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Efficacy of Rituximab Therapy for Oculomotor Nerve Palsy and Vision Loss Due to Granulomatous Polyangiitis

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Abstract

Completely reversible oculomotor palsy VI and the vision restoration after only rituximab-corticosteroid therapy in granulomatosis polyangiitis with intraorbital mass presented with CT documentation. Short review of literature about treatment options follows.

Keywords: Granulomatous polyangiitis; Oculomotor Sixth Nerve Palsy; Vision Loss; Rituximab; Corticosteroids

Introduction

Sixth nerve palsy is a disorder that affects eye movement. That is due to damage of the sixth cranial nerve (n. abducens). The primary function of the sixth cranial nerve is to send signals to lateral rectus muscle. This small muscle is located on the outer side in our eye. It is responsible for turning your eye away from our nose. When the lateral rectus muscle weakness, your eye crosses inward toward your nose. If inferior oblique ocular muscle (n. III) localized closely to lateral rectus is affected, the eye deviated down inward.

A 67-year-old man with overweight, diabetes mellitus type II, arterial hypertension, congestive heart failure and paroxismal atrial fibrillation developed left ocular nerve pulsy with inward deviation of ocular nucleus and vision loss. Last three years he has treated with corticosteroid pulses and rituximab (RTX) (Anti-CD20) due to granulomatous polyangiitis (GPA) and untill December 2020 was in remission. Because corona pandemic last course postponed and half year after missed RTX for the first time he developed left retro orbital mass with internal inferior rotation of his eye with vision loss (Figure 1). IV Methylprednisolone 1g/day pulse therapy N3 with further IV 100mg/day was reinstituted and he got infusions of RTX 500mg/week N4. Oral prednisone continued with tapering to 20mg/day. Three moths later full resolution of ocular palsy followed (Figure 2) and his vision restored. One can see parallel restoration of anterior posterior axis (white lines) of right and left eyes.

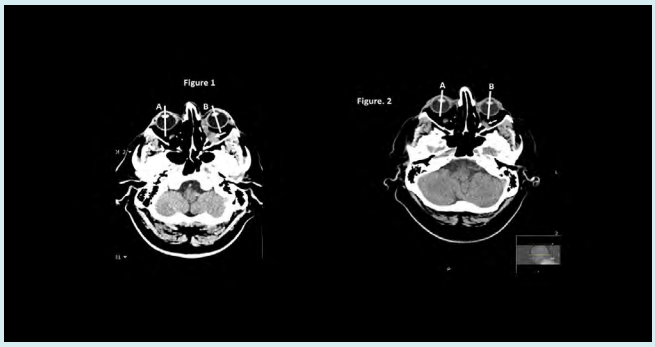

Figure 1: Computer tomography of a head shows left posterior intraorbital mass (GPA infiltrate) (white arrow), obstructing entrance of ocular nerve with internal deviation of left ocular nucleus (white A-B lines).

Figure 2: Three months later after rituximab and corticosteroid therapy an angle between axial ocular lines of both eyes vanished and significant reduction of the inflammatory mass (arrow) followed.

Completely reversible oculomotor palsy VI and the vision restoration after only rituximab-corticosteroid therapy in GPA with intraorbital mass have not described. All patients also had immunosuppressive and cytotoxic treatment along with RTX [1-5]. Fifting years ago a case of abducens nerve palsy resulting from GPA pachymeningitis reported, with response to thirty mg per day of prednison only [6]. Recently, a case with headache and new left-sided hearing loss as relapse of GPA after failure of CS+IV endoxan (CY) reported. The patient got GC and RTX, and a 1-year remission followed [7]. The molecular mechanism of RTX is not fully understood. In this case, RTX was more effective with rapid and strong suppressing B cells than CY. Since the B cells count was proportional to the patients clinical manifestations, B cells represent a suitable target for the treatment of GPA with cranial neuropathies.

Anti-CD20 therapy used successfully in refractory granulomatous disease affecting the CNS: RTX is currently the drug most frequently used for remission induction in refractory orbital granuloma. Whereas an initial series on five patients reported inefficacy of rituximab, further case series composed of five to seven patients reported remission rates of 80–100% [8-9]. Anti-TNF therapy (infliximab) may be option in refractory GPA [10].

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