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Involvement of temporal artery as a rare presentation of IgG4-related disease

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Abstract

Immunoglobulin G4-related disease (IgG4-RD) is a chronic systemic condition characterized by fibroinflammatory features that can affect multiple organs. Ophthalmic involvement commonly presents with tumor-like masses, particularly in the lacrimal glands and orbital tissues. However, involvement of temporal artery as a rare manifestation of IgG4-RD has been infrequently reported in the literature. Herein, we present a case demonstrating neuro-ophthalmic involvement in which involvement of temporal artery emerged as a clinical manifestation of IgG4-related disease.

Keywords: Immunoglobulin G4-related disease, temporal arteritis

We herein report a case characterized by temporal artery involvement in the context of Immunoglobulin G4-Related Disease (IgG4-RD), presenting with an atypical clinical manifestation.

CASE REPORT

A 63-year-old male patient presented with complaints of diplopia, prominent vessels in the right temporal area, and pain and swelling around the right eye, which had started 15 days prior.

On examination, his arterial blood pressure was 120/75 mmHg, pulse 78/min, and body temperature 36.3˚C. Neurologically, he was conscious, cooperative, and oriented. His pupils were isochoric, with intact bilateral direct and indirect light reflexes. Visual acuity was LogMAR 0.4 in the right eye and LogMAR 0.0 in the left eye. Fundus examination and intraocular pressures were within normal limits. Proptosis was observed in the right eye, with restricted upward and inward gaze (Figure 1). The remainder of the neurological examination was unremarkable. His medical history included well-controlled hypertension and diabetes mellitus. He also had a history of herpes keratitis, dacryocystitis and allergic rhinitis, which had resolved after treatment. Family history was unremarkable.

Routine biochemistry showed normal liver and kidney function, electrolytes, and lipid profile. However, amylase (146 Units/Liter (U/L)), lipase (178 U/L), C-reactive protein (CRP) (1 milligrams/deciliter (mg/dL)), and erythrocyte sedimentation rate (ESR) (59 millimeters/hour (mm/h)) were elevated. Hemogram showed an increased white blood cell count (12.5×10^3/microliter (μL)) and eosinophil count (1.4×10^3/μL).

Thyroid function tests and autoantibodies were within normal limits, ruling out thyroid

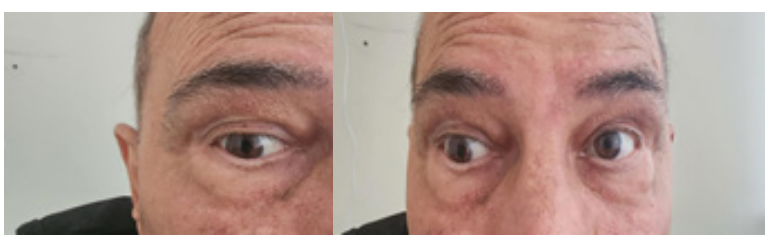

Figure 1. Proptosis and restricted inward and upward gaze were observed in the right eye.

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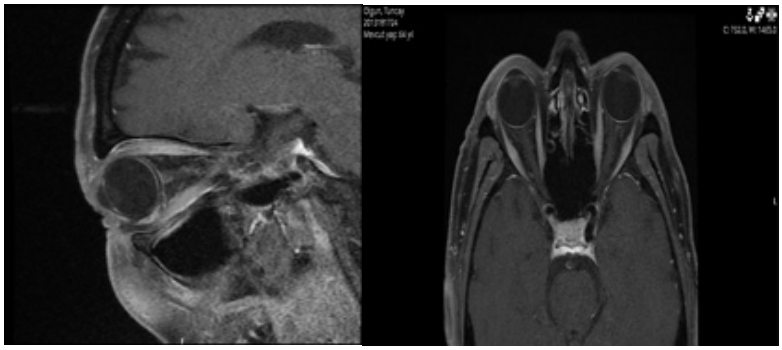

Figure 2. Proptosis in the right eye, increased T2 signal compatible with edema in the eyelids, and significant thickening in the muscle belly of the extraocular muscle groups were observed.

ophthalmopathy. Vasculitis, tumor markers and infectious markers (syphilis, brucella, toxoplasma) were negative. Complement levels were within normal limits and no atypical cells were seen in the peripheral smear. Thoracic and abdominal tomography showed no pathology.

Evaluation for IgG4-RD revealed elevated IgG4 (394 mg/dL; normal: 3–200 mg/dL) and total IgE (1755 Units/milliliter (IU/mL); normal: 0–87 IU/mL). Cranial MRI was unremarkable, while orbital MRI showed proptosis of the right eye, T2 hyperintensity indicating periorbital edema, thickening of the extraocular muscle groups and lacrimal gland degeneration (Figure 2).

Due to pain in the temporal area and prominent vessels, temporal artery ultrasonography (USG) was performed, showing arterial wall thickening up to 1 mm along a 2 centimeter (cm) segment of the bilateral superficial temporal artery with reduced luminal flow (Figure 3). Temporal artery biopsy revealed eosinophilic infiltration in the adventitia, extending into the muscular layer and intima, with additional inflammatory infiltration in the adventitia (Figure 4).

Based on clinical and histological findings, the patient was diagnosed with involvement of temporal artery associated with IgG4-related vascular involvement. Treatment with methylprednisolone (0.5 milligrams/kilogram/day) and clopidogrel (75 mg once daily) was initiated, leading to significant improvement. Approximately one month later, significant improvement was noted in the patient's proptosis and restriction of eye movements. Visual acuity had improved to LogMAR 0.1 in the right eye and LogMAR 0.0 in the left eye. At the follow-up visit two months later, the neurological examination was entirely normal.

DISCUSSION

Immunoglobulin G4-RD is an immune-mediated fibroinflammatory condition affecting multiple organs.[1] The most frequently involved organs include the esophagus, lacrimal glands, major salivary glands, pancreas, bile ducts, retroperitoneum, lungs, kidneys, aorta, and thyroid gland.[2,3] Neurological manifestations including orbital disease, hypertrophic pachymeningitis, hypophysitis and parenchymal involvement are less common but clinically significant.[4] Temporal artery inflammation can result from

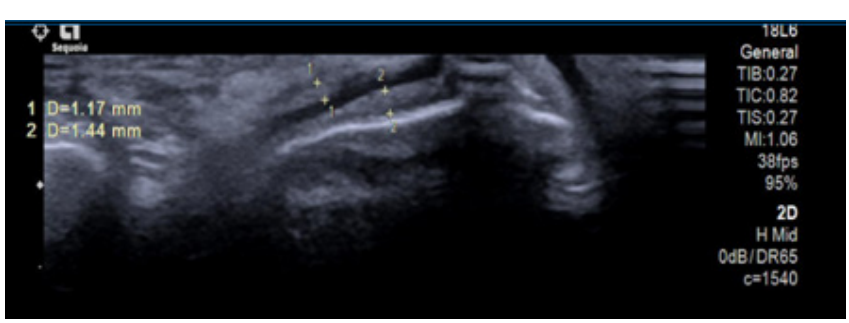

Figure 3. In temporal artery USG, an increase in arterial wall thickness of up to 1 mm is observed in the superficial artery in a segment approximately 2 cm from the medial of the tragus.

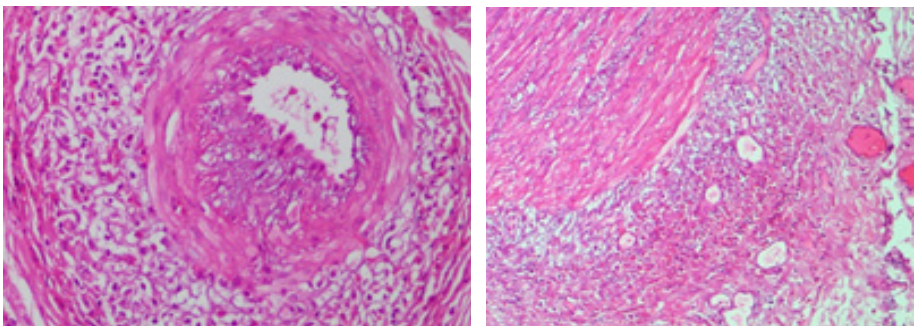

Figure 4. In the vascular biopsy material, eosinophils infiltrating the adventitia and occasionally advancing into the muscular layer and intima were observed. This is accompanied by chronic inflammatory cell infiltration in the adventitia. Intimal proliferation was observed.

a variety of underlying conditions, including IgG4-RD, amyloidosis, polyarteritis nodosa, eosinophilic granulomatosis with polyangiitis, and granulomatosis with polyangiitis.[5] In other words, not all cases of temporal artery inflammation are attributable to "temporal arteritis" or "giant cell arteritis." Although temporal arteritis may be considered the most likely diagnosis in such cases, early exclusion of less common etiologies is crucial for ensuring an accurate diagnosis and appropriate treatment process.

In temporal arteritis, a significant reduction in visual acuity is typically noted upon examination. Vision loss often reaches the level of counting fingers or even complete absence of light perception. This impairment is primarily attributed to the frequent involvement of the ophthalmic artery and/or its branches, which can result in ocular or even orbital ischemia.[6] Furthermore, fundus examination in temporal arteritis may reveal signs of ischemic optic neuropathy, such as a mildly pale and edematous optic disc, soft exudates, and small retinal hemorrhages.[7,8] In contrast to the typical presentation of temporal arteritis, our patient did not exhibit notable visual impairment at initial assessment. Fundus findings were within normal limits.

Elevated or significantly elevated levels of inflammatory markers, such as CRP and ESR, represent key laboratory findings in the diagnosis of temporal arteritis. In the majority of cases, ESR values equal to or exceeding 100 mm/hour are observed as characteristic findings, and serum CRP levels usually show a parallel increase. However, in approximately 8–22.5% of patients, the acute-phase response may remain within normal limits.[9] In our case, both CRP and ESR levels were not significantly elevated—an atypical finding for classic temporal arteritis, but one that aligns with features of IgG4-RD.

In the diagnosis of temporal arteritis, biopsy findings play a crucial role alongside clinical and laboratory data. Characteristic histopathological features of temporal arteritis include intimal thickening causing luminal narrowing, mononuclear inflammatory cell infiltration, intima-media invasion, disruption of the internal elastic lamina, and the presence of multinucleated giant cells.[10] In our case, the histopathological findings were not consistent with classic temporal arteritis. The temporal artery biopsy revealed inflammatory cell infiltration within the adventitia, along with eosinophils extending from the adventitia into the muscular layer and, in some areas, into the intima—findings suggestive of IgG4-related temporal artery involvement.

An important distinction between temporal arteritis and IgG4-RD lies in their treatment response and overall prognosis. In cases where temporal artery involvement is due to alternative causes such as IgG4-RD, the risk of vision loss is minimal or even absent—unlike in classical temporal arteritis.[11,12] In our patient, the clinical presentation differed from typical temporal arteritis, with the arterial inflammation attributed to IgG4-RD. As a result, there was no evidence of rapidly progressive or severe visual impairment. Notably, visual acuity improved significantly following corticosteroid therapy, reaching LogMAR 0.1 in the right eye and LogMAR 0.0 in the left. In contrast, temporal arteritis often progresses rapidly, and the extent of vascular involvement critically affects treatment outcomes and prognosis. Despite early initiation of high-dose corticosteroids, visual outcomes in temporal arteritis are frequently unfavorable.[6]

Cases of IgG4-related periaortitis and periarteritis have been reported in the literatüre.[13] With our case, we aimed to highlight that, unlike previously reported clinical presentations in the

literature, involvement of the temporal artery and associated inflammation may occur in IgG4-RD, and that this condition should be considered in the differential diagnosis. By identifying involvement of temporal artery in IgG4-RD, we present it as a distinct clinical entity not previously emphasized in the literature.

DISCLOSURE

Ethic: The participant has consented to the publication.

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Conflict of interest: None

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