Diagnosing and Rehabilitating a Rare Case of Cerebral Amyloidosis

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Introduction

A 76 year old F with a history of dementia who presented to the hospital after months of progressive autonomic dysfunction, mental status decline, bladder incontinence, shuffling gait, and weakness.

Initially, weakness involved the left side, but progressed to the right side as well. EEG was inconclusive. MRI of brain showed no evidence of hydrocephalus and revealed persistent non suppression of FLAIR in right parietal and frontal sulci with some focal leptomeningeal enhancement suggesting meningeal involvement.

Background

Cerebral amyloidosis is most often silent. When symptomatic it most commonly presents with spontaneous lobar hemorrhage. However, there are cases of cerebral amyloidosis which fall in between these two most common presentations. In these cases, a wide variety of confounding symptoms can make diagnosis difficult.

Most commonly, cerebral amyloidosis presents as a peripheral/autonomic neuropathy. The most common symptoms include numbness, paresthesias, pain, and autonomic symptoms such as bowel/bladder dysfunction and orthostatic hypotension.[1]

CNS involvement is unusual but can lead to extensive cortical pathology and dementia as well as spontaneous intracranial bleeding.[2]

References

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Results

Due to involvement of the leptomeninges in combination with autonomic dysfunction, the diagnosis was narrowed to Cerebral Amyloidosis in the setting of progressive dementia, but leptomeningeal carcinomatosis could not be ruled out especially in the setting of progressive weight loss. Several months later, the patient presented back in the rehab unit and repeat MRI showed the same findings which had remained stable since her initial admission.

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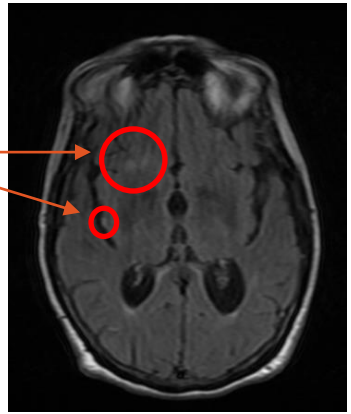
Possibly due to chronic microhemorrhage characteristic of cerebral amyloidosis, which can be seen on T2 weighted MRI[3]. Cortical distribution is more suggestive of cerebral amyloidosis.



Conclusion

With the stability of MRI findings seen during repeat MRI months after initial admission, carcinomatosis became a much less likely diagnosis. While a definitive diagnosis was not achieved due to requiring a cranial biopsy, this is an interesting case of diagnosing a rare and unusual presentation of cerebral amyloidosis, involving both the central and peripheral nervous system (autonomic dysfunction).

Rehabilitation

Upon discharge, patient mobility had improved from 0 to 4 for most mobility categories and her mentation had returned to near baseline. Throughout the admission, the patient maintained high motivation during therapy and made steady progress.

Autonomic and cognitive dysfunction posed unique challenges during inpatient rehabilitation of the patient, requiring close monitoring. Autonomic dysfunction primarily manifested as orthostatic hypotension which was monitored closely and managed pharmacologically. Cognitive dysfunction improved significantly with therapy and patient had returned to her baseline mentation well before discharge.

Despite the challenges illustrated in this case, the potential for improvement is demonstrated clearly. The patient was discharged home after finishing her course of rehabilitation.

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This research was supported (in whole or in part) by HCA and/or an HCA affiliated entity. The views expressed in this publication represent those of the author(s) do not necessarily represent the official views of HCA or any of its affiliated entities.