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Potential Misdiagnosis of Bell's Palsy in the Emergency Department

Jahan Fahimi, MD, MPH,
Department of Emergency Medicine, University of California, San Francisco; San Francisco, CA, USA, Department of Emergency Medicine, Alameda County Medical Center – Highland Hospital; Oakland, CA, USA

Babak B. Navi, MD, and
Department of Neurology, Brain and Mind Research Institute, Weill Cornell Medical College; New York, NY, USA

Hooman Kamel, MD
Department of Neurology, Brain and Mind Research Institute, Weill Cornell Medical College; New York, NY, USA

Abstract

Study Objective—We evaluate the incidence of potentially incorrect emergency department (ED) diagnoses of Bell's palsy and identify factors associated with identification of a serious alternative diagnosis on follow-up.

Methods—We performed a retrospective cohort study from California's Office of Statewide Health Planning and Development (OSHPD) for 2005–2011. Subjects were adult patients discharged from the ED with a diagnosis of Bell's palsy. Information related to demographics, imaging use, and comorbidities were collected. Our outcome was one of the following diagnoses made within 90 days of the index ED visit: stroke, intracranial hemorrhage, subarachnoid hemorrhage, brain tumor, central nervous system infection, Guillain-Barre syndrome (GBS), Lyme disease, otitis media/mastoiditis, or herpes zoster. We report hazard ratios (HR) and 95% confidence intervals (CI) for factors associated with misdiagnosis.

Results—A total of 43,979 patients were discharged with a diagnosis of Bell's palsy. Median age was 45. On 90-day follow-up 356 patients (0.8%) received an outcome diagnosis, and 39.9% were made within 7 days. Factors associated with the outcome included increasing age (HR 1.11, 95% CI 1.01–1.21, every 10 years), black race (HR 1.68, 95% CI 1.13–2.48), diabetes (HR 1.46, 95% CI 1.10–1.95), computed tomography or magnetic resonance imaging use (HR 1.43, 95% CI 1.10–1.85). Private insurance was negatively associated with an alternative diagnosis (HR 0.65, 95% CI 0.46–0.93). Stroke, herpes zoster, GBS, and otitis media accounted for 85.4% of all alternative diagnoses.

Conclusion—Emergency providers have a very low rate of misdiagnosing Bell's palsy. The association between imaging use and misdiagnosis is likely confounded by patient acuity. Increasing age and diabetes are modest risk factors for misdiagnosis.

Conflicts of Interest: None

Author Contributions:
BBN and HK conceived of the study. HK obtained the data and performed the statistical analysis. JF and HK interpreted the results. JF drafted the manuscript, and all authors contributed significantly to its editing and review.

Introduction

Background

Bell's palsy represents an idiopathic, unilateral facial paralysis with an annual incidence of approximately 15 per 100,000.[1] It remains the most common cause of unilateral facial paralysis, affecting men and women equally, and while the median age at onset is 40, it can occur at any age.[2] Patients with Bell's palsy commonly present with partial or complete weakness of the muscles of one-half of the face, resulting in an inability to raise eyebrows, wrinkle the forehead, or close the eyelid; loss of the nasolabial fold; and drooping of the angle of the mouth. Other associated symptoms include alterations in taste, hyperacusis, inability to produce tears, and a subjective feeling of facial numbness, though objective sensation is preserved.

The pathophysiology of Bell's palsy involves inflammation of the facial nerve at the geniculate ganglion causing compression and associated ischemia or demyelination.[3] Symptoms are often acute and progressive in nature, evolving over one day to one week. It has been postulated that the herpes simplex virus type 1 may play a causative role, though this theory is still in question.[4] If left untreated, approximately 85% of patient's with Bell's will experience some recovery,[5] and treatment with corticosteroids, with or without antiviral medications, has been shown to improve outcomes.[6,7]

Importance

Unilateral facial paralysis may be caused by a host of diseases other than Bell's palsy, some more serious or potentially life threatening. Due to the dramatic and distressing nature of acute facial paralysis, patients often seek evaluation in emergency departments (EDs). There are no established guidelines for diagnosing Bell's palsy; therefore, clinicians must rely on thorough history taking and physical examination to distinguish patients who can be safely discharged from those who need evaluation for alternative diagnoses with laboratory testing, imaging, or consultation. The differential diagnostic considerations may include stroke, Lyme disease, human immunodeficiency virus (HIV) infection, diabetes mellitus, sarcoidosis, Ramsey-Hunt syndrome (herpes zoster), lymphoma, or cerebellopontine tumors, among others. A misdiagnosis may lead to inappropriate treatment or morbidity.

Goals of This Investigation

The objective of this study is to describe the incidence and risk factors of potentially incorrect ED diagnosis of Bell's palsy, defined by the subsequent diagnosis of a serious and likely alternative disease.

Methods

Study Design and Setting

This is a retrospective cohort study using administrative claims data from the California Office of Statewide Health Planning and Development (OSHPD) for the years 2005–2011, provided in a publicly available and deidentified format by the Healthcare Cost and Utilization Project. All California hospitals are required to submit detailed information related to encounters and discharges among a wide array of domains through automated reporting software. OSHPD provides detailed reporting and formatting specifications used by hospital analysts to collect and aggregate the data in a uniform manner that are then transmitted via an online system. Uploaded data is then thoroughly verified and validated using software in a multi-step process,[8,9] including correcting errors in data entry (i.e., incorrect formatting, blank or missing fields), flagging significant changes in trends from previous reporting periods, or identifying inconsistent information (i.e., incorrect sex when a

sex-specific procedure was performed). OSHPD does not independently audit individual facility data abstraction processes, but does dedicate analysts to facilities in order to assist with reporting and correcting errors. Additional details related to data collection, as well as detailed documentation for datasets, can be found at OSHPD's Healthcare Information Division website.[10]

For this analysis, we used the State Inpatient Database and Emergency Department Database, both of which contain demographic, clinical, payer, and facility information. These data include visit-level information for California ED and inpatient encounters, excluding federal hospitals. Data within OSHPD represent all such visits and are not sampled.

To protect patient confidentiality in these publically available data, records with unique combinations of demographic variables will have masked one or more variables such that the record becomes de-identifiable. Masked variables are reported as missing. Any missing data was coded as such for analysis, and no values were imputed. Within OSHPD data, a patient-specific unique linkage number allows anonymous tracking of patients across visits. This unique number is based on the social security number and provides a method for linking ED encounters and hospitalizations.

Selection of Participants

We identified a cohort of adult patients (age > 17 years), who were California residents, discharged from the ED with a primary diagnosis of Bell's palsy using *International Classification of Diseases, Ninth Revision* (*ICD-9*) code 351.0. We excluded patients who concurrently were diagnosed with one of the conditions defined as an outcome of interest (see below).

Methods and Measurements

We collected information included in the database related to age, sex, race/ethnicity, primary insurance status, day of the week, computed tomography (CT) or magnetic resonance imaging (MRI) use, and comorbidities (including hypertension, diabetes mellitus, coronary artery disease, congestive heart failure, chronic kidney disease, or chronic obstructive pulmonary disease [COPD]). These covariates have been defined in the OSHPD data and definitions were unchanged during our study period.

Outcomes

The unique linkage number was used to link subjects from the index ED visit to subsequent ED encounters or hospitalizations within 90 days. Our primary outcome was a composite of the following serious ED or inpatient discharge diagnoses (by *ICD-9* code) within 90 days of the index ED Bell's palsy diagnosis: ischemic stroke (*ICD-9* 433.01, 433.11, 433.21, 433.31, 433.81, 433.91, 434.01, 434.11, 434.91, and 436, combined with an algorithm previously validated for identifying acute ischemic stroke[11]), intracranial hemorrhage (*ICD-9* 431), or subarachnoid hemorrhage (*ICD-9* 430); brain tumor (*ICD-9* 191); central nervous system (CNS) infection, including meningitis, encephalitis, and brain abscess (*ICD-9* 320-326); HIV (*ICD-9* 042); Guillain-Barre syndrome (GBS) (*ICD-9* 357.0); Lyme disease (*ICD-9* 088.81); otitis media or mastoiditis (*ICD-9* 381-383); or herpes zoster (*ICD-9* 053). These diagnoses were chosen a priori to represent distinct and clinically important disease entities that may present in a similar fashion to Bell's palsy. We excluded some common mimics from our outcomes (e.g., diabetic neuropathy, sarcoidosis), because while they may initially be confused with Bell's palsy, the clinical significance of misdiagnosis is unclear and patient outcomes would generally not be expected to worsen

from delayed diagnosis. Additionally, we excluded mortality as a separate outcome since any death related to one of the outcome diagnoses would already be captured.

Because there is variability in the clinical significance of individual outcome diagnoses, we considered a secondary outcome limited to life-threatening conditions associated with a central cause of facial paralysis (ischemic stroke, intracranial hemorrhage, subarachnoid hemorrhage, brain tumor, and CNS infection).

Analysis

We report overall counts and proportions of covariates and predictors stratified by our outcome of interest. Continuous data is represented as medians with interquartile ranges (IQRs) and categorical data as percentages with 95% confidence intervals (CIs). We performed survival analysis to determine the combined cumulative incidence of our outcome diagnoses. The risk period started on the date of the index ED visit where a diagnosis of Bell's palsy was made and ended on the date when an outcome diagnosis was made, the patient died, or at 90 days after the index ED visit. A multivariable Cox proportional hazards model was used to identify hazard ratios (HR) and 95% CIs for demographic characteristics, imaging use, and comorbidity-related variables that may be associated with misdiagnosis of Bell's palsy; given the lack of population-level data on this topic, these predictors were chosen based on face validity. We used robust standard errors to account for clustering by ED. All statistical analysis was performed using Stata SE (Version 12, StataCorp, College Station, TX).

We received an exemption from review for this study from our local institutional review board.

Results

In California EDs between 2005–2011, there were 43,979 total patients discharged with a diagnosis of Bell's palsy. Median age at diagnosis was 45 years (IQR 32 to 57). A total of 6,224 (14.2%) underwent neuroimaging as part of their ED evaluation; 5,763 (13.2%) had CT alone, 238 (0.6%) had MRI alone, and 223 (0.5%) had both. On 90-day follow-up, 356 patients (0.8%) received one of our predefined serious alternative diagnoses. When restricted to only life-threatening alternative diagnoses associated with central facial paralysis, 127 patients (0.3%) were identified.

Demographic characteristics as well as the frequency of comorbid conditions and ED imaging utilization of the overall cohort stratified by outcome are found in Table 1. Subjects who subsequently received a serious diagnosis other than Bell's palsy had a higher prevalence of comorbidities and were more likely to have had imaging as part of their initial evaluation.

Table 2 shows the results of our multivariate model, which identified multiple factors associated with the likelihood of receiving an alternative diagnosis. Demographic factors or comorbid conditions positively associated included increasing age (HR 1.11, 95% CI 1.01–1.21, for every 10 years), black as compared to white race (HR 1.68, 95% CI 1.13–2.48), and diabetes mellitus (HR 1.46, 95% CI 1.10–1.95). The use of CT or MRI imaging on the index visit was also associated with an increase in the misdiagnosis of Bell's palsy (HR 1.43, 95% CI 1.10–1.85). Private insurance, as compared to Medicare, was negatively associated with an alternative diagnosis (HR 0.65, 95% CI 0.46–0.93).

The alternative diagnoses made on follow-up are listed in Table 3, stratified by 7-, 14-, 30-, and 90-day follow-up. Of the 356 individuals with an alternative diagnosis within 90 days,

142 (39.9%) were within 7 days. Most common, at all four time intervals, were ischemic stroke, herpes zoster, GBS, and otitis media, accounting for 85.4% of all alternative diagnoses.

Limitations

This study was done as a secondary analysis of a large administrative dataset, and therefore suffers from many limitations inherent to such retrospective cohort studies. Most importantly, some relevant clinical factors such as details of the patient's history, neurological examination, or concurrent medical complaints are not assessed in this study. Additionally, the dataset does not include information on patient acuity, an important confounding variable. In particular, the positive association between CT or MRI use and our outcome may be related to this unmeasured confounding as sicker or more complex patients may be more likely to both receive acute imaging and an alternative diagnosis on follow-up.

Our outcome is the diagnosis of one or more a priori decided conditions within 90 days of the original ED diagnosis of Bell's palsy. We assume that this second diagnosis is the correct etiology of the facial paralysis mistaken for Bell's palsy on the first visit. Certainly, two independent conditions may co-exist in a relatively short time period, and the original diagnosis of Bell's palsy may have been correct. However, well over half of the alternative diagnoses were made within 14 days of the Bell's palsy diagnosis, suggesting that the alternative diagnosis accounted for the initial presentation. Alternatively, it is possible that a misdiagnosis of Bell's palsy went unrecognized during the 90-day follow-up period or indefinitely. These two misclassifications would have countervailing effects on the true rate of misdiagnosis.

Finally, our outcome diagnosis is assessed either on subsequent ED visit or hospitalization, leaving out a potential subgroup of patients who received an alternative diagnosis in an outpatient visit with a primary care physician, otolaryngologist, or neurologist. Likewise, patients who migrated out of California may have been lost to follow-up. As such, the true misdiagnosis rate may be underestimated.

Discussion

This study using administrative data from a large and demographically heterogeneous state from 2005 to 2011 found that emergency providers have a high accuracy for diagnosing Bell's palsy in the ED. A misdiagnosis rate less than 1% appears acceptable when compared to other disorders commonly presenting to the ED. For example, the diagnostic accuracy in evaluation of migraine headaches,[12] acute appendicitis,[13] and pulmonary embolism[14] in acute or ambulatory care settings appear to be lower than that of Bell's palsy in the ED. One study found that emergency providers frequently misdiagnosed neurocardiogenic syncope, peripheral vertigo, or primary headaches, among others, labeling them incorrectly as stroke or seizure.[15] Overall, the study suggested that misdiagnosis or diagnostic uncertainty was present in 36% of neurologic consultations in the ED. Few studies have evaluated the accuracy of diagnosing relatively benign conditions; conversely, most research has focused on missing serious diseases among common presenting complaints. For instance, among patients with headache, misdiagnosis rates are 5% for subarachnoid hemorrhage, a highly deadly condition.[16] Similarly, for other dangerous neurologic conditions, higher rates of incorrect diagnoses have been reported for central vertigo,[17,18] spinal cord compression,[19] transient ischemic attack,[20] GBS,[21] and seizures.[22]

The accuracy of diagnosis in Bell's palsy is likely related to its distinctive and specific presentation. However, facial weakness is a cardinal symptom of stroke[23] and in some cases it may be difficult to differentiate peripheral versus central facial nerve weakness. We found

that more than one-quarter of the Bell's palsy misdiagnoses were ischemic stroke. Cases of strokes mimicking Bell's palsy have been reported in the literature.[24,25] Infarcts of the dorsal pons may cause ischemia to the motor component of the facial nerve resulting in isolated ipsilateral palsy including the forehead. Additionally, a radiologic analysis of stroke syndromes further challenges the dogma that dual facial innervation leads to sparing of the upper face with upper motor neuron lesions.[26] With these caveats in mind, detailed history and neurological examination is typically sufficient to evaluate unilateral facial paralysis and rule in or out Bell's palsy in the vast majority of cases.

Certainly, not all misdiagnoses of serious conditions are the same – a missed subarachnoid hemorrhage may have profoundly different consequences than a missed initial presentation of HIV infection. Our analysis included a relatively wide range of important clinical entities in order to best evaluate the diagnostic accuracy of emergency providers. However, the significance of labeling entities such as Lyme disease or herpes zoster as "idiopathic" and delaying the definitive diagnosis is unclear. In reality, the correct diagnosis may not be apparent or accessible to the emergency provider during the index visit. Other alternative diagnoses, such as otitis media, are of questionable causal relevance if made weeks or months after onset of facial paralysis. When limiting our outcome to only potentially catastrophic diagnoses such as stroke, hemorrhage, tumor, or CNS infection, the misdiagnosis rate is even lower at 0.3%, providing further reassurance that emergency providers' performance in diagnosing Bell's palsy is adequate and appropriate.

Interestingly, we found that 10.9% of patients misdiagnosed with Bell's palsy went on to receive a diagnosis of GBS on 90-day follow-up. GBS encompasses multiple heterogeneous immune-mediated neuropathies, generally presenting with progressive and symmetric motor weakness and loss of deep tendon reflexes. Multiple variants of GBS exist, with acute inflammatory demyelinating polyneuropathy, the classic form with ascending paralysis, accounting for the majority of cases.[27] Variants of GBS may involve the cranial nerves, including Miller Fisher syndrome, typically presenting with ophthalmoplegia, ataxia, and areflexia with subsequent descending paralysis. Miller Fisher syndrome accounts for approximately 5% of all cases of GBS and may progress in half of cases to cause various other cranial nerve abnormalities.[28,29] Additional variants of GBS that may overlap with Miller Fisher syndrome and initially present with facial symptoms include Bickerstaff encephalitis and pharyngeal-cervical-brachial weakness, though both of these entities may have multiple associated symptoms that would generally preclude a diagnosis of Bell's palsy.[30,31] More detailed research may be warranted to investigate the association between ED misdiagnosis of Bell's palsy and subsequent diagnosis of GBS.

We also found that the use of CT or MRI imaging was positively associated with a subsequent diagnosis other than Bell's palsy. This finding is likely due to unmeasured confounding by indication due to differences in the acuity or complexity of patients or their presentation. In practice, imaging should be reserved for cases in which the history and physical examination findings raise concern for an upper motor neuron facial nerve paralysis or when there are findings beyond isolated cranial nerve seven involvement. Our results suggest that particular attention should be paid to older patients with diabetes, both of which are commonly associated with vascular complications; therefore, facial paralysis in these patients should raise more concern for stroke than Bell's palsy. However, the pathophysiology of Bell's palsy may also involve small vessel ischemia to the facial nerve. In fact, previous research has shown that cardiovascular risk factors, particularly hypertension, are significantly more prevalent in Bell's palsy as compared to patients with peripheral facial paralysis of other etiologies.[32] We postulate that cardiovascular risks, while potentially related to development of Bell's palsy, have even stronger associations with other diagnoses such as stroke, intracranial hemorrhage, or subarachnoid hemorrhage.

Conclusion

Emergency providers adequately diagnose Bell's palsy in the ED with a low rate of missing serious or life-threatening alternate diagnoses based on subsequent 90-day follow-up. Careful history and physical examination, with rational use of imaging, remains the mainstay of ED evaluation of patients with isolated peripheral facial nerve weakness. Increasing age and diabetes mellitus are modest risk factors for misdiagnosis of Bell's palsy.

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Table 1

Demographic, comorbid, and imaging use characteristics of patients with an ED diagnosis of Bell's palsy in California, 2005–2011, stratified by 90–day alternative diagnosis.

	No alternative diagnosis made n = 43,623		Alternative diagnosis made n = 356	
	No.	**% (95% CI)**	**No.**	**% (95% CI)**
Age quartile				
18–31	10,890	25.0 (24.6–25.4)	70	19.7 (15.9–24.1)
32–44	10,748	24.6 (24.2–25.0)	69	19.4 (15.6–23.8)
45–56	11,339	26.0 (25.6–26.4)	93	26.1 (21.8–30.9)
≥ 57	10,646	24.4 (24.0–24.8)	124	34.8 (30.1–39.9)
Sex				
Male	21,280	48.8 (48.3–49.2)	166	46.6 (41.5–51.8)
Female	22,343	51.2 (50.8–51.7)	190	53.4 (48.2–58.5)
Race				
White	15,164	38.4 (38.0–38.9)	140	41.1 (36.0–46.4)
Black	2,545	6.45 (6.21–6.70)	38	11.1 (8.1–15.1)
Hispanic	17,900	45.4 (44.9–45.9)	134	39.3 (34.3–44.6)
Asian	2,492	6.32 (6.08–6.56)	18	5.28 (3.37–8.19)
Other	1,341	3.40 (3.23–3.58)	11	3.23 (1.81–5.69)
Insurance status				
Medicare	6,845	15.7 (15.4–16.0)	92	25.8 (21.6–30.6)
Medicaid	6,123	14.0 (13.7–14.4)	69	19.4 (15.6–23.8)
Private	19,502	44.7 (44.2–45.2)	117	32.9 (28.2–37.9)
Self–pay	8,725	20.0 (19.6–20.4)	57	16.0 (12.6–20.2)
Other	2,415	5.54 (5.33–5.76)	21	5.90 (3.89–8.85)
Visit day				
Weekday	30,842	70.7 (70.3–71.1)	245	68.8 (63.8–73.4)
Weekend	12,781	29.3 (28.9–29.7)	111	31.2 (26.6–36.2)
Comorbidity				
Hypertension	8,439	19.3 (19.0–19.7)	105	29.5 (25.0–34.4)

	No alternative diagnosis made n = 43,623		Alternative diagnosis made n = 356	
	No.	% (95% CI)	No.	% (95% CI)
Diabetes Mellitus	5,169	11.8 (11.6–12.1)	73	20.5 (16.6–25.0)
Coronary heart disease	666	1.53 (1.42–1.65)	6	1.69 (0.78–3.63)
Congestive heart failure	170	0.39 (0.34–0.45)	1	0.28 (0.05–1.57)
Chronic kidney disease	281	0.64 (0.57–0.72)	5	1.40 (0.60–3.24)
Chronic obstructive pulmonary disease	250	0.57 (0.50–0.65)	4	1.12 (0.44–2.85)
Imaging				
CT and/or MRI	6,149	14.1 (13.8–14.4)	75	21.1 (17.2–25.6)

Table 2

Multivariate associations between demographics/comorbidities and alternative diagnosis made on 90-day follow-up for patients with an initial ED diagnosis of Bell's palsy.

	HR (95% CI)
Age, every 10 years	1.11 (1.01–1.21)
Female sex	1.02 (0.80–1.29)
Race	
White	Ref
Black	1.68 (1.13–2.48)
Hispanic	0.82 (0.62–1.07)
Asian	0.75 (0.45–1.27)
Other	0.90 (0.50–1.64)
Insurance status	
Medicare	Ref
Medicaid	1.26 (0.85–1.87)
Private	0.65 (0.46–0.93)
Self-pay	0.77 (0.52–1.15)
Other	0.84 (0.46–1.55)
Weekend	1.12 (0.89–1.41)
Comorbid conditions	
Hypertension	1.22 (0.94–1.59)
Diabetes	1.46 (1.10–1.95)
Coronary heart disease	0.64 (0.30–1.38)
Congestive heart failure	0.36 (0.49–2.58)
Chronic kidney disease	1.33 (0.54–3.29)
Chronic obstructive pulmonary disease	1.23 (0.45–3.36)
Imaging use (CT and/or MRI)	1.43 (1.10–1.85)

Table 3

Cumulative counts of alternative diagnoses made on 7, 14, 30, and 90-day follow-up for 356 patients* with an initial ED diagnosis of Bell's palsy.

	7-day	14-day	30-day	90-day	
Diagnosis	**No.**	**No.**	**No.**	**No.**	**%**
Ischemic stroke	53	62	77	98	27.3
Intracranial hemorrhage	4	4	5	10	2.8
Subarachnoid hemorrhage	0	0	0	2	0.6
Brain tumor	0	1	2	4	1.1
Central nervous system infection	4	5	7	13	3.6
Human immunodeficiency virus infection	3	6	7	19	5.3
Guillain-Barre syndrome	23	30	37	39	10.9
Lyme disease	1	0	0	3	0.8
Otitis media or mastoiditis	20	38	56	86	24.0
Herpes zoster	34	57	69	83	23.2
Total	142	204	261	358	

* Two patients received more than one alternative diagnosis.