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Published In/Presented At

Moran, J. Prousi, G. Smith, S. (2017, October 28). *AMPA-R Antibody Positive Autoimmune Encephalitis: An Under Recognized Cause of Acute Mental Status Changes*. Poster Presented at: American College of Physicians (ACP), Harrisburg, PA.

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AMPA-R Antibody Positive Autoimmune Encephalitis: An Under Recognized Cause of Acute Mental Status Changes

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INTRODUCTION

- Considered to be a rare culprit of acute mental status changes, autoimmune encephalitis accounts for only 4% of encephalitis cases¹
- The presentation of Anti-AMPA-R encephalitis is acute in nature and symptomatology consists of psychiatric symptoms, behavioral abnormalities and movement disorders
- Women over the age of 50 with known autoimmune disorders have been most commonly affected
- At present, 58% of women and 23% of men who develop this pathologic process are found to have an underlying malignancy²

CASE PRESENTATION

- A 60 year old female with a history of rheumatoid arthritis and depression presented with a one-month history of altered mental status, abnormal behavior and unsteady gait
- Presenting to an outside hospital, she underwent metabolic, infectious, and neuroimaging studies that were unremarkable. It was suspected that she had either a rapidly progressive dementia or a brief psychotic disorder triggered by severe depression
- Her mental status did not improve over a 15-day hospital course and she was transferred to our facility. On presentation the patient was not oriented to person place or time, was writhing, moaning and completely non verbal.
- Differentials included CJD, Vasculitis, and paraneoplastic / autoimmune encephalitis. Diagnostic studies returned showing positivity for Anti-AMPA-R antibodies, indicating the patient had a rare form of autoimmune encephalitis. Extensive work up for underlying malignancy was negative
- The patient made significant improvement with plasma exchange therapy, cyclophosphamide and steroids. Her mental status and behavior returned to baseline and she was discharged in stable condition

TREATMENT

- Treatment is largely based on the evaluation of case reports and case series with the majority of approaches revolving around combined therapies.
- Immunotherapy with either corticosteroids alone or in conjunction with IV immunoglobulin or plasma exchange is the mainstay of therapy^{4,5}
- Second line therapies with rituximab, cyclophosphamide, or combined approach are possibilities for those who fail to improve within 4 weeks of immunotherapy^{4,5}
- It should also be noted that given a significant association with primary tumor diagnoses, tumor removal is suggested and has lead to a reduction of antibody levels^{4,5}

DISCUSSION

- This case in combination with review of several case series in literature suggests that the detection of AMPA-R antibodies be considered in patients, particularly women, older than 50 years who present not only with limbic encephalitis but also with rapidly progressive abnormal behaviors resembling acute psychosis¹
- This patient's history of a previously diagnosed autoimmune disease should also have prompted consideration. Literature suggests that up to 50% of patients diagnosed with AMPA-R Ab positive autoimmune encephalitis had history of another systemic autoimmune disease²
- Early detection is crucial due to the high association AMPA-R Ab positive autoimmune encephalitis has with underlying malignancy. An underlying tumor was discovered in 64% of patients in a case series of 22 patients²
- Patients without malignancy tend to respond very well to immunotherapy; however, close follow up is indicated due to a relapse rate of up to 50% and diagnosis of a primary tumor has occurred during relapses^{2,3}
- Overall, the prognosis for this patient is very favorable with literature showing a 100% 5 year survival in patients without underlying malignancy compared with a 50% 5 year survival in those with underlying malignancy²

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