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Clinical challenges

What's in a name?

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(In keeping with the format of a clinical pathological conference, the abstract and key words appear at the end of the article.)

1. Case report

A 51-year-old man underwent bronchoscopy at an outside hospital for evaluation of cough and pulmonary fibrosis. His medical history was significant for stable rheumatoid arthritis (RA) diagnosed 2 years prior and type 2 diabetes. He had no history of extra-articular RA. His medications were celecoxib, sulfasalazine, metformin, glimepiride, and pantoprazole. He had a history of alcohol abuse and was a previous smoker with a 25 pack-year history. He denied other drug use.

Bronchoscopy under monitored anesthesia care revealed that the arytenoids were mildly erythematous, but was otherwise unremarkable. In the recovery room he became agitated requiring propofol and fentanyl. He was then admitted for observation. Overnight, he had a fever of 103°F (39.44°C) and became tachypneic with irregular breathing and required intubation.

His serum white cell count was variably elevated from 8 to 15 billion cells/L (3.5–10.5 billion cells/L). Arterial blood gases were normal. Initial chest X ray showed chronic changes of pulmonary fibrosis with no evidence of pneumothorax or pneumonia. Computed tomography of the brain was normal. He was treated empirically for meningitis with ceftriaxone 2 g intravenous (IV) every 12 hours, vancomycin 1 g IV every 12 hours, and ampicillin 2 g IV every 4 hours and for seizures with phenytoin. He was extubated the following day and had worsening mental status changes and progressive weakness. He was then transferred to our hospital.

He was alert, but oriented only to person and place. He had mild dysarthria with "mumbling" speech and intact repetition. His pupils were equally round and reactive to light. Facial sensation was intact to light touch, and facial movements were symmetric. On motor examination, his right upper extremity

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was 4/5 proximally and 4/5 distally compared with left upper extremity strength 4−/5 proximally and 4/5 distally. In the lower extremities, strength on the right was 4+/5 proximally, 4+/5 distally versus 4−/5 proximally and 4/5 distally on the left. Sensation was intact to light touch in all 4 extremities individually, but he had extinction on the left with double simultaneous stimulation. He had difficulty with finger-to-nose testing. He was also noted to have a rash on his ankles bilaterally.

His neuro-ophthalmology examination showed visual acuity of 20/25 in each eye. The pupils were equal, round, and reactive to light with no relative afferent pupillary defect. Intraocular pressure measurements were 10 mm Hg in both eyes. Humphrey visual field testing 24-2 showed a left incongruous macular splitting homonymous hemianopsia (Fig. 1). Ocular motility was full. External examination, slit-lamp biomicroscopy, and dilated ophthalmoscopic examinations were normal.

What is the differential diagnosis?

What testing should be performed at this time?

2. Comments

2.1. *Comments by Steven A. Newman, MD*

Although a computed tomography scan had been normal, the obvious next step would be to obtain a magnetic resonance imaging (MRI) scan. This would be far more sensitive than computed tomography to recent infarct (diffusion weighted imaging) or abnormalities in white matter tracts (fluid-attenuated inversion recovery) and certainly should be used if microvascular disease is expected. The findings on MRI will likely direct additional testing depending on whether this looks like a local problem or a secondary problem from thromboembolism or systemic pathology.

One other test that might be helpful here would be angiography. This can often give clues to a primary vasculitis, not seen on other forms of imaging study (computed tomography, MRI, etc.). When ordering these tests, it would be important to have a neurologist on board and discuss the potential value of angiography with neuroradiology. More recent studies point out that biopsy and angiography remain the gold standard for diagnosis of small to medium size vessel vasculitis.[22]

3. Case report (continued)

MRI of the brain showed multiple areas of diffusion restriction in both cerebral hemispheres, but to a greater extent on the right, including the right frontal, right parietal, and right occipitotemporal gyri consistent with middle cerebral artery territory infarction with relative sparing of the white matter (Fig. 2). Electroencephalography showed lateralized discharges

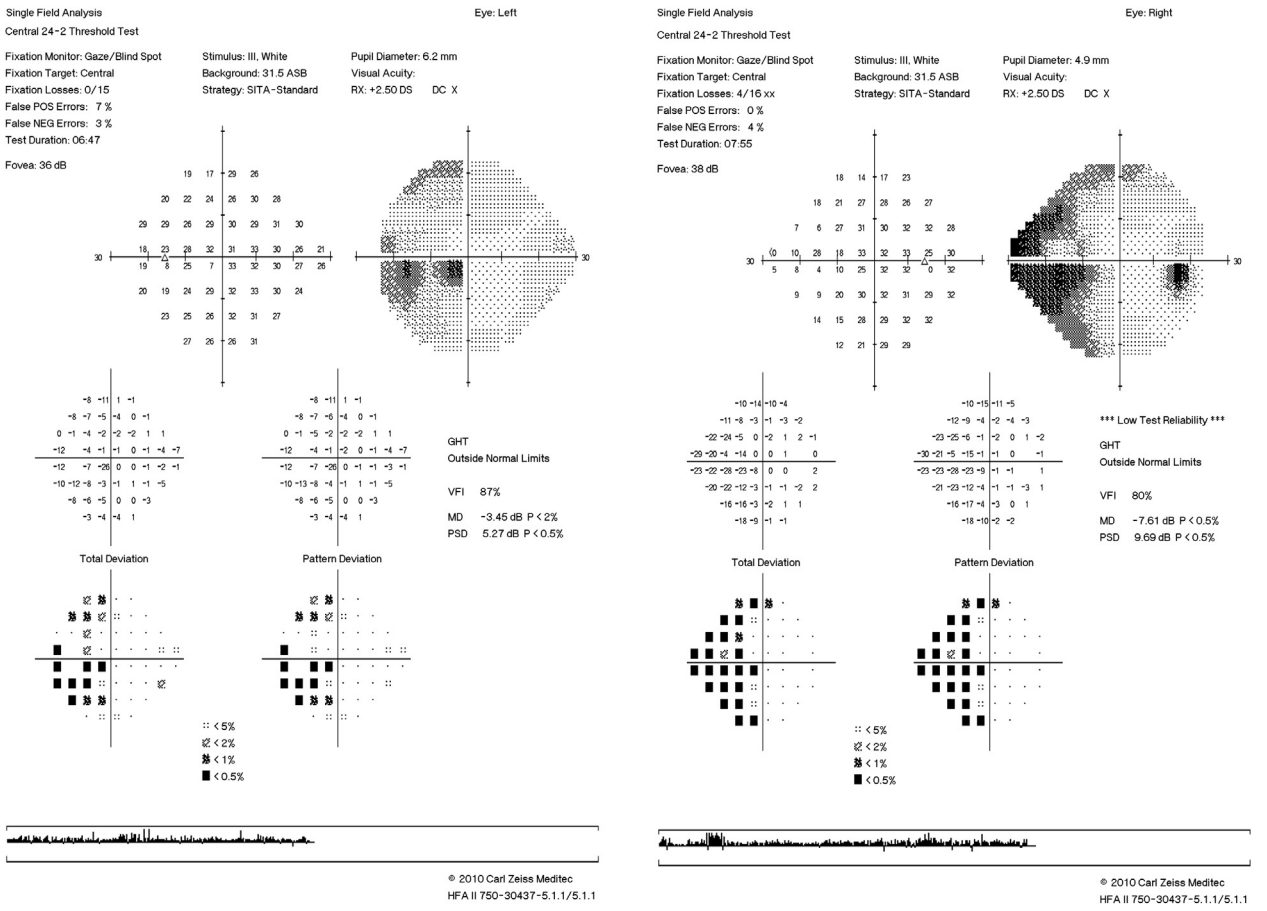

Fig. 1 – Humphrey visual field 24-2. Left incomplete incongruous macular splitting homonymous hemianopsia at initial presentation.