Cytomegalovirus retinitis in Good Syndrome

Retinite por citomegalovírus em paciente com Síndrome de Good

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ABSTRACT

Good syndrome is a paraneoplastic syndrome characterized by the association of thymoma and hypogammaglobulinemia, with immunosuppression. We report a rare case of cytomegalovirus retinitis in a patient with this syndrome.

Keywords: Cytomegalovirus Retinitis; Retinitis; Cytomegalovirus; Thymoma; Agammaglobulinemia; Case reports

RESUMO

A Síndrome de Good é uma síndrome paraneoplásica caracterizada pela associação de timoma e hipogamaglobulinemia, cursando com imunossupressão. Relatamos um caso raro de retinite por citomegalovírus em paciente com esta síndrome.

Descritores: Retinite por citomegalovírus; Retinite; Citomegalovírus; Timoma; Agamaglobulinemia; Relatos de casos

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**INTRODUCTION**

Thymoma is a rare malignant neoplasm accounting for 20 to 30% of mediastinal tumors in adults. Most have slow and sluggish growth. This tumor may present respiratory symptoms due to compression of the upper airways and superior vena cava syndrome. It may also cause paraneoplastic syndromes such as myasthenia gravis, red cell aplasia, or hypogammaglobulinemia.

Good syndrome was first described by the American oncologist Dr. Robert Good in 1956. He observed the association between thymoma and hypogammaglobulinemia leading to immunosuppression in these patients. The syndrome typically occurs in middle-aged adults, and has frequent recurrent pneumonia and sinusitis, cytomegalovirus infections (especially retinitis), and fungal infections.

The occurrence of cytomegalovirus retinitis in patients with Good syndrome is rare, with only nine cases described in the literature to date. We report here the case of a patient with cytomegalovirus retinitis and Good syndrome.

**CASE REPORT**

C.E.P., 49 years old, female, with complaint of low visual acuity in the left eye with two weeks of evolution. Thymoma diagnosed 6 months before, having already undergone thymectomy and 6 cycles of chemotherapy (Pemetrexede). He presented two previous episodes of pneumonia and one of sinusitis in this period. The exam showed right eye with visual acuity 20/20 and no changes. Left eye with vision 20/30, intraocular pressure 32mmHg, anterior chamber reaction 3+ and vitreitis 2+. Eye fundoscopy presented extensive area of peripheral retinitis with vasculitis and superior justadiscal necrotic lesion (Figure 1).

The exams showed: normal hemogram with absence of leucopenia; VDRL and FTA-ABS negative; toxoplasmosis IgM positive (0.93 Index) and IgG positive; Herpes simplex and zoster IgM negative and IgG positive; Cytomegalovirus IgM negative and IgG positive; HIV negative.

The diagnostic hypothesis was acute herpetic retinal necrosis, and the patient was treated with intravenous acyclovir (10mg/kg every 8 hours) for 14 days associated with systemic corticosteroid (1mg/kg), topical corticosteroids and topical medications to control the intraocular pressure (timolol maleate 0.5% and brimonidine tartrate 0.2%). After discharge, she presented a discrete improvement of the retinal lesions (Figure 2), and was orally medicated with valaciclovir (3g/day) and prednisone (0.5mg/kg/day).

In the ambulatory follow-up two weeks after the beginning of the oral medication, the clinical condition of the left eye progressed, with the onset of a new lesion (Figure 3).

Valaciclovir was then suspended, and intravenous ganciclovir started (5mg/kg every 12 hours). This dose was maintained for 21 days, and the patient presented significant improvement of the condition, with retinal lesions practically cured (Figure 4).

Ganciclovir was used in maintenance dose (5mg/kg/day) for 18 weeks. When the medication was discontinued, the patient presented visual acuity 20/70, residual vitreitis 2+, and healed lesions (Figure 5).

Before the diagnosis of cytomegalovirus retinitis, the patient was investigated for Good Syndrome: immunoglobulin G (IgG) decreased 676mg/dL (reference values 800-1600mg/dL) and immunoglobulins A and M (IgA and IgM) at the lower limits. The CD4 count was also low, 348/μL (reference values 430-1010 / μL), consistent with the syndrome.

**DISCUSSION**

We report a case of cytomegalovirus retinitis in a patient with Good Syndrome. This is a rare case with only nine descriptions in the literature. Downes et al. recently reviewed these published cases and found the following characteristics: average age of presentation of 56 years (48-68 years); higher prevalence in women (55.5%); unilateral condition (88.9%); anterior chamber inflammation (62.5%); vitreitis present in 88.8% of cases and described as moderate to severe in 55.5% of the cases; diagnosis of cytomegalovirus retinitis after diagnosis of thymoma (88.8%); opportunistic respiratory infections (77.8%).

All these characteristics were also present in our case. Some particularities, however, were observed. Retinal involvement is more frequent in zones I and II, with only 2 cases with involvement of the periphery of the retina (zone III) described in the literature, which occurred in our case. Another difference observed was related to the visual acuity of presentation of the retinitis condition. The initial visual acuity of our patient was 20/30, the best found among the cases described. Downes et al. had a vision of 20/40 at baseline, but the other seven patients had a vision of less than or equal to 20/100 at diagnosis. The best visual acuity at presentation, however, did not provide our patient with a better final vision when compared to the other cases. Of these, 55.5% had vision between 20/40 and 20/200 at the final condition, where our patient also fits with final vision of 20/70.

In the vast majority of reports, diagnostic confirmation was made through aqueous humor PCR, which was not performed in our case. At the onset of our patient’s condition, extensive retinal necrosis and major inflammatory reaction, both anterior and posterior, led to the diagnosis of herpetic retinitis, which may also occur in Good Syndrome. Insufficient response to intravenous acyclovir and progression of the disease with the use of oral valaciclovir led us to the diagnosis of cytomegalovirus. Aqueous humoral PCR could have helped in this differential diagnosis. However, after the evolution of the case, we believe that there is no doubt about the diagnosis of our patient facing the expressive response of the retinal lesion to ganciclovir and all the other commemorations of the immune response. Surgery, 1956; 40(6):1010–7.

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It is also important to observe the peculiarities of a cytomegalovirus retinitis in an HIV negative patient. Several authors report more exuberant vitreitis in this situation, with conditions that even remember an acute retinal necrosis, just like ours. Good syndrome is a cause of cytomegalovirus retinitis in HIV-negative patients, and should always be investigated in this situation. Retinitis usually appears after the diagnosis of the syndrome, but there are reports in which the systemic condition was opened with retinitis.

**REFERENCES**


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