

## CLINICAL VIGNETTE

# A Case of Bilateral Peripheral Facial Nerve Palsy

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### Case Report

A 32-year-old Caucasian female with a history of idiopathic thrombocytopenic purpura, benign thyroid nodules, and left-sided Bell's palsy presented to her primary care provider with new-onset right-sided facial droop. One day prior, the patient woke up with right facial muscle tightness and inability to close her right eye. Over the next day, she developed severe right-sided otalgia. She denied weight loss, facial numbness, tingling, pain, diaphoresis, and auditory or visual changes. She denied taking new medications.

Eight weeks prior, the patient noted left-sided facial droop, inability to close her left eye, and significant left-sided otalgia. She was diagnosed with Bell's palsy and prescribed a 12-day course of prednisone and valacyclovir, with mild improvement. She subsequently visited her ophthalmologist for left eye dryness and had near complete left-sided CNVII palsy with 50% lid function and poor recovery. Initial laboratory work up, including rheumatoid factor (RF), antinuclear antibody, angiotensin-converting enzyme (ACE), and comprehensive metabolic panel, was unremarkable. Brain magnetic resonance imaging (MRI) was ordered. The patient reported having left-sided Bell's palsy four years prior that resolved within two weeks with valacyclovir and prednisone.

Given the patient's rapid progression of symptoms and the low incidence of bilateral Bell's palsy, she warranted further neurological work up. Brain MRI with and without contrast revealed enhancement along the fundus and labyrinth segments of the bilateral seventh cranial nerves, more conspicuous on the right, indicating more recent inflammation. Her serologies were positive for herpes simplex virus (HSV) immunoglobulin G (IgG), Mycoplasma pneumonia IgG, and Epstein Barr Virus (EBV)-EBNA IgG. Of note, the HSV1/2 IgM, Mycoplasma pneumonia IgM, and EBV-Viral Capsid Antigen (VCA) IgM were negative. Her bloodwork was otherwise unremarkable, including negative human immunodeficiency virus (HIV), varicella zoster virus (VZV), *B.burdorferi* antibodies, HSV antibodies, anti-Sjogrens syndrome-related antigens A and B antibodies, Epstein Barr Virus polymerase chain reaction (PCR), rapid plasma reagin, and interferon gamma release assay tests. A lumbar puncture (LP) demonstrated increased lymphocytes (95%) and an elevated EBV-VCA IgG level. The cerebral spinal fluid (CSF) did not reveal oligoclonal bands or elevated ACE level, and was negative for HSV1/2 IgG, VZV IgG, VZV DNA PCR, cytomegalovirus (CMV) DNA PCR and albuminocytologic dissociation. She completed a 12-day course

of prednisone and acyclovir with improvement in her right facial droop. Based on her labwork, it was felt that her Bell's palsy was likely EBV-related. Two months later, her ophthalmologist noted complete resolution of her symptoms.

### Discussion

The two most common causes of acute facial paralysis are ischemic stroke and Bell's palsy.<sup>1</sup> Given the time-critical treatment for acute stroke, physicians must quickly make the distinction in order to avoid permanent damage. The difference in clinical findings between the two conditions may be subtle.

Bell's palsy was first described by Sir Charles Bell in the 1920s as facial paralysis caused by trauma to the peripheral branches of the facial nerve.<sup>2</sup> Today it is defined as an idiopathic acute peripheral facial nerve palsy. The incidence is 25-30 cases/100,000 people/year, accounting for 60-75% of all cases of facial paralysis.<sup>3</sup> It affects men and women equally with the highest rate between the ages of 15-60.<sup>3,4</sup> Deficits accumulate over hours to days. Symptom severity peaks between three to seven days. Roughly 85% of patients show partial recovery within three weeks and full recovery within six months.<sup>1</sup> Bell's palsy is caused by facial nerve damage at the level of or below the brainstem.<sup>4,5</sup> The majority of proposed etiologies are viral infections, including HSV, VZV, EBV, CMV, HIV and influenza.<sup>6</sup> Other possible triggers include microvascular insult from uncontrolled diabetes, third trimester of pregnancy, malignancies, Lyme disease, and Guillain-Barre Syndrome (GBS).<sup>7</sup>

Patients with Bell's palsy present with weakness in the ipsilateral upper and lower face that progress over hours to days. Clinical findings include the inability to wrinkle the forehead, close the eyes completely, or smile on the affected side. The facial nerve also innervates the stapedius muscle of the ear and the nervus intermedius, which provides sensation for anterior two-thirds of tongue. Therefore, these patients may also experience ipsilateral otalgia from stapedius muscle inflammation and impaired sense of taste.<sup>8</sup>

Facial weakness caused by strokes is largely dependent on the location of occlusion within the brain and can develop within seconds to minutes.<sup>9</sup> Patients with central facial palsy present with the inability to smile or purse lips, but retain the ability to wrinkle the forehead and close the eye on the affected side. This finding is attributed to the dual innervation of the upper face,

which allows sparing of the contralateral upper facial muscles if the lesion damages the upper motor neurons in the motor cortex or internal capsule.<sup>4,5</sup> Strokes involving the cortex generally cause central facial weakness, sparing the forehead and eyes, but strokes involving the brainstem can mimic a peripheral lesion. Here, the patient often presents with other focal neurologic deficits, such as numbness, dysarthria, visual impairment, vertigo, and ataxia.<sup>9,10</sup> Patients suspected of having a stroke should visit the emergency department immediately for further neurological evaluation and imaging.

Bell's palsy is classified into five categories: unilateral non-recurrent, unilateral recurrent, simultaneous bilateral, alternating bilateral, and recurrent bilateral.<sup>11</sup> While unilateral non-recurrent Bell's palsy is often assumed to be idiopathic or associated with HSV, recurrent unilateral Bell's palsy has been loosely linked to malignant hypertension, diabetes, pregnancy, and brain malignancies. Melkersson Rosenthal syndrome, characterized by recurrent unilateral facial paresis, facial edema, and fissured lips, should also be considered.<sup>12</sup> An estimated 4-7% of cases of Bell's palsy have recurrent facial palsy.<sup>8</sup> In contrast, bilateral Bell's palsy is exceedingly rare, noted only 0.3-2% of cases, and often implicates an underlying systemic disease.<sup>7</sup> Both sides of the face must be involved within a 30-day period to be defined as bilateral facial paralysis.

The most common causes of bilateral facial palsy are Lyme disease (36% of cases), GBS (5%), trauma (4%), sarcoidosis (0.9%), and AIDS (0.9%).<sup>13</sup> Lyme disease is the leading cause of bilateral facial palsy; approximately 30-35% of infected individuals suffer from facial paralysis.<sup>7</sup> Serologies may show IgM antibodies against *B. burgdorferi* and CSF lymphocytosis. GBS is typically a post infective polyneuropathy characterized by progressive limb weakness and areflexia. Of patients with GBS, 35% present with bilateral facial weakness, increasing to 50-60% in severe cases.<sup>14</sup> Diagnosis is made clinically and with CSF revealing albuminocytologic disassociation. In neurosarcoidosis, cranial nerve neuropathy, particularly of the facial nerve, is the most common complication. Upwards of 50% of patients with neurosarcoidosis experience a form of facial palsy, and 25-50% of these patients have either simultaneous or sequential bilateral facial nerve involvement.<sup>14</sup> Chest radiography, (CXR) ACE serologies, and biopsy of non-caseating granulomas are considered in diagnosing sarcoidosis. However, the remainder of the physical exam, labwork, and imaging may be unremarkable.<sup>15</sup> In HIV-1, it is estimated that 40-90% of patients with primary HIV-1 infection experience acute retroviral syndrome with seroconversion.<sup>16</sup> However, facial paralysis may precede seroconversion by four to six weeks.<sup>17</sup> In immune-depressed patients, paralysis is more likely caused by an AIDS-defining illness. Thus, patients with bilateral facial paresis who test HIV-negative, should repeat testing after six weeks.<sup>16</sup>

Recently, EBV is gaining attention as a cause of bilateral facial palsy. Neurological complications are reported in about seven percent of EBV infections, most commonly affecting the facial nerve. Approximately 40% of EBV-associated facial nerve

palsy cases are reported as bilateral.<sup>16</sup> EBV-induced facial palsy is well documented in the pediatric population, but there are only rare reports amongst adults. One case report of a 14-month-old girl with bilateral facial palsy presumed to be caused by EBV showed negative EBV-VCA IgM and EBV-EBNA IgM, which should rule out acute EBV. However, her serologies revealed positive EBV-VCA IgG after one month and positive EBV-EBNA IgG after 5 months.<sup>18</sup> Another case describes a 43-year-old female who presented with bilateral facial droop progressing over three days with serological tests that also revealed negative EBV-VCA IgM but positive EBV-VCA IgG<sup>19</sup> with no clinical signs of EBV. Although there is limited literature, one theory is that uncontrolled, chronic EBV may induce B-lymphocytosis, leading to Non-Hodgkin's lymphoma (NHL). Facial paralysis has also been linked to early stages of NHL.<sup>19</sup>

Diagnostic work up for Bell's palsy is extensive, including brain MRI, serologic and CSF evaluation, CXR, electromyography, nerve conduction tests, and organ biopsy in severe cases. Currently, the treatment recommended is typically prednisone 60-80mg/day for one week within 72 hours of onset, and only advancing to the addition of antivirals with severe or persistent symptoms.<sup>20-22</sup> In cases of suspected systemic diseases, such as Lyme disease, GBS or neurosarcoidosis, the underlying disease should be treated. With EBV, there is insufficient evidence for treatment beyond supportive care. In a study of 94 patients with acute infectious mononucleosis, acyclovir and prednisone reduced oropharyngeal shedding, but did not affect symptom duration.<sup>23</sup>

In our patient, there were eight weeks between the development of her left and right facial palsy. It is difficult to assess whether this is overlapping alternating unilateral facial palsy, recurrent unilateral Bell's palsy with coincidental development of the contralateral side, or recurrent unilateral Bell's palsy on her left side within eight weeks and subsequent development of bilateral Bell's palsy within 30 days of the recurrence. Nonetheless, her case warranted emergent imaging and work-up to definitively rule out an acute ischemic stroke. Extensive investigation, including serologic workup, lumbar puncture, and imaging should be performed to rule out an underlying systemic disease that may require alternative treatment.

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