CASE REPORT

Unusual aetiology of isolated lower motor neuron facial palsy: systemic lupus erythematosus presenting with cranial nerve palsy and nephritis

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SUMMARY

A 20-year-old woman presented with common cold symptoms was found to have a left-sided facial droop. On examination, peripheral facial nerve palsy was confirmed. Subsequent testing showed nephrotic range proteinuria and positive serologies including antinuclear antibody and anti-smith antibody. Kidney biopsy showed stage III lupus nephritis. Treatment with pulse steroids along with mycophenolate mofetil for her lupus nephritis resulted in concomitant improvement of her facial palsy.

BACKGROUND

This is a rare presentation of systemic lupus erythematosus (SLE) with a cranial palsy, specifically facial nerve palsy.

CASE PRESENTATION

A 20-year-old woman presented to the emergency department with a 10-day history of fatigue, myalgia, diffuse body aches, high-grade fevers, coryzal symptoms, pleuritic chest pain and haematuria. She also noted drooling out of the left side of her mouth as well as inability to close her left eye. On examination, she had complete paralysis of the upper as well as the lower parts of the left side of the face. General examination was otherwise unremarkable; in particular, there were no rashes, mucocutaneous ulcers or signs of active arthritis.

INVESTIGATIONS

Biochemical and haematological investigations showed microcytic hypochromic anaemia, basic metabolic panel showed blood urea nitrogen and serum creatine within normal range, however, routine urinalysis was remarkable for 4+ protein, 3+ blood; spot urine showed a protein-creatinine ratio of 3.4 mg/mg. HIV and hepatitis B/C serologies were negative, and fasting blood glucose, glycosylated haemoglobin were normal.

Autoimmune serological tests were positive for antinuclear antibody with a titre 1:20 480, anti-smith (SM), anti-ribonucleoprotein (RNP), anti-SM/RNP, anti-Sjogren's syndrome A and antichromatin along with elevated C reactive protein and erythrocyte sedimentation rate-WESTERGREN of 2.86 and 91, respectively. In addition, patient labs did not indicate antiphospholipid syndrome given normal dilute Russell viper venom time and anticardiolipin antibodies. However, she had normal complement levels C3, and C4 as well as negative IgM antibodies for herpes simplex virus

(HSV), Epstein-Barr virus, cytomegalovirus and varicella-zoster virus, influenza A/B PCR.

Kidney biopsy showed early transformation to focal proliferative lupus nephritis.

DIFFERENTIAL DIAGNOSIS

Unilateral peripheral facial nerve palsy is a common entity with the most common aetiology being HSV. However, peripheral neuropathy could also be a manifestation of an underlying autoimmune disease. A similar case of facial palsy has been reported previously in association with systemic lupus erythematosus as described by Gupta *et al.*¹

TREATMENT

The patient was initially started on steroids and valacyclovir as an inpatient for presumed Bell's palsy. When the diagnosis of lupus nephritis was established, she was switched to pulse dose steroids, and mycophenolate mofetil was added to her treatment regimen for induction.² Given the rarity of lupus presenting as a cranial nerve palsy; the valacyclovir was continued for 10 days despite the negative HSV serologies. She was later discharged on a slow steroid taper.

OUTCOME AND FOLLOW-UP

The patient had resolution of her facial palsy and improvement in markers of active nephritis at 4 months of follow-up.

DISCUSSION

Neurological involvement in SLE is not uncommon. It varies from seizures, stroke, transverse myelopathy, to peripheral neuropathy resembling Guillain-Barre syndrome. Hence it may include both the peripheral and the central nervous system.³⁻⁵ However, cranial palsy is a very rare feature of SLE and is only described in case reports or case series. Moreover, most of the cases described in the literature actually developed neurological manifestations through the course of their disease rather than on initial presentation.⁶ Saleh *et al* described a case of lupus presented initially with isolated sixth cranial nerve (CN) palsy; later developed classic manifestations of SLE confirmed by serologies. Gupta *et al* described a woman with recurrent facial nerve palsy that was subsequently diagnosed with SLE. Our patient presented with seventh CN palsy that was initially presumed to be post viral Bell's palsy. However, she was incidentally found to have nephrotic range

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proteinuria with active urine sediment that led to the diagnosis of active stage III lupus nephritis and SLE based on kidney biopsy and positive serologies. It would be challenging to confirm the direct association of the CN VII palsy to SLE. However, it is plausible that her facial palsy is a manifestation of underlying SLE. One of the suggested pathophysiological mechanisms of SLE involve immune dysregulation with potential activation of the disease process by infections which may also be the trigger for CN VII palsy.⁷ Potential mechanisms for CN VII palsy in SLE include autoimmune antibodies against neuronal antigen or inflammatory cytokines; both of which may be triggered by an infectious process, viral or bacterial. Another mechanism involve thrombotic vasculopathy mediated by anti-phospholipid antibodies.³

The mainstay of treatment in SLE-related cranial palsy is immunosuppression. Most of the studies evaluating the efficacy of different therapeutic regimens are case reports that included only small number of patients. However, Baril Fabris *et al* described a randomised controlled trial of patients with severe neuropsychiatric manifestations of SLE to either intravenous cyclophosphamide or pulse steroids. There was much higher response rates in intravenous cyclophosphamide group at 2 years of follow-up.²

According to American College of Rheumatology nomenclature definitions, when describing neuropsychiatric systemic lupus erythematosus (NPSLE), it is required to have a certain NPSLE in addition to three or more other lupus manifestations. Pertaining cranial nerve presentations it depends more on clinical gestalt given rarity of this presentation. Therefore in our case we ruled out initially other common causes such as viral. Also, given the simultaneous presentation of the cranial nerve palsy and lupus we made that association.⁸

On review of European League against Rheumatism recommendations in diagnosis and management, it might be helpful to delineate is an inflammatory process versus coagulopathic. However, it is usually challenging and not highly accurate especially in peripheral neuropathies. Knowing that, our case anti-phospholipid serology negativity and other lab studies indicating active inflammation favours the inflammatory process. It is recommended that in either inflammation or coagulopathy to initiate steroids and immunomodulators, but in case of antiphospholipid syndrome intravenous route is preferable with glucocorticoids administered in a pulse dose.⁹

Learning points

- ▶ Consider autoimmune disease among the differential diagnosis of neuropathy even isolated cranial nerve palsies.
- ▶ Keep high index of suspicion for lupus given its varied clinical presentations which could unveil an underlying process such as lupus nephritis.
- ▶ When managing cranial nerve palsy with unclear etiology, initially treat broadly.

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