Background

• Posterior Cortical Atrophy (PCA) is a neurodegenerative syndrome that changes one’s ability to process visuospatial information. It typically presents with changes to visuospatial skills and progresses to more global cognitive decline.1

• Until recently, there was no classification framework for PCA, which has limited not only diagnosis and treatment, but also research endeavors. Given that there are no established instruments to guide the assessment of PCA symptoms, the syndrome is often evaluated qualitatively, delaying diagnosis.2

• The aim of this project is two-fold: 1. to conduct a comprehensive literature review to create a simplified and consolidated resource to help primary care providers recognize the signs of PCA; and 2. determine the best clinical tools for PCA diagnosis by identifying what tests are currently being used by clinicians and researchers around the world.

Methods

Part 1 – Literature Review

Systematic literature review carried out on PubMed, Google Scholar, and JAMA Network using the following search terms: ‘posterior cortical atrophy,’ ‘Alzheimer’s disease variant,’ and ‘atypical Alzheimer’s disease.’ Publications from June 2002 to July 2022 were included and came from American and international publications. References of relevant publications were screened to ensure all appropriate publications were included.

Part 2 – Survey

A 23-question REDCap survey was sent out to members of the Alzheimer’s disease Professional Interest Area. The survey consisted of four sections: A. establishing the role and clinical experience of the survey participant; B. reporting on tests used in diagnosis and/or assessment of PCA and the frequency of their use; C. reporting on tests used in diagnosis and/or assessment of features that are not included in the Core PCA Cognitive Features; and D. reporting on preferred global measures for following patients over time.

The results of the survey were then analyzed to identify the testing methodologies being currently used. For the purposes of this project, analysis of the survey focused on the most commonly used metrics, specifically from the clinicians who replied to the survey. A cutoff of 30% was determined to identify the “most used clinical tools”; tools that were used by less than 30% of clinicians were not included in the analysis.

COMIRB approval was obtained prior to sending out the survey (protocol number: 20-1045).

Results

Survey - Results

- The survey was sent out to experts in the field. Analysis identified 23 separate tests as being commonly used for PCA assessment.

- From here, a battery of tests can be created that could be used to screen and diagnose effectiveness in diagnosing PCA. By having a comprehensive set of tools, future studies can be facilitated, as this creates a comprehensive and objective measure for data collection and diagnosis.

- A provider education resource summarizing the ‘must-know’ points regarding PCA was created to help primary care providers better identify the syndrome in their patients.

Conclusions

- PCA is a complex syndrome with a poorly understood pathophysiology. Until recently, there was no agreed-upon classification system, which limited diagnosis and treatment options.3 An additional roadblock identified in current literature was the lack of standardized clinical instruments that could be used to screen and diagnose patients.

- Based on this gap in information, a survey was sent out to experts in the field. Analysis identified 12 separate tests as being commonly used for PCA assessment.

- From here, a battery of tests can be created that could be distributed and tested for effectiveness in diagnosing PCA. By having a comprehensive set of tools, future studies can be facilitated, as this creates a comprehensive and objective measure for data collection and diagnosis.

- A provider education resource summarizing the ‘must-know’ points regarding PCA was created to help primary care providers better identify the syndrome in their patients.

Limitations and Implications

- Limitations: 1. The survey was sent out to a limited sample of clinicians and researchers; 2. It is possible that there are clinicians who have a favorite set of tests that they wish to continue using, despite an agreed-upon standard. This would defeat the purpose of standardization and could lead to missed (or false positive) diagnoses.

- 3. A battery of tests does little good if patients do not have access to care, or if clinicians do not know how to look for PCA in the first place.

- Implications: Though there has been an upward trend in literature for PCA in the last decade, the syndrome continues to be underdiagnosed.

- Educating primary care providers on PCA as well as creating a standardized and accessible battery of tests would facilitate diagnosis, treatment, and research, improving quality of life for patients.

Acknowledgements

I would like to thank Dr. Steven Lowenstein for his mentorship and guidance in solidifying a direction for the overarching aims of the project. I would like to thank Dr. Victoria Pelak for her mentorship on this project as well as her expertise in the field and Ms. Jerri Lusk for all her help in facilitating meetings and communications.

Disclosures

Elizaveta Orysheva has no conflicts of interest to disclose.

References

1 Schott JM, Crutch SJ. Posterior Cortical Atrophy. Continuum (Minneap Minn). 2019 Feb;25(1):183-205. PMID: 30376747