Title of Presentation: Rapidly Shifting into Third Gear

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I Please type 200-300 word history & exam abstract below

A 67-year-old right-handed woman developed the acute onset of visual loss in her left eye, two days after the resection of two basal cell carcinomas from her nose. An ophthalmologic evaluation three days after the onset of her visual loss revealed a corrected visual acuity of 20/40 OD and counting fingers OS. She had a 2+ left APD. Ophthalmoscopic examination revealed arterial narrowing and left disc pallor. She was diagnosed with a central retinal artery occlusion. Evaluation included a normal ECG and carotid ultrasound study. Cardiac echo showed mild left ventricular hypertrophy.

Two weeks later she developed speech difficulties and right-sided weakness, which gradually progressed over several days. The patient was admitted to another hospital with a presumptive diagnosis of stroke and started on heparin. A subsequent brain MRI revealed a large heterogeneously enhancing periventricular left frontal lesion. Mild enhancement of the left optic nerve was also noted. MR spectroscopy was suspicious for a neoplastic lesion. The patient was transferred to the Neurosurgery service at our institution for further evaluation and management. A metastatic work-up, including a CT of her chest, abdomen, and pelvis was normal. A lumbar puncture was attempted twice under fluoroscopic guidance, but was unsuccessful. On neuro-opthalmologic examination, her VA was 20/25 OD and CF OS. Right eye visual fields were full with a markedly depressed field OS, particularly inferiorly. A 2+ left APD was present. The left optic nerve was pale. She had full extradural movements except for decreased pursuit to the right. She had a prominent right central 7th nerve palsy, and a moderate right hemiparesis with increased tone and reflexes on that side.

A diagnostic procedure was performed

ABSTRACT AUTHOR'S ACKNOWLEDGMENT - CONCURRENCE AND DISCLOSURE STATEMENT:
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PROGRAM FORM (ANSWER) 33rd ANNUAL MEETING, April 21-22, 2001, Ann Arbor, Michigan

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Final Diagnosis: **Tumefactive Multiple Sclerosis**

SUMMARY OF CASE INCLUDING PATHOLOGY AND REFERENCES (200-300 WORDS):

†A stereotactic biopsy of the frontal lesion revealed a diffuse infiltration of foamy macrophages and lymphocytes with numerous reactive astrocytes. Luxol fast blue stain showed demyelination. The final biopsy diagnosis was demyelination. Of note, the patient also had two separate lesions on MRI, one in the right pontomedullary junction, and a spinal cord lesion at C7-T1. She was treated with high-dose steroids and her condition stabilized. Her symptoms increased as her steroids were tapered. She was then successfully treated with high-dose steroids and plasmapheresis.

There have been several case reports of large demyelinating destructive lesions mimicking a tumor (1-6). Our case is unusual, however, given the size of the lesion, the extent of the edema, and the midline shift. In our patient, the initial concern was for a lymphoma or glioma, and an inflammatory demyelinating process was much lower down on the differential, especially given the patient’s age and lack of prior history consistent with a diagnosis of MS. One study suggests that such large tumor-like demyelinating lesions may represent a separate entity; one that is intermediate between MS and ADEM, and may carry a different prognosis (2). The majority of these patients did not go on to develop additional lesions in a follow-up period ranging from nine months to twelve years.

The decision of when to perform a biopsy in these cases may be a difficult one. On MRI, lesions that are well-defined, rounded, and occur along the ventricle, particularly when coupled with other lesions, may lessen the need to obtain pathologic confirmation (5). Newer MRI techniques such as magnetization transfer, perfusion MRI, and MR spectroscopy may also be helpful in differentiating between a tumefactive demyelinating lesion and a neoplasm (1). Unfortunately, in our patient, the MR spectroscopy was more suggestive of a tumor.

Even in cases that are biopsied, tumefactive MS lesions may be mistaken for neoplasms. One report describes five patients whose biopsies were initially diagnosed as tumors. Four of these patients had full dose radiation therapy, and had an unexpectedly poor outcome, suggesting that XRT may be harmful in patients with demyelinating disease (3). Finally, although plasmapheresis in demyelinating disease is controversial, there have been anecdotal case reports of success, particularly in cases of ADEM that are refractory to high-dose steroids (7-11). Furthermore, a small, but randomized, double-blind trial of plasmapheresis in cases of acute demyelinating events (either MS or other inflammatory conditions) demonstrated significant improvement in neurological disability (12).

References: