Positive Antibodies in Atypical Dementia: A Case Report Exploring the Intricacies of Diagnosing Rapid Cognitive Decline

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INTRODUCTION

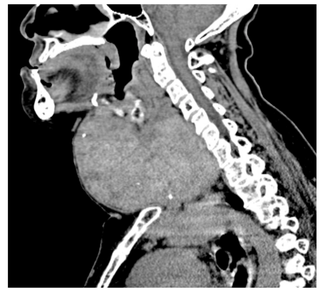
Autoimmune encephalitis (AIE) can be extremely difficult to differentiate from general dementia syndromes and often results in a time-intensive and costly diagnostic dilemma.

CASE DESCRIPTION

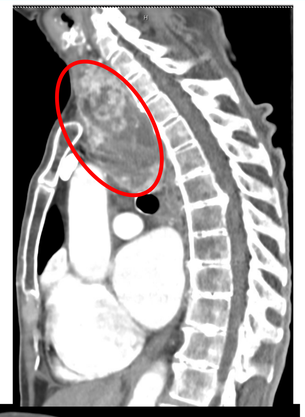
A 70-year-old woman with PMH of atrial fibrillation, thyroid disease, and recent illness with Covid-19 was brought in by her husband for weeks of worsening mental status with personality change, severe agitation, rapid cognitive decline, mania and psychosis.

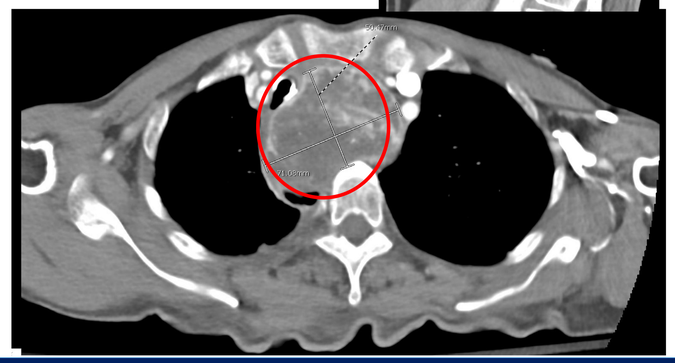
Workup for strongly suspected AIE was initiated, including lumbar puncture, paraneoplastic/AIE CSF panel, serum studies, pan-CT, brain MRI w/&w/o contrast, and alternate causes workup including infectious, thyroid, toxicities, and vitamin deficiencies. The patient was immediately started on high-dose methylprednisolone for empiric treatment of AIE given months prior workup from outside hospital had positive serum titer for AChR Ab. The CL Psychiatry team was consulted for severe agitation management requiring restraints—she had paranoid and grandiose delusions, severe cognitive impairment, and delirium, and her MoCA Test was 17/30. She was placed on valproic acid, lorazepam at bedtime. After steroid course she received intravenous immune globulin (IVIG) with minimal improvement in symptoms. Three weeks later, the send-out panel CSF returned, but her CSF was negative for any paraneoplastic/autoimmune antibodies for AIE and she never had elevated inflammatory markers. Her mediastinal mass was ultimately thought to be a large goiter that had been present for over 15 years, and family declined biopsy.

IMAGING



"Normal Goiter"





TIMELINE

July 2022: Covid-19 infection

August 2022: Symptom onset: personality change, forgetfulness

September 2022: Outside hospitalization:
- Mediastinal mass discovered (known to patient)
- Serum autoimmune encephalitis panel showed AChR Ab postive

October 2022: Psychiatric hospitalization, started on valproic acid, quetiapine
*AChR antibody came back positive, however patient lost to follow-up

October 2022-January 2023: Continued to exhibit psychosis and manic symptoms at home

February 2023: admitted to Dell Seton Medical Center, Psych Consult Liaison team consulted for severe agitation

February 2023: empirically treated for AIE with steroids, agitation managed with valproic acid, lorazepam

March 2023: CSF send-out encephalitis panel resulted - *negative*

DISCUSSION

Due to the lack of clear laboratory evidence for AIE and lack of empiric treatment response, the patient was diagnosed with **atypical dementia**.

Factors contributing to initial concern for AIE:
- Patient-specific: presence of autoantibodies in serum panel, atypical rapid decline
- Historical factors: limited collateral
- Systemic factors: fragmented healthcare system, delay in encephalitis panel results

CONCLUSIONS

There have been numerous case reports describing how AIE can mimic dementia, but this case provides an example of presumed AIE that was eventually excluded in favor of an atypical dementia syndrome.

Take-home points:

1. **Presence of red flags for AIE warrants a thorough investigation**
2. **Beware of anchoring bias**
3. **Empiric treatment for AIE may further complicate the presentation but should not be delayed**

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