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POSTER PRESENTATION

A Case of Giant Cell Arteritis with Anterior Ischemic Optic Neuropathy Resistant to Corticosteroid Therapy

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Giant cell arteritis (GCA) is the most common primary systemic vasculitis in adults 50 years or older. GCA is an immune-mediated vasculitis of medium to large-sized arteries. It predicates the aorta and its major branches, including the carotid and vertebral arteries. Symptoms of GCA commonly include headache, fever, scalp tenderness, vision loss, and jaw claudication. Severe unilateral or bilateral loss of vision is the most common and feared complication of GCA related to occlusion of the short posterior ciliary arteries with resulting ischemia to the optic nerve. We know this condition as arteritic anterior ischemic optic neuropathy. In addition, there are other ophthalmic manifestations such as amaurosis fugax, diplopia, ptosis, and eye pain. The cornerstone of GCA treatment is glucocorticoids, which must be initiated immediately after diagnosis at a high dose. Nevertheless, glucocorticoids in aging people frequently lead to side effects, and some patients are refractory to this therapy.

CASE PRESENTATION

A 71-year-old male patient has known diagnoses of cured bladder cancer, early-stage Alzheimer's, and previous right leg deep vein thrombosis. He was under apixaban 2x2.5 mg and acetylsalicylic acid 1x100 mg. The patient complained of fatigue, headache, and jaw pain that started one month ago. One week later, vision loss and diplopia occurred in the right eye. In the examination of the patient at the emergency service, with the diagnosis of pharyngitis, antibiotic (amoxicillin and levofloxacin) treatment was applied for two weeks. Jaw claudication and vision loss continued after antibiotic therapy. When his symptoms did not regress, the patient was admitted to the Hacettepe University Hospital. In laboratory evaluation; Hemoglobin: 11.1 gr/dl, leukocyte:5500 x 10^3 / µL, thrombocyte:224000 x 10^3 /µL, erythrocyte sedimentation rate: 112 mm/hour, CRP: 2.5 mg/dL, procalcitonin: 0.029 ng/mL. Temporal Doppler USG resulted in atherosclerotic involvement. Brain MRI showed peripheral contrast enhancement and thickening of the arterial wall in the right temporal artery in favor of giant cell arteritis. No ischemic pathology was found in brain diffusion MRI, brain CT angiography, or carotid vertebral Doppler USG examinations. As a result of the ophthalmology consultation, extensive cotton wool spots were observed in the right retinal evaluation of the patient, and it was evaluated as giant cell arteritis. A biopsy sample containing a 1.5 cm section of the right temporal artery was taken, and giant cell arteritis was detected. The treatment was started urgently without waiting for the biopsy result. 1 g of pulse steroid therapy was given for three days. 48 mg metilprednizolon (MPZ) treatment was given as maintenance steroid treatment. The vision loss in the patient's right eye decreased. After the biopsy result, weekly subcutaneous tocilizumab 162 mg treatment was started. New onset of vision loss occurred in the patient's left eye ten days after the start of pulse steroid treatment. Giant cell arteritis was considered in the ophthalmologic re-evaluation. One gram pulse steroid treatment was applied again for three days. The patient's vision improved after three pulse steroid therapy. After that, maintenance steroid therapy was continued. On the 15th day of the treatment, vision loss occurred again in the left eye. Pulse steroid therapy for five days was planned for resistant giant cell arthritis eye involvement. The patient's vision

partially improved after five pulse steroid therapy. It was planned to take 80 mg mpz as a maintenance treatment. The patient's treatment is still ongoing.

DISCUSSION

The case is remarkable for different aspects, particularly for the dramatic aggressiveness of the disease despite intravenous MPZ, manifesting by A-AION progression in the first eye and rapid second eye involvement. There is no consensus in the literature concerning the optimal therapeutic approach in GCA patients with corticosteroid-resistant A-AION. Methotrexate could provide a long-term, but modest CS is sparing effect. However, this treatment is not an option for rapid control of the refractory disease. Cyclophosphamide may represent an option for patients with the resistant disease despite high doses of CS or requiring prolonged medium- to high-dose CS therapy. Data are nevertheless limited. We did not plan cyclophosphamide treatment because of bladder cancer.

Recently, it has been shown that TCZ can rapidly induce and maintain GCA remission in a prospective study, several case reports, and some retrospective studies. A phase III trial of tocilizumab showed sustained GC-free remission in 56% of patients receiving weekly subcutaneous tocilizumab alongside the 52-week tapered course of GC, compared with 18% in the 52-week GC taper placebo arm, respectively. Patients treated with tocilizumab required over 40% lower cumulative doses of GC and experienced fewer GC-related adverse events than the placebo group. These results led to the approval of subcutaneous tocilizumab (162mg/week) by NICE for relapsing and refractory GCA.

TCZ was given subcutaneously to all patients included in the GiACTA trial, whereas it was administered intravenously to 79.1% of the clinical practice series. Despite these differences in the route of administration, the number of patients achieving sustained remission was almost similar in both groups (54.6% in the GiACTA trial and 70.4% in the clinical practice group (iv tocilizumab every four weeks); p=0.42) [1].

REFERENCES

[1] Stone JH, Tuckwell K, Dimonaco S, Klearman M, Aringer M, Blockmans D, et al. Trial of Tocilizumab in Giant-Cell Arteritis. N Engl J Med. 2017;377(4):317-28.