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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

REFERENCES:

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 therapy4,5
• Second line therapies with rituximab, cyclophosphamide, or combined approach are possibilities for those who fail to improve within 4 weeks of immunotherapy4,5
• 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 levels4,5

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 relapses2,3
• 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