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## Comparison of Fixed Cell-based Assay to Radioimmunoprecipitation Assay for Acetylcholine Receptor Antibody Detection in Myasthenia Gravis

Ario Mirian, Michael Nicolle, Adrian Budhram

### Objective

To compare specificity and sensitivity of a commercially available fixed cell-based assay (F-CBA) to radioimmunoprecipitation assay (RIPA) for acetylcholine receptor antibody (anti-AChR) detection in myasthenia gravis (MG).

### Background

Approximately 50% of ocular and 85% of generalized MG are anti-AChR positive by RIPA, the current gold standard test. Clustered live cell-based assay (L-CBA) can detect low-affinity anti-AChR that are missed by RIPA, but the costly and time-consuming nature of L-CBA has restricted its use to specialized centres. A commercial F-CBA has become available for anti-AChR detection, but its diagnostic performance compared to RIPA requires evaluation.

### Design/Methods

In this retrospective diagnostic cohort study we reviewed the clinical information of suspected MG patients evaluated at London Health Sciences Centre MG clinic, who were clinically classified as MG or non-MG and who had anti-AChR RIPA and then F-CBA performed. Classification of each patient as anti-AChR F-CBA-negative/positive, RIPA-negative/positive, and MG/non-MG permitted specificity and sensitivity calculations for each assay.

### Results

Six-hundred-eighteen patients were included in study analysis. The median patient age at time of sample collection was 45.8 years (range: 7.5–87.5 years) and 312/618 (50.5%) were female. Of 618 patients, 395 (63.9%) were classified as MG. Specificity of both F-CBA and RIPA was excellent (99.6% vs. 100%,  $P > 0.99$ ). One F-CBA-positive patient was classified as non-MG, although in retrospect ocular MG with functional overlay was challenging to exclude. Sensitivity of F-CBA was significantly higher than RIPA (76.7% vs. 72.7%,  $P = 0.002$ ). Overall, 20/97 (21%) otherwise SNMG patients after RIPA evaluation had anti-AChR detected by F-CBA.

### Conclusions

In our study anti-AChR F-CBA and RIPA both had excellent specificity, while F-CBA had 4% higher sensitivity for MG and detected anti-AChR in 21% of SNMG patients. Our findings indicate that F-CBA is a viable alternative to RIPA for anti-AChR detection.

**Disclosure:** Dr. Mirian has nothing to disclose. Dr. Nicolle has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Alexion. Dr. Budhram has nothing to disclose.

## Piloting an Advanced Neuroimmunology Elective for Neurology Residents

Sonia Kaur Singh, Rohini Samudralwar

### Objective

To describe the creation of an Advanced Neuroimmunology elective for residents with a special interest in clinical neuroimmunology.

### Background

There has been a dramatic change in the landscape of neuroimmune conditions with the discovery of new pathogenic autoantibodies, disease modifying therapies and wider availability of multidisciplinary care systems for patients. Most residencies do offer exposure to multiple sclerosis but with increasing interest in neuroimmunology and autoimmune neurological conditions, there is a gap in resident education to meet needs of this changing landscape.

### Design/Methods

A curriculum for advanced neuroimmunology (NI) was developed for residents with special interest in clinical neuroimmunology. This two-week elective consisted of rotations through NI and affiliated multidisciplinary clinics to increase exposure to immune mediated neurological illnesses, appreciate their heterogeneity, and aid multidisciplinary approach. Department experts in various disease states related to neuroimmunology were contacted and based on interest and resident elective time, a schedule was set up for rotations through neuroinfectious diseases, pulmonary sarcoidosis clinic, neuro-oncology, neuropathology and rheumatology. An additional expectation was to work with the fellow on inpatient consults that came in through the 2 weeks. In addition to multiple sclerosis/neuroimmunology division didactics, residents are encouraged to attend other affiliated department conferences as well as present at interdepartmental meetings, such as neuro-rheumatology conference.

### Results

The availability of this elective allowed increased exposure to neuroimmunological conditions outside the typical Multiple Sclerosis elective at UTHHealth. It also has allowed for additional inter-departmental collaboration clinically. Since the initial pilot elective, more residents have requested this as an elective and will be surveyed about their experience.

### Conclusions

There is an unmet need for MS and NI subspecialists. Exposure to the broad spectrum of neuroimmunological conditions through multidisciplinary collaborations during residency is instrumental to ensure future specialists have the foundations to adapt to this rapidly advancing field.

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## A Case of Pembrolizumab (Anti-PD-1) Induced Encephalitis

Anza Zahid, Meryim Poursheykhi, Mujtaba Saeed, Ivo Tremont

### Objective

N/A.

### Background

PD-1 Immune checkpoint inhibitors (ICI) have been associated with neurologic immune-related adverse events including meningoencephalitis and limbic encephalitis that can manifest as paraneoplastic syndromes. We present a case of suspected pembrolizumab (anti PD-1) induced limbic encephalitis presenting as episodic aphasia.

### Design/Methods

N/A.

## Results

A 62-year-old man with poorly differentiated papillary thyroid carcinoma with extensive brain, lung, lymphatic metastases post thyroidectomy, radioiodine therapy, chemotherapy with dabrafenib and trametinib, and stereotactic radiation surgery presented with recurrence of brain metastases one year after the diagnosis of a 2.1 cm metastatic lesion in the left parieto-occipital region causing right homonymous hemianopsia. The metastases initially improved in size and number with radiation and serial brain MRIs were stable. Biopsy of the mets revealed a poorly differentiated carcinoma. He was subsequently treated with pembrolizumab for six months. Two months after treatment initiation, he reported episodic behavioral arrest, confusion, and expressive aphasia concerning for seizures. Continuous EEG monitoring revealed a left-sided focus without seizures, and the episodes persisted despite levetiracetam and clobazam. Four lumbar punctures revealed lymphocytic pleocytosis (10–14 cells), elevated protein (48–68 mg/dl), negative cytology, flow cytometry and viral studies including JC virus. Paraneoplastic panels in serum and cerebrospinal fluid were negative. Repeat MRIs findings were most consistent with radionecrosis and noted improvement of the metastatic lesions. Suspicion was raised for pembrolizumab-induced encephalitis and he received high-dose steroids with minimal response however, clinical improvement noted with reduced episode frequency after intravenous immunoglobulin induction therapy and rituximab maintenance therapy.

## Conclusions

PD-1 ICI-related encephalitis is a diagnosis of exclusion that should be considered in patients with encephalopathy or other neurological deficits following 3 months of treatment initiation and response to immunosuppressive therapy. Higher incidences are reported in males. Early recognition is crucial to prevent long-term neurologic damage. Outcomes are depend on patient characteristics and clinical presentation.

**Disclosure:** Dr. Zahid has nothing to disclose. Dr. Poursheykhi has nothing to disclose. Dr. Saeed has nothing to disclose. Dr. Tremont has nothing to disclose.

## Neuroblastoma Presentation With Multiple Cranial Nerve Involvement

Aysha Arshad, Janetta Arellano, Anastasia Chumakova, Sharief Taraman

### Objective

NA.

### Background

We report a case of neuroblastoma, a pediatric neuroendocrine tumor of the sympathetic nervous system, in a 3-year-old female with multiple cranial nerve involvement.

### Design/Methods

A 3-year-old afebrile, lethargic female presented with bilateral eyelid droop, right head tilt, slurred speech, gagging, abnormal walking and no bowel movement. Neurological examination noted bilateral ptosis, dysarthria, left tongue deviation, proximal weakness in upper and lower extremities, areflexia in biceps and patellar tendons, dysmetria, and wide-based gait. MRI of the brain showed heterogeneous appearance of the clivus and MRI of the spine showed right adrenal mass and heterogeneous enhancement of multiple vertebrae, suggesting possible metastatic disease. Serum and cerebrospinal studies were unremarkable. Patient was treated with intravenous methylprednisolone and plasmapheresis for suspected paraneoplastic syndrome; however, she continued to clinically progress. Adrenal mass biopsy results and elevated urine VMA and HVA levels were consistent with the diagnosis of neuroblastoma. Nuclear imaging and meta-iodobenzylguanidine scan were negative. Paraneoplastic panels and Lambert Eaton panel were negative for autoantibodies. Total resection of

the abdominal mass and right adrenal gland with continued steroid taper, resulted in reported near total symptom resolution.

## Results

Our patient presented with antibody negative paraneoplastic polyneuropathy due to neuroblastoma. Cranial nerve involvement in neuroblastoma can result from tumor involvement of the sympathetic chain, paraneoplastic syndromes, or metastasis to the skull. Our patient's initial imaging suggested potential metastatic or inflammatory involvement of the clivus, suspected of causing her cranial nerve symptoms. However, nuclear studies were negative for metastatic disease and patient's symptoms resolved after resection of the main mass suggesting a paraneoplastic etiology. Although her paraneoplastic panel and Lambert-Eaton panel were negative for common associated antibodies, it is likely an unmeasured autoantibody, cytokine, hormones, or peptide was at play.

## Conclusions

Neuroblastoma should be considered as a differential for a neurological presentation involving multiple cranial nerves in a child.

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## Co-Occurring NMDA-Receptor and Anti-GAD65 Antibodies in the CSF of a Patient With Encephalitis: Case Report

Caleb McEntire, Giovanna Manzano, Jenny Linnola

### Objective

We describe a 58-year-old woman who presented with rapid cognitive changes and was found to have concurrent CSF NMDA and GAD65 receptor antibodies.

### Background

Antibodies against NMDA and GAD65 receptors are associated with highly morbid autoimmune encephalitides. One case of co-occurring NMDA-R and anti-GAD65 antibodies in a patient with progressive cognitive changes and type I diabetes has previously been described. Herein, we describe a case of a previously high-functioning woman who experienced rapidly progressive cognitive changes secondary to autoimmune encephalitis (AE) with co-existent NMDAR and GAD65 antibodies.

### Design/Methods

NA.

## Results

A 58-year-old woman with adult-onset insulin-dependent diabetes mellitus, hypertension, and prior left-sided Bell's palsy presented to medical care for subacute cognitive decline characterized initially by inattention and difficulty with activities of daily living, progressing to profound global aphasia and seizures. Diagnostic testing revealed GAD65 antibody positivity in serum (33 nmol/L) and CSF (17.3 nmol/L) and negative serum but positive CSF NMDAR antibody. CSF showed pleocytosis (37 nucleated cells, 97% lymphocytes), elevated glucose (109 mg/dL), and normal protein (41 mg/dL). EEG showed right temporal epileptiform discharges. MRI was unrevealing. She was treated with IV steroids, IVIg, and rituximab, and has slowly improved on follow-up.

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Anza Zahid, Meryim Poursheykhi, Mujtaba Saeed, et al.

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