Case 1

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Clinical Abstract:

A 75-year-old right-handed white male was admitted to the hospital with a one-year history of episodic left retro-orbital pain and diplopia.

The patient had been well until one year prior to admission when he first complained of left-sided headache. He was treated for sinusitis and otitis with antibiotics. A more localized left retro-orbital constant throbbing pain associated with horizontal diplopia with left lateral gaze returned three months later. At that time his neurologic exam was normal, ESR was 70 mm/hr. Despite a normal temporal artery biopsy he was treated with prednisone 60mg/day. The pain and diplopia resolved shortly after treatment was initiated. Five months later he returned to medical attention with recurrence of left retroorbital pain and diplopia. Evaluation showed a depressed left nasolabial fold, but no ophthalmoplegia. An angiogram was normal and a CT scan showed diffuse atrophy. A lumbar puncture revealed protein 61 mgm %, glucose 81 mgm %, and 1+ oligoclonal bands. When steroids were increased the patient again became pain-free.

Six months later he complained of sharp retro-orbital pain which worsened with eye movements, diplopia in all fields of gaze, and patchy left facial numbness. He also had anorexia, nausea, vomiting, dysphagia and had lost 50 lbs. in weight.

He had had a TIA with transient global amnesia three years prior to admission and pneumonia several months prior to admission.

Admission physical examination showed a cachectic male with depressed affect, and mild dementia. He had scalp tenderness, proptosis and chemosis. Both globes easily ballotted. There was no meningismus. He had a pupil-sparing left III, IV, VI, and VII paresis. There were full EOM's in the right eye, fundi were normal. He showed general paratonia, diffuse weakness in the right upper and right lower extremities with slight reflex asymmetry. Deep tendon reflexes and proprioception below the knees were absent in the lower extremities. He had a frontal gait with wide-based, reluctant, shuffling steps, no ataxia, and no Romberg sign. ESR was 21mm/hr. A lumbar puncture was done which showed an opening pressure of 60 cm H2O, protein 110 mgm %, glucose 51 mgm % (serum 118 mgm %), 1 RBC and 46 WBC per mm (3P7M90L), oligoclonal bands were present, IgG index 2.9; cytology was normal, crypto Antigen, AFB, and fungal cultures were negative, as were serum RPR, and FTA. A CT scan showed a "thickened left cavernous sinus."

Following a craniotomy, Decadron and Imuran were begun. The patient became pain-free and horizontal EOM's improved. His later course was complicated by gastrointestinal bleeds, a hypertensive episode and urosepsis. His renal function deteriorated, he developed pulmonary edema and expired.

Material Submitted: One hemotoxylin and eosin stained section and a gross photograph of the base of the skull at autopsy.

Points for Discussion: 1. Diagnosis 2. Pathogenesis.