

The Dual Catastrophe of Behçet's Disease Visual Loss Followed by Acute Spinal Shock After Lumbar Drain Removal

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Abstract : Context: Increased intracranial pressure and associated symptoms such as headache, papilledema, motor or sensory deficits, seizures, and conscious disturbance are well-known in acute CVT. However, visual loss is not commonly associated with this disease, except in the case of secondary IIH associated with it. Process: We report a case of a 40-year-old male with Behçet's disease and cerebral venous thrombosis, and other multiple comorbidities admitted with a four-day history of increasing headache and rapidly progressive visual loss bilaterally. The neurological examination was positive for bilateral papilledema of grade 3 with light perception on the left eye and counting fingers on the right eye. Brain imaging showed old findings of cerebral venous thrombosis without any intraparenchymal lesions to suggest a flare-up of Behçet's disease. The lumbar puncture, followed by the lumbar drain insertion, gave no benefit in headache or vision. However, he completely lost sight. The right optic nerve sheath fenestration did not result in vision improvement. The acute spinal shock complicated the lumbar drain removal due to epidural hematoma. An urgent lumbar laminectomy with hematoma evacuation undertook. Intra-operatively, the neurosurgeon noted suspicious abnormal vessels at conus medullaris with the possibility of an arteriovenous malformation. Outcome: In a few days following the spinal surgery, the patient vision started to improve. Further improvement was achieved after plasma exchange sessions followed by cyclophosphamide. In the recent follow-up in the clinic, he reported better vision, drove, and completed his Ph.D. studies. Relevance: Visual loss in patients with Behçet's disease should always be anticipated and taken reasonable care of, ensuring that they receive well-combined immunosuppression with anticoagulation and agents to reduce intracranial pressure. This patient's story is significant for a high disease burden and complicated hospital course by acute spinal shock due to spinal lumbar drain removal with a possible underlying spinal arteriovenous malformation.

Keywords : Behcet disease, optic neuritis, IIH, CVT

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