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Case Report

Intracranial and systemic manifestations of familial leptomeningeal amyloidosis, as seen on CT and MRI

Saralyn Beckius, BS[a,*], *Kamran Shah, MD*[b,c,1]

[a] *Wayne State University School of Medicine, 540 E Canfield St., Detroit, MI 48201, USA*
[b] *Department of Radiology, Wayne State University School of Medicine, 540 E Canfield St., Detroit, MI 48201, USA*
[c] *Department of Radiology DMC Detroit Receiving Hospital 3L-8, 4201 St Antoine St., Detroit, MI 48201, USA*

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ABSTRACT

Leptomeningeal amyloidosis is a subset of familial transthyretin amyloidosis, a family of diseases occurring in conjunction with multiple known mutations of the transthyretin gene. Though this is primarily a disease of the central nervous system, amyloid deposition is multisystemic. We describe a case of a 61-year-old man with known central nervous system amyloidosis presenting to the emergency room with stroke-like symptoms, including left hemineglect, right gaze paresis, and left hemiplegia, atop baseline dementia. A noncontrast CT head demonstrated ventriculomegaly and no acute hemorrhage. Urinalysis indicated an underlying urinary tract infection, ultimately believed to have prompted a breakthrough seizure. Electroencephalogram revealed diffuse encephalopathy. Contrast-enhanced MRI demonstrated hallmarks of intracranial amyloid with no new infarct. Previously taken noncontrast CT neck and thorax demonstrated evidence of systemic disease.

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Background

Amyloidosis is a disease of accumulation, characterized by the extracellular deposition of abnormally folded protein fibrils. It is most commonly multisystemic, with immunoglobulin light chain, chronic inflammatory amyloidosis, and the familial amyloidoses all occurring in multiple organ systems (central nervous system, kidneys, heart, lungs, and gastrointestinal system). Transthyretin-associated amyloidoses (ATTR, occurring in conjunction with abnormal mutation of the *TTR* gene) cause multisystemic disease, typically with predominance of cardiomyopathy or peripheral neuropathy. Leptomeningeal or oculoleptomeningeal amyloidosis, however, is a transthyretin

* Corresponding author.
E-mail addresses: sbeckius@med.wayne.edu (S. Beckius), kshah@dmc.org (K. Shah).
[1] Acting Chairman, Wayne State Department of Radiology; Program Director, DMC Diagnostic Radiology Residency, Neuroradiology, Harper Hospital; Medical School: Chicago Medical School 2001-2005; Residency Training: University of Maryland, Baltimore, MD 2007-2011. Fellowships: Neuroradiology (Northwestern University, Chicago, IL 2011-2012), Medical Informatics (University of Maryland, Baltimore, MD 2010-2011), and Nuclear Medicine (Rush University, Chicago, IL 2006-2007).
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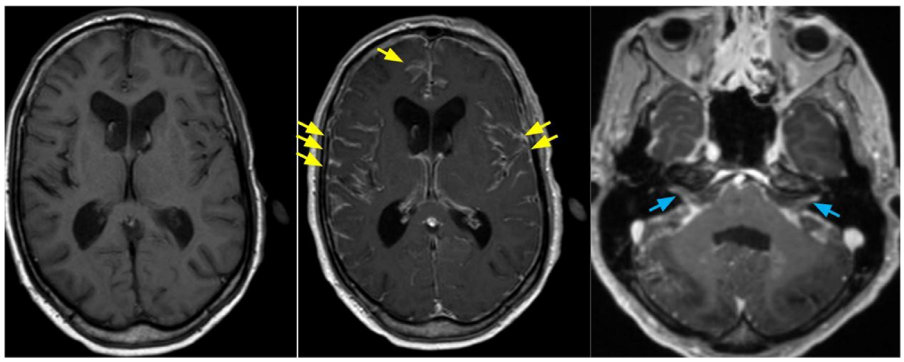

Fig. 1 – T1-weighted brain MRI without (left) and with (right) contrast demonstrate marked leptomeningeal enhancement (yellow arrows) with involvement of the seventh and eighth cranial nerve complexes, particularly on the right (blue arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article).

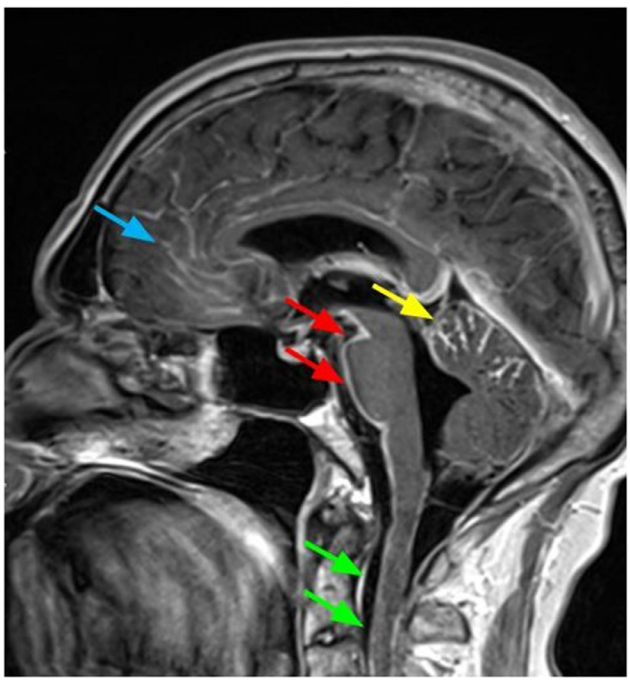

Fig. 2 – T1-weighted brain MRI demonstrates marked leptomeningeal enhancement in supratentorial (blue arrows) and infratentorial (yellow arrows) compartments, as well as along the surface of the brainstem (red arrows) and cervical spinal cord (green arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

amyloidosis with extensive manifestation in the central nervous system. While the disease was initially thought to occur exclusively in European populations, incidence (with the causative mutation, Leu12Pro) has also been documented in patients of African origin [1]. Presentations include dementia, seizure disorders, spasticity, ataxia, and sensorineural hearing loss [2], secondary to amyloid deposition in leptomeningeal vessels. Here, we present a case of a man of European origin with oculoleptomeningeal amyloidosis, whose imaging is

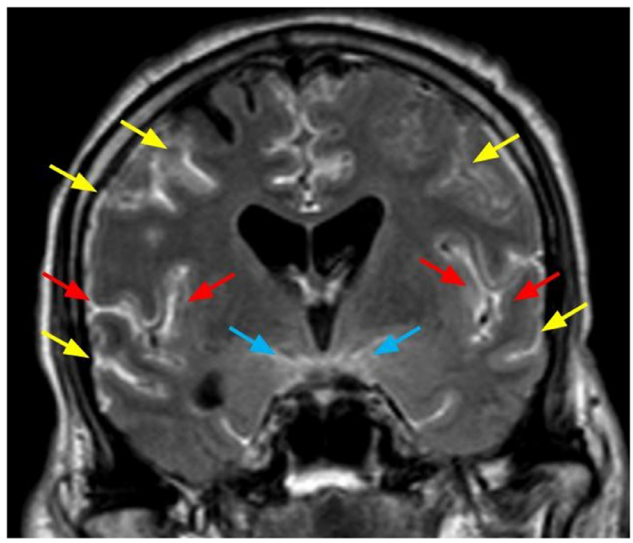

Fig. 3 – Contrast-enhanced coronal FLAIR image demonstrates diffusely abnormal signal and enhancement throughout the sulci (yellow arrows), particularly within the Sylvian fissures (red arrows), as well as within the basal cisterns (blue arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

demonstrative of amyloid deposition in the CNS, as well as in the larynx, trachea, bronchi, and lungs. The following imaging and presentation will allow readers to appreciate both the intra- and extracranial manifestation of amyloidosis.

Case presentation

A 61-year-old man, diagnosed at age 49 in 2004 with leptomeningeal amyloidosis, presented to the emergency room with altered mental status, hemineglect, and gaze paresis. Per his wife, he had been displaying worsening confusion and staring episodes for the past months. Initial noncontrast head

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