Systemic Lupus Erythematosus Presenting as "One-and-a-Half Syndrome"

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"One-and-a-half syndrome" is a lesion in the parapontine region involving both the parapontine reticular formation (PPRF) and the medial longitudinal fasciculus ipsilaterally. There is a failure of lateral conjugate gaze to one side, impairment of adduction of the eye on the same side, and nystagmus on abduction of the opposite eye. This syndrome is more commonly seen with multiple sclerosis and rarely presents in patients with systemic lupus erythematosus (SLE)[1,3,8], as in this case. In December 1999 a 26-year-old African-American woman with SLE diagnosed in 1996 presented with complaints of lightheadedness and double vision. She was taking tapering doses of prednisone. She had history of pericardial effusion (status post pericardial window 1998) and had had a relapse of diffuse arthralgia 6 months before. She was hypertensive and hypothyroid taking supplementation medication. At presentation the pupils were equally reactive to light and accommodation, with mild extopia and diplopia. The field of vision was normal. Other significant clinical findings included "paralytic strabismus" of the right eye and gaze-evoked horizontal nystagmus on abduction. There was complete loss of left eye abduction and right eye adduction. Funduscopic examination was normal. All other cranial nerves were intact with normal higher central nervous system (CNS) functions. Laboratory findings included erythrocyte sedimentation rate (ESR) 85 mm/h, complement C3 and CH50 normal, C4 13 low. Magnetic resonance imaging T2 weighted sequence showed 7 mm rounded focus of increased signal in the upper pons of the lower midbrain on the left. The lesion was not seen on the T1 weighted sequence and showed no enhancement with contrast, suggesting lupus vasculitis[4]. Considering the clinical and laboratory findings and history of SLE, the acute onset of left intranuclear ophthalmoplegia with left abducent nerve palsy confirmed the diagnosis of "one-and-a-half-syndrome." She was initially given high doses of iv steroids and then changed to oral steroids[5-7]. She responded well and extraocular eye movements were normal within 3 days.

"One-and-a-half syndrome" is a lesion in the parapontine region (involving the PPRF and medial longitudinal fasciculus) presenting with failure of adduction of ipsilateral eye and nystagmus on abduction of the contralateral eye resulting in loss of lateral conjugate gaze. Multiple sclerosis, which is more common with this presentation, was ruled out with the negative funduscopic result, increased ESR, and MRI results. Supranuclear gaze palsies due to vasculitis in SLE have been rarely reported. Our literature review revealed one case report of "one-and-a-half-syndrome" from Turkey. In the few case reports of internuclear ophthalmoplegia in SLE[1-3], ocular motor signs are uncommon and transitory and there was poor correlation with clinical imaging. Our patient had classical clinical signs and we were able to support the diagnosis with clinical imaging. A common lupus related abnormality of the CNS is microfocal scarring associated with intimal changes in small arterioles[6]. Although multiple sclerosis is more common with the above symptoms, SLE vasculitis involving brainstem should

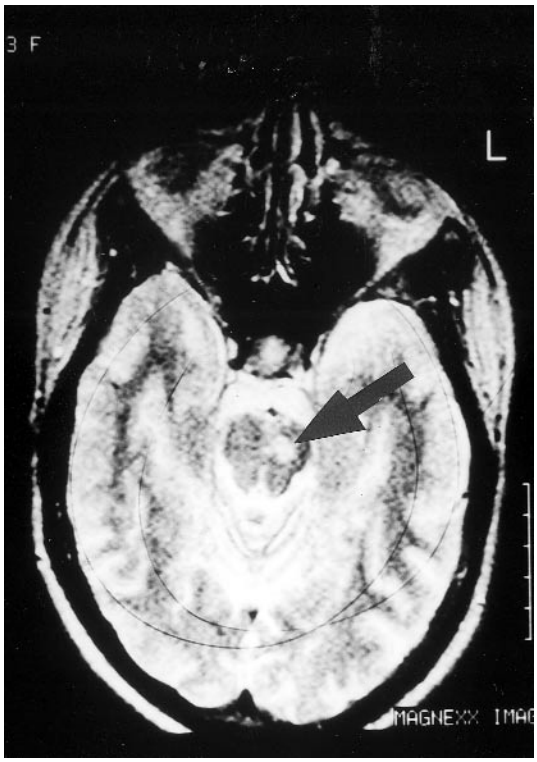

Figure 1. MRI T2 weighted sequence reveals 7 mm rounded focus of increased signal in the upper pons of the lower midbrain.

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always be suspected in young women[7,8]. Steroid treatment resulted in excellent improvement. Other treatment modalities available for refractory cases are cyclophosphamide, immunoglobulins, and plasmapheresis.

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