CASE REPORT

Limited Wegener's Granulomatosis Presenting with an Isolated Abduction Deficit

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ABSTRACT

Wegener's granulomatosis often affects the orbit, typically presenting with painful proptosis. The authors describe a 14 year-old girl, with limited Wegener's granulomatosis, who initially presented with an isolated painless abduction deficit that spontaneously resolved over several weeks. She subsequently developed painful proptosis and diplopia, followed by facial and oral nodules. This case demonstrates that limited Wegener's granulomatosis can rarely present with an isolated painless abduction deficit.

Keywords: Abduction deficit, diplopia, limited Wegener's granulomatosis, proptosis, vasculitis

INTRODUCTION

Wegener's granulomatosis is a vasculitis affecting small- and medium-sized vessels with a predilection for pulmonary and renal involvement. Limited forms of Wegener's granulomatosis affect non-life-threatening organ systems and carry a better prognosis than systemic Wegener's granulomatosis.¹ Orbit involvement is common and typically presents with painful proptosis. We report a patient with limited Wegener's granulomatosis who presented with an isolated painless abduction deficit and subsequently developed more characteristic symptoms and signs, including painful proptosis and diplopia, followed by facial and oral nodules.

CASE REPORT

A 14 year-old girl with an unremarkable past history, presented with a 10-day history of painless binocular horizontal diplopia. Examination revealed visual

acuity of 20/20 OU and full visual fields. Anterior segment, posterior segment, external, and pupil examinations were unremarkable. There was no proptosis, periorbital oedema, chemosis, or redness OU (Figure 1). Ocular motor examination revealed a –3 abduction deficit OD with a large incomitant esotropia measuring 20 prism dioptres in primary position (Figure 1). Her cranial nerves were otherwise intact. Magnetic resonance imaging (MRI) of the brain was unremarkable, but the orbits were obscured by artifact from her braces (Figure 2). She was observed. Two weeks later, her diplopia had improved. Examination revealed a –1 abduction deficit OD with an incomitant esotropia measuring 12 prism dioptres in primary position. Her diplopia fully resolved 7 weeks after her initial presentation.

Two weeks later, however, she developed pain and swelling OD, binocular oblique diplopia, and a lateral canthus nodule OD. Examination revealed periorbital oedema, erythema, ptosis, and axial proptosis OD (Figure 3). Ductions were full, but there was a

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small-angle esotropia and incomitant left hypertropia measuring 16 prism dioptres in primary position. Her braces were removed so that a better imaging scan could be obtained. MRI of the orbits showed enlargement of the right lacrimal gland and the lateral, superior, and inferior rectus muscles, with preseptal oedema and enhancement (Figure 4). There was enhancement in the right superior orbital fissure with expansion of the right cavernous sinus (Figure 4). Laboratory studies, including antineutrophil cytoplasmic antibodies (ANCA) and angiotensin-converting enzyme (ACE), and chest x-ray were unrevealing.

Orbital biopsy was considered, but the patient requested empiric steroid treatment. Her symptoms and signs rapidly resolved on prednisone (1 mg/kg/day). The prednisone was slowly tapered over 4 months. Within days of stopping the steroids, she developed several non-tender subcutaneous facial and oral nodules. Biopsy of the nodules showed necrotizing granulomatous inflammation with multinucleated giant cells (Figure 5). Some granulomas had a linear and branching appearance, but no overt vasculitis was found. There were no acid-fast bacilli (AFB) or fungi on AFB and Gömöri methenamine silver (GMS) stains. Polymerase chain reaction (PCR) for AFB and fungi was negative. Repeat laboratory studies, including ANCA and erythrocyte

sedimentation rate (ESR), were unrevealing. Lyme and *Histoplasma* serologies were negative. *Mycobacterium tuberculosis* antigen was not detected by QuantiFERON-TB (QFT) assay. Chest computed tomography (CT) was unremarkable. Urinalysis and urine microscopy were unremarkable. On the basis of her presentation, biopsy findings, negative ANCA, and response to steroids, she was diagnosed with limited Wegener's granulomatosis and was treated with methotrexate. The nodules resolved. With the exception of one mild relapse that resolved with steroids, her disease has remained quiescent.

COMMENT

Orbital involvement occurs in approximately 15% of patients with Wegener's granulomatosis,² but up to 65% of cases with orbital involvement have the limited variant of the disease.³ The triad of granulomatous inflammation, tissue necrosis, and vasculitis on biopsy confirms the diagnosis of Wegener's granulomatosis, but the full triad is often not found in the limited form. In a review of 126 biopsies from patients with Wegener's granulomatosis, the full triad was present in only 16% of all head and neck specimens.⁴ In limited Wegener's granulomatosis, vasculitis is frequently not identified,⁵ as in our

FIGURE 1 Clinical photograph at presentation showing esotropia.

FIGURE 3 Clinical photograph at follow-up showing periorbital oedema, erythema, and ptosis OD.

FIGURE 2 Magnetic resonance imaging with contrast (A, axial; B, coronal) at presentation showed normal cavernous sinuses. The orbits were completely obscured by artifact from braces.

FIGURE 4 Magnetic resonance imaging with contrast (A, axial; B, coronal) at follow-up showed right periorbital inflammation with enlargement of the lacrimal gland, lateral rectus, superior rectus, and anterior portion of inferior rectus. There was enhancement in the right superior orbital fissure (arrow) with expansion of the right cavernous sinus (arrowhead).

FIGURE 5 (A) Biopsy of an oral nodule showed granulomatous inflammation (arrowheads) with multinucleated giant cells (arrows). (B) Some necrotizing granulomas had a linear and branching appearance (arrows), but no overt vasculitis was found.

patient. Patients with the limited form are more likely to be female, present at a younger age, and to have a negative ANCA.^{5,6} In addition, they have a better prognosis and respond more favourably to treatment, often going into remission after initiation of treatment or requiring only a small dose of maintenance therapy.¹ Our patient's presentation and investigation results were most consistent with limited Wegener's granulomatosis. The presence of necrotizing granulomatous inflammation, a normal chest CT, and normal ACE level argued against sarcoidosis. An infectious aetiology was also considered, but extensive testing for possible infectious aetiologies was unrevealing. Furthermore, the rapid and sustained clinical improvement with prednisone was felt to be more consistent with an inflammatory than infectious aetiology.

Our patient's presentation with an isolated abduction deficit was unusual for limited Wegener's granulomatosis. Orbital involvement most commonly presents with subacute painful proptosis.⁷ Over 50% of patients with orbital involvement develop diplopia

at some stage of the disease, but less than 7% present with isolated diplopia.³ The diplopia can be due to inflammation resulting in tethering of or mass effect on extraocular muscles, direct infiltration of muscles, vasculitis of the vasa nervorum of muscles, or cranial nerve involvement.³

Because our patient's initial MRI was suboptimal due to artifact from her braces, we can only speculate as to the cause of her abduction deficit. Wegener's granulomatosis can cause cranial neuropathies, most often affecting the second, sixth, and seventh nerves.^{8,9} There is only one prior case report of a patient with a sixth nerve palsy as the initial manifestation of limited Wegener's granulomatosis.¹⁰ The patient's sixth nerve palsy improved with oral prednisone,¹⁰ whereas our patient's abduction deficit improved spontaneously. A potential mechanism for sixth nerve palsy in our patient would be ischemia, since the spontaneous recovery of her abduction deficit over several weeks was similar to that seen with microvascular sixth nerve palsy.¹¹ In summary, our case demonstrates that limited Wegener's

granulomatosis can rarely present with an isolated abduction deficit.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figures 1 and 5 of this article is available in colour online at www.informahealthcare.com/oph

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