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Case study

Clinical pathologic case report: A 70-year-old man with inflammatory cerebral amyloid angiopathy causing headache, cognitive impairment, and aphasia

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ABSTRACT

A 70-year-old man presented with two months of worsening cognitive impairment, hallucinations, and difficulty speaking, with superimposed headaches. Cerebrospinal fluid analysis was notable for lymphocytic pleocytosis and elevated protein. Imaging studies revealed multiple acute and subacute infarcts with cortical microhemorrhages. The patient underwent a stereotactic brain biopsy. In this article, we discuss the patient's differential diagnosis, pathologic findings, ultimate diagnosis, and clinical outcome.

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1. Case description

A 70-year-old man with diabetes mellitus, hypertension, and hyperlipidemia was in his usual state of health when he developed generalized malaise for one week followed by left retro-orbital headache with associated nausea and vomiting. He attended urgent care and was treated with intravenous analgesics with relief. A week later, he noticed word-finding difficulties, left-sided vision loss and inability to function at his job. He presented to a hospital, where a computed tomographic (CT) scan of his head was normal, and he was discharged home. Two days later, he re-presented and was admitted with new onset speech arrest and visual hallucinations. He had no history of fevers. Brain magnetic resonance imaging (MRI) was reportedly abnormal, but this was not thought to fully explain his clinical decline. He had an extensive evaluation, and was diagnosed with a viral meningoencephalitis and treated with IV acyclovir until the CSF HSV PCR returned negative. Over the course of two weeks his clinical status improved. He was discharged to a rehabilitation facility. One week later, he was readmitted after a fall with head strike in the setting of ongoing agitation and hallucinations. Repeat Brain MRI and lumbar puncture were again abnormal. He was transferred to our hospital for further evaluation and management.

On admission to our hospital his neurologic examination was abnormal. He intermittently picked at the air. He had a paucity of speech and impaired word retrieval. He was oriented to year, but not to month or hospital. He perseverated and was inattentive. His long-term memory was intact when speaking about his prior employment. Visual fields and extraocular movements were intact but optic ataxia was present. Tone, power, and reflexes were normal bilaterally. Plantar reflexes were flexor.

Throughout his hospitalization, his neurological and mental status examinations waxed and waned. At his best, he was attentive, fully oriented and demonstrated moderate insight into his care and treatment plan. Episodically, he experienced acute agitation, hallucinations, expressive aphasia, and right-sided facial weakness that resolved within hours.

2. Results

Laboratory testing of electrolytes, complete blood count, renal function and liver function was normal. The following serum studies were also negative or normal: thyroid-stimulating hormone,

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ammonia, vitamin B12, C-reactive protein, erythrocyte sedimentation rate, thyroid peroxidase antibodies, human immunodeficiency virus antibodies, lyme enzyme-linked immunosorbent assay, rapid plasma reagin, antinuclear antibody, anti-neutrophil cytoplasmic antibody (ANCA), lupus anticoagulant, cryoglobulins. The following antibodies were negative or normal: Anti-smooth muscle, Anti-smith, double-stranded DNA, anti-Ro, anti-La, Beta-2 glycoprotein IgG and IgM, ribonucleoprotein (RNP), myeloperoxidase (MPO), proteinase 3 (PR-3). Complement C3 was normal at 132 mg/dl. C4 was mildly elevated at 44 mg/dl. Serum and urine protein electrophoresis were normal.

Three lumbar punctures were performed during his hospitalizations and are reported in Table 1. All were inflammatory with a lymphocytic predominance. CSF was negative for herpes simplex virus (HSV) 1 and 2 on serial studies as well as at least one negative result for Eastern equine encephalitis virus (EEE) IgM, Powassan virus IgM, West Nile virus (WNV) IgM and IgG, Cryptococcus antigen, and Treponemal IgG antibody. CSF bacterial culture grew no organisms. CSF flow cytometry revealed 90% of the cells were CD3-positive T cells (CD4:CD8 ratio of 4.1). CSF cytology showed an increased number of lymphocytes, including occasional reactive forms.

Serum and cerebrospinal fluid (CSF) paraneoplastic panels were negative. CSF neuron-specific enolase and 14-3-3 protein were negative.

The patient's brain MRI on repeat admission to the outside hospital is shown in Fig. 1 and notable for bilateral deep white matter hyperintensities consistent with small vessel disease as well as restricted diffusion in the left parietal lobe. A follow-up MRI ten days later at our institution is depicted in Fig. 2 and demonstrates new areas of restricted diffusion in bilateral hemispheres. Additionally, multiple susceptibility-weighted imaging (SWI) lesions suggestive of cortical microhemorrhages were found. These microhemorrhages were not seen on outside imaging which used gradient echo sequence rather than SWI. Findings from CT and repeat MRI studies also revealed a new left subdural hematoma (SDH) with associated mild midline shift. Of note, this was after the fall sustained at the rehabilitation facility. CT and MR angiogram of neck and head showed no evidence of a vasculopathy, dural arteriovenous fistula or cortical venous sinus thrombosis.

Initial electroencephalogram (EEG) performed on day 9 at the outside hospital demonstrated diffuse encephalopathy with intermittent frontotemporal rhythmic slowing. EEGs performed following discovery of left SDH at our hospital revealed occasional left temporal focal delta slowing and bilateral semi-rhythmic slow waves consistent with left temporal cerebral dysfunction and bihemispheric encephalopathy. No epileptiform discharges or electrographic seizures were seen. We did not capture any acute episodes of agitation, aphasia or right facial weakness while on EEG.

Transthoracic echocardiogram (TTE) was normal. Transesophageal echogram (TEE) was aborted due to inability to traverse the esophagus. Transcranial doppler High-Intensity Transient signals (HITs) were negative for cerebral emboli. CT examination of the chest, abdomen, and pelvis revealed no evidence of malignancy.

3. Discussion

This patient's rapid evolution from highly functional to markedly cognitively impaired over a course of two months focuses the differential diagnosis on a rapidly progressive dementia (RPD). The inflammatory CSF and abnormal brain imaging suggest infectious, inflammatory, or vascular etiologies as most likely, but also possible are autoimmune, neoplastic, prion disease, and non-prion neurodegenerative etiologies. According to data from 1160

Table 1
Laboratory data – Lumbar puncture results.

Variable	Reference range	Hospital Day 9	Hospital Day 24	Hospital Day 33
Total nucleated cells (/μL)	0.00–0.02	62	24	1
Neutrophils (%)	–	–	4.0	11
Lymphocytes (%)	–	–	80.0	54
Monocytes/Macrophages (%)	–	–	16.0	26
RBC Count (/μL)	–	28	1600	42
Xanthochromia	Negative	NA	Negative	Present
Total Protein (mg/dl)	10–44	221	113	62.9
Glucose (mg/dl)	40–70	49	50	137

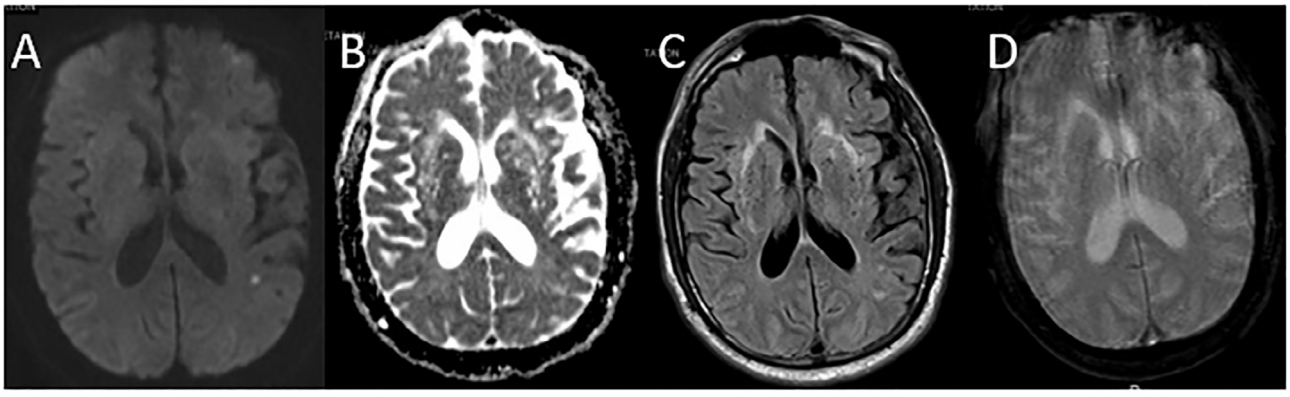

Fig. 1. Brain MRI, axial cuts. Images obtained hospital day 24 from initial hospitalization. (A) Diffusion-weighted imaging (DWI) and (B) apparent diffusion coefficient (ADC) sequences show small area of restricted diffusion in left parietal lobe. (C) Fluid attenuated inversion recovery (FLAIR) sequence demonstrates T2 hyperintensities in deep white matter. (D) Gradient echo sequence is without evidence of microhemorrhages.

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