Case Report

Blindness, ophthalmoplegia and extensive radiculopathy: An unusual clinical syndrome in intracranial sino-venous thrombosis

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Isolated intracranial hypertension is a common manifestation of intracranial sino-venous thrombosis (ISVT). Markedly elevated intracranial tension presents with unusual features including cranial neuropathies and radiculopathy. We report two cases with ISVT, which presented with headache, papilledema, progressive visual loss, complete ophthalmoplegia and flaccid areflexic quadriparesis along with a normal sensorium. Magnetic resonance imaging (MRI) of the brain and cervical spinal cord showed no lesions that could account for the neurological deficits. Markedly elevated lumbar CSF pressure was noted in both cases. Nerve conduction study favored radiculopathy in one case and was normal in the other. Raised intracranial pressure was found to be the sole cause for the clinical manifestations. Visual impairment persisted in one patient despite lumbo-peritoneal shunting while the other died of septicemia. To our knowledge, such a presentation has not been reported in literature till date.

Key Words: Intracranial sino-venous thrombosis, Raised intracranial pressure, Radiculopathy, Guillain-Barre syndrome.

Case Report

Case 1
A 24-year-old woman presented with neck pain, diplopia of 4 weeks duration and progressive visual loss and quadriparesis of 3 weeks duration. She also reported shoulder and back pain. At admission, her vitals were stable and she had a clear sensorium. Optic fundi showed bilateral papilledema. Vision was restricted to perception of hand movements in both the eyes. Bilateral complete ophthalmoplegia with mid-dilated pupils poorly reacting to light and accommodation was noted. Gag reflex was diminished on the left. She had generalized areflexia and hypotonia with Grade 2 power (Medical Research Council grading) in the proximal muscles and Grade 3+ distally along with moderate trunk and neck weakness. Flexor plantar responses were noted bilaterally. Sensory examination was normal. Neck stiffness and Kerning’s sign were present.

Her hematological and basic biochemical parameters were normal. CSF study showed elevated opening pressure with CSF spurting above the 400 mm mark of the manometer. CSF analysis revealed 5 lymphocytes, protein of 0.40 g/L and sugar of 0.50 g/L. Digital subtraction angiography (DSA) showed thrombosis in the posterior superior sagittal sinus, torcular and right transverse sinuses. Urine homocysteine, anti-nuclear antibody, anti-phospholipid antibody and sickling test were negative. Visual evoked potential to strobe stimulation showed major positivity at 187 ms in the right eye and was inelicitable in the left eye. The nerve conduction study including F-waves, was normal.

She underwent a lumbo-peritoneal shunt following which her ophthalmoplegia and limb weakness improved over two weeks. She was continued on anticoagulants. At one-year follow-up, she had bilateral optic atrophy and only a marginal improvement in vision. Her hematological and basic biochemical parameters were normal. CSF study showed elevated opening pressure with CSF spurting above the 400 mm mark of the manometer. CSF analysis revealed 5 lymphocytes, protein of 0.40 g/L and sugar of 0.50 g/L. Digital subtraction angiography (DSA) showed thrombosis in the posterior superior sagittal sinus, torcular and right transverse sinuses. Urine homocysteine, anti-nuclear antibody, anti-phospholipid antibody and sickling test were negative. Visual evoked potential to strobe stimulation showed major positivity at 187 ms in the right eye and was inelicitable in the left eye. The nerve conduction study including F-waves, was normal.

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Case 2
A 33-year-old woman presented with headache and vomiting of two months duration and unsteadiness of gait and seizures of 1 month duration. She also reported shoulder and back pain. At admission, her vitals were stable and she had a clear sensorium. Optic fundi showed bilateral papilledema. Vision was restricted to perception of hand movements in both the eyes. Bilateral complete ophthalmoplegia with mid-dilated pupils poorly reacting to light and accommodation was noted. Gag reflex was diminished on the left. She had generalized areflexia and hypotonia with Grade 2 power (Medical Research Council grading) in the proximal muscles and Grade 3+ distally along with moderate trunk and neck weakness. Flexor plantar responses were noted bilaterally. Sensory examination was normal. Neck stiffness and Kerning’s sign were present.

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Discussion

The two cases of angiographically proven ISVT reported here had remarkably similar clinical features, consisting of acute bilateral visual loss, complete internal and external ophthalmoplegia, areflexic flaccid quadriparesis and normal sensorium along with raised ICP. MRI brain and spine showed no abnormalities that could directly account for the deficits. Reduction in CSF pressure was associated with partial recovery suggesting a possible role of intracranial hypertension in the pathogenesis. Though blindness and ophthalmoplegia are well-known manifestations of raised ICP, flaccid quadriparesis is extremely rare.

We initially considered the possibility of midbrain infarction secondary to deep cerebral venous thrombosis as the cause for areflexic quadriparesis with ophthalmoplegia. However, MRI brain, done twice in the second case, showed no lesion in the brainstem. Cavernous sinus thrombosis was excluded on angiography. Various cranial nerve palsies due to raised ICP are well described in the literature. This has been attributed to a direct effect of raised ICP causing distortion or displacement of brainstem structures and or cranial nerves. Kuchniet al described cases of transverse and sigmoid sinus thrombosis presenting with isolated cranial nerve palsies, presumably due to thrombosis of pontine and medullary veins. But the prompt reversal of oculomotor paralysis in our first case and immediate recovery of pupillary paralysis after the lumbo-peritoneal shunt in our second case favors raised ICP as the cause.
cause of oculomotor palsy.

With a normal MRI of brain and spinal cord, flaccid areflexic quadriplegia in our cases is likely to be due to a lower motor neuron involvement. The inelicitable F-waves with normal peripheral conduction study in our second case supported a radicular involvement. Needle electromyography could not be performed as our patients were on anticoagulants. The normal nerve conduction study in the first case could be due to milder involvement and dilution factor.

Needle electromyography could not be performed as our patients were on anticoagulants. The normal nerve conduction study in the first case could be due to milder involvement and dilution factor. Minor manifestations of radiculopathy in the form of acral paraesthesias and spinal and radicular pain have been described in patients with pseudotumour cerebri. Recently, Obeid et al reported two patients with extensive radiculopathy due to intracranial hypertension; one patient had pseudotumour cerebri and the other ISVT. Both had papilledema and impending visual loss. Electrodiagnostic study showed features of a diffuse radiculopathy. Both cases were diagnosed initially as Guillain-Barre syndrome (GBS) and were treated with intravenous immunoglobulin without benefit. They responded well to lumbo-peritoneal shunt supporting the view that raised ICP was responsible for the radiculopathy. However, unlike our cases, ophthalmoplegia was absent in these cases. Ophthalmoplegia with quadriplegia as seen in our cases may resemble Miller Fisher-Guillain- Barre overlap syndrome clinically. However our cases had normal CSF protein and nerve conduction study was not diagnostic of GBS in either of them. Longstanding radiculopathy can lead to axonal loss and simulate axonal forms of GBS electrophysiologically also. Though it is well known that papilledema rarely does occur in GBS, cases with atypical or normal electrophysiological study should be viewed with suspicion; CSF pressure testing and MRV or DSA would be advisable to exclude ISVT in such cases.

We suspect that the mechanism of radicular dysfunction is due to elevated CSF pressure distending the subarachnoid space around the nerve roots resulting in mechanical stretching and pressure effect. Enlargement of the spinal subarachnoid space along with distended root pouches has been reported in a patient with radiculopathy due to idiopathic intracranial hypertension. Venous ischemia of nerve roots secondary to impaired venous outflow through the thin-walled radicular veins may also play a role. Motor roots appear to be more vulnerable as weakness was the major deficit in our cases and in those reported by Obeid. We propose that a marked rise in ICP produces a potentially reversible dysfunction of the cranial nerves and spinal nerve roots leading to this unusual clinical syndrome. In our experience, such cases appear to be at risk of irreversible visual loss.

In conclusion, we report that the syndrome of visual impairment, ophthalmoplegia and flaccid quadriplegia in patients with ISVT signifies a markedly raised ICP. Hence, patients with these manifestations should be treated early and aggressively to decrease the ICP to safe levels.

References


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