consensual photo motor reflex. Sicca syndrome was confirmed in Schirmer test. Rheumatoid factor was

positive (23.6) and anti-RO/LA antibody positive. Cranial MRI showed very thin and intense contrast enhancement along both optic nerve sheath and parotid edema associated (Figure 1) meeting the criteria of Sjogren syndrome (SS) [19]. We found no evidence of secondary SS suggesting a form primary.

Patient had clinical improvement of visual acuity and retroocular pain partially after pulse therapy with methylprednisolone. Rituximab maintenance patient every 6 months. SS is a rare cause and should not be forgotten as a differential diagnosis of optic perineuritis.

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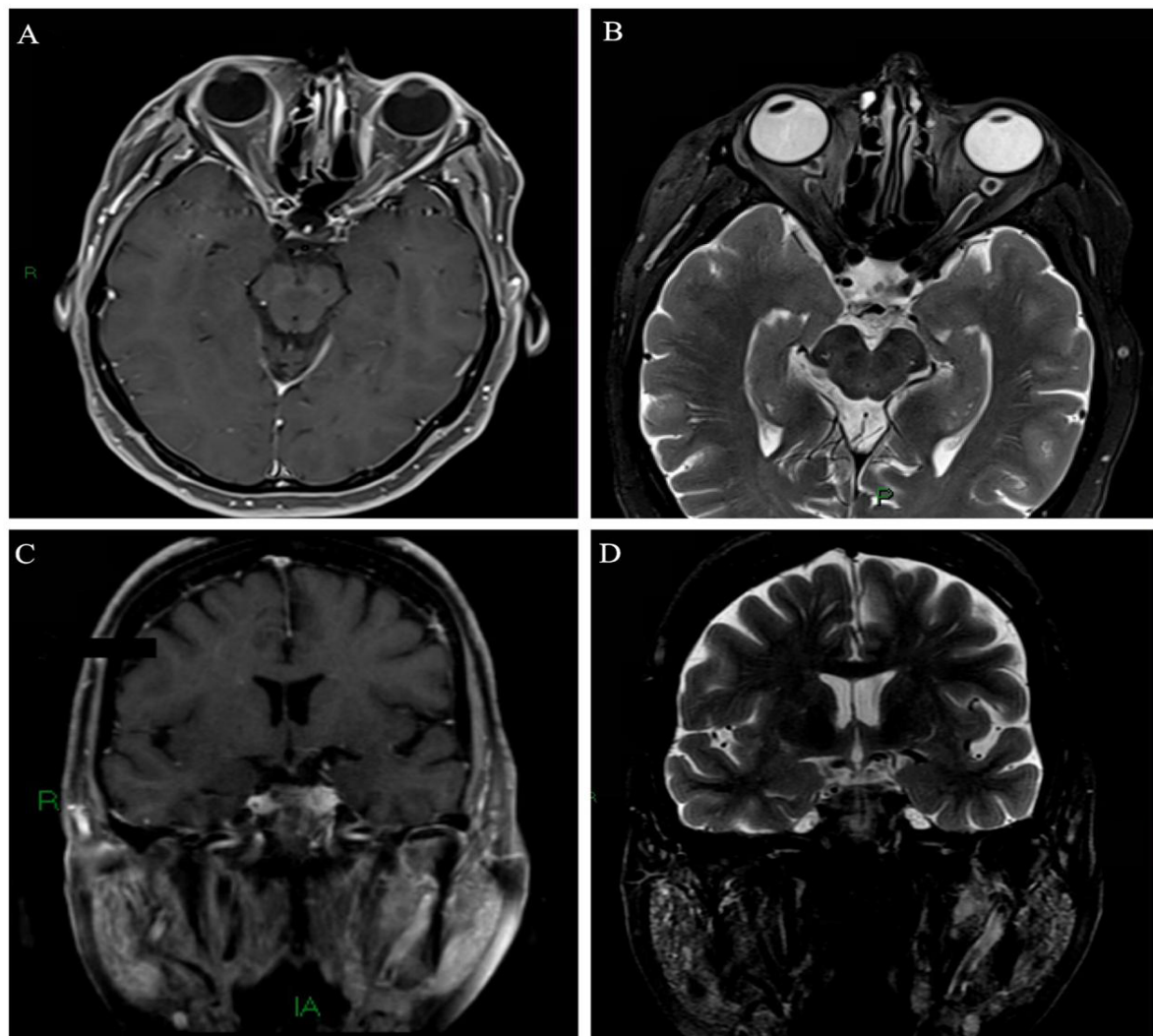


Figure 1: Axial A) T1-spin echo, after gadolinium iv injection and axial; B) T2 with fat saturation show a very thin and intense symmetrically contrast enhancement along both optic nerve sheath as well a high intensity thickening along of both optic perineural CSF space. Coronal C) T1-spin echo, after gadolinium i.v. injection and coronal; D) T2 with fat saturation reveal a symmetrical increase of parotidite glands.

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