Miyazaki syndrome is a cervical myelopathy or cervical radiculopathy caused by cervical epidural venous congestion, due to shunt overdrainage. The complex pathophysiology includes CSF pressure-changes consistent with the Monro-Kellie hypothesis and a non-functional Starling resistor, leading to spinal epidural venous plexus enlargement and dilation. This venous congestion may be significant enough to exert compression on the spinal cord or nerve roots. The typical clinical and imaging findings together with a history of ventricular CSF shunting may establish the diagnosis, proven by a successful treatment. The aim of treatment is the abrogation of CSF over-drainage. The eligible interventions may be the followings: the increase of the opening-pressure of the valve system by the insertion of a new programmable valve if necessary, closing or removing the shunt.

In 1997 Miyazaki et al. described a case of intracranial hypotension syndrome due to overdrainage of cerebrospinal fluid presented with hearing loss after ventriculoperitoneal shunting procedure. A 69-year-old man suffering from subarachnoid hemorrhage presented with an angiogram showing two aneurysms, one of the right internal carotid artery and one of the middle cerebral artery. Neck clipping was performed. One month later, he developed normal pressure hydrocephalus (NPH), which was treated by ventriculoperitoneal shunting procedure using low pressure Pudenz valve system. Trias of NPH were improved by insertion of shunt system. However, he complained of hearing loss which was worsened by upright position and improved by lying down. Such kinds of phenomenon were demonstrated by audiogram showing that the transitory decrease of hearing and electrocochleography showing the elongation of N1 latency at upright position. These data suggested that his hearing loss was caused by inner ear or auditory nerve lesion. After the shunt system was replaced into the antisiphon device, his hearing disturbance improved. Axial computed tomography of bone window at the level of orbitomeatal line demonstrated widely perilymphatic duct on both sides. This finding suggested that the fluctuation of intracranial pressure was easily transmitted into the cochlear through the widened perilymphatic duct, resulting in hearing disturbance.

Várallyay et al. want to call attention to this rare iatrogenic condition with potentially severe consequences.

They performed a systematic literature-review and presented ther five cases.

Once recognized in time, Miyazaki syndrome can be well taken care of.

Patients with chronic ventricular shunt need monitoring for CSF over-drainage to recognise potential complications such as cervical myelopathy or radiculopathy.
In 2018 a 33-year-old patient had undergone placement of a ventriculoperitoneal shunt with a pressure-adjustable valve for communicating hydrocephalus years before presenting to our department with the complaints of constant headache and unsteady gait. On the basis of the clinical picture and her history, plain and contrast-enhanced cranial and whole spine magnetic resonance imaging and magnetic resonance angiography examinations were performed, with the scans revealing signs indicative of cerebrospinal fluid hypotension typical of Miyazaki syndrome.

In 2015 Caruso et al. reported one case.

Overshunting associated myelopathy is a rare complication of CSF diversion that should be familiar to physicians who routinely evaluate patients with intracranial shunts.

Only 12 previous cases have been reported in the literature.

OSAM has to be considered according to the Monro-Kellie hypothesis and is affected by an engorgement of the cervical epidural venous plexus, which can produce cervical myelopathy. Since it can be treated simply by increasing the shunt resistance, surgeons should be aware of the rarely detected overdrainage complication.

Classically, patients present with positional headache, but less common symptoms include neck pain and cranial nerve palsies.

A 45-year-old-patient with shunt-dependent, congenital hydrocephalus presented with an 8-year history of progressive tetraparesis and gait disorder in the Department of Neurosurgery, University of Tübingen, Germany. The patient was wheelchair-dependent. A new MRI scan of the head revealed slit ventricle syndrome and dural enhancement due to shunt overdrainage. An MRI and a CT-Phlebography of the cervical spine revealed engorgement of the epidural venous plexus with secondary compression of the spinal cord and myelomalacia. Surgery was performed during which we implanted a shunt valve. The patient recovered from surgery without any new deficits. The tetraparesis improved during the inpatient hospital stay. CT-Phlebography was performed 5 days after surgery and showed that the epidural venous plexus anterior to the cervical spinal cord had returned to nearly normal size. On follow-up examination 3 month after surgery, the patient´s strength had improved, and he was able to walk short distances with assistance and with ankle foot orthosis on the right side.

OSAM has to be considered according to the Monro-Kellie doctrine and is affected by an engorgement of the epidural cervical venous plexus, which can produce cervical myelopathy. Since it can be treated simply by increasing the shunt resistance, surgeons should be aware of the rarely detected overdrainage complication.

Ho et al., presented 2 cases of cervical myelopathy produced by engorged vertebral veins due to
overshunting. Overshunting-associated myelopathy is a rare complication of CSF shunting. Coexisting cervical degenerative disc disease may further increase the difficulty of diagnosing the condition. Neurosurgeons and others who routinely evaluate patients with intracranial shunts should be familiar with this rare but possible diagnosis.

A 26-year-old woman with shunt-dependent, congenital hydrocephalus, presented with rapidly progressive cervical myelopathy following ventriculoperitoneal shunt revision. Imaging revealed engorgement of the cervical epidural venous plexus and mass effect on the cervical spinal cord. “Over-shunting associated myelopathy” is a rare complication of CSF diversion that should be familiar to physicians who routinely evaluate patients with intracranial shunts.

References


