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Neurology Case Series

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Dear Doctor,

Greetings from Micro Labs Limited.

We are happy to bring you "Neurology case series", which contains latest and interesting cases in the field of Neurology.

This issue contains

1. **A case report on chronic CSF leak from the lumbar-peritoneal shunt tract.**
2. **The significance of spinal cord perfusion in delayed, transient quadriplegia.**
3. **A case on rare co-occurrence of multiple sclerosis and Wilson's disease**
4. **A Case Report of Delayed Hydrocephalus Detection in Corticobasal Degeneration Following Mildly Traumatic Subarachnoid Hemorrhage**

Should you require any specific article or medical information, please feel free to contact us at medicalservices@microlabs.in

Looking forward to your valuable feedback

Best Regards,

Medical Team,

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1. A case report on chronic CSF leak from the lumbar-peritoneal shunt tract.

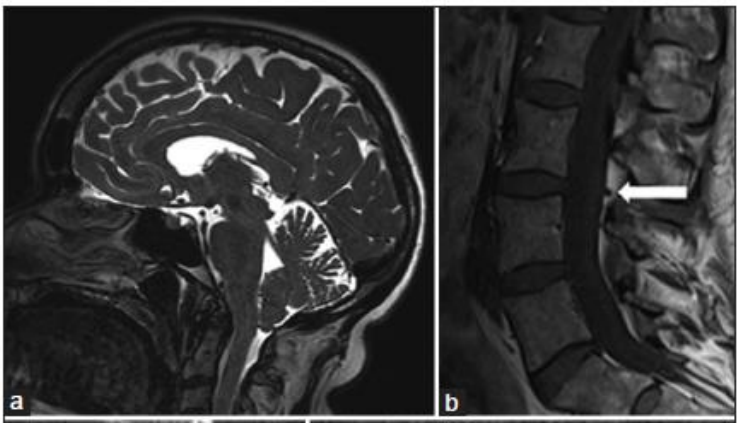
Introduction:

Cerebrospinal fluid (CSF) is a transparent, protein- and glucose-rich liquid found in the central nervous system's subarachnoid region. CSF controls the temperature of the central nervous system and protects the brain and spinal cord. As a result, a leak in this system can compromise brain blood flow and function, as well as increasing the risk of direct damage to the brain parenchyma due to a lack of fluid cushion. Furthermore, the occurrence of a CSF leak implies that additional investigation and treatment are required.[1] CSF leaks with symptoms are widespread and have a well-established diagnostic approach.[2] here is a case of long-term severe low-pressure symptoms caused by high-flow CSF escaping through an acquired arachnoid-epidural venous plexus To the author's knowledge, no such mechanism has been described in the literature.

Asurgical repair with post-operative precautions is more likely to achieve permanent repair in CSF Leak

Case presentation:

An 18-month history of developing postural holocranial headaches, dizziness, brain fog, dull neck discomfort, left face numbness and twitching, and pain in both arms in a C7–C8 distribution was presented by a 55-year-old female. The patient was confined to bed for the whole day since her symptoms were severe when upright but disappeared when reclined. A diagnosis of idiopathic intracranial hypertension (IIH) had been confirmed 20 years prior, and a lumboperitoneal (LP) shunt had been implanted. For continued symptoms, a fixed-pressure valve ventriculoperitoneal (VP) shunt was implanted the next year. The low-pressure symptoms listed above originally developed 5 years prior to the current presentation. Because of concerns about CSF overdraining, both the LP and VP shunts were removed. After the LP and VP shunts were eventually removed, the low-pressure symptoms faded away for a while before reappearing again for present episode. A Chiari I malformation was discovered on MRI (CMI). On sagittal MRI, however, a slight dural disruption could be noticed on closer inspection [Figure 1]. A CSF bleb L3/4 was discovered on a subsequent CT myelography. The



(a) Sagittal T2W MRI scan showing tonsillar descent with effacement of cisterna magna. (b) Sagittal T1W MRI reported as normal but reveals dural defect (white arrow) at L3/4.

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LP shunt was removed at the same level as this dural defect. An procedure was performed on the patient. A posterior and midline route was used to reach the L3/L4 dural level. With an overlaying connection of adipose tissue and a vascular, a dural defect and arachnoid bleb were discovered. The vessel was coagulated and cut . The adipose tissue on the dura was dissected and cauterised. The arachnoid was retained. The dural defect was ultimately closed and TISSEEL adhesive was used to cover it. Acetazolamide 500 mg BD was begun postoperatively and was continued for long duration however no improvement was seen in patient's symptoms A follow-up MRI revealed a considerable pseudomeningocele with a persistent leak. CSF was detected leaking around the prior sutures. intraoperatively, The arachnoid was reopened once all sutures were removed. The dural defect was extended again and closed. On the suture line, a little portion of muscle was crushed and positioned. An adjacent portion had a lumbar drain inserted for 5 days and then removed. The patient's major complaints of postural headache and dizziness have significantly improved four months after the surgery. She is now mobilising on her own and is being tapered off of the acetazolamide.

Discussion:

This is a rare instance of chronic CSF over drainage caused by a persistent dural defect that developed after the removal of an LP shunt. This case differs from previously reported occurrences of spontaneous intracranial hypotension[3] and CSF bypassing in situ LP shunts.[4] This case demonstrates how, in the first instance, a long-standing CSF leak may benefit from definitive surgical correction.In the case of chronic symptoms and a verified spinal CSF leak, a research by Haini et al recommends surgical therapy no later than 12.4 weeks following onset.[5]

Conclusion:

An acquired arachnoid-epidural venous plexus appears to have caused excessive CSF absorption. In patients with low blood pressure symptoms and a CMI, a strong index of caution for cerebral hypotension is indicated. A surgical repair with post-operative precautions are more likely to result in a permanent repair.

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2. The significance of spinal cord perfusion in delayed, transient quadriplegia

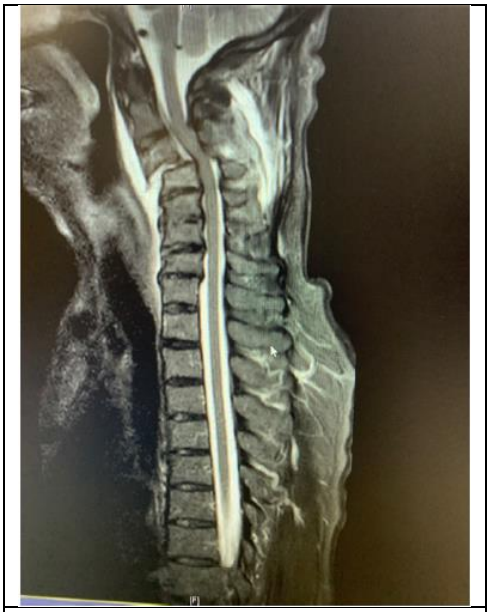
Introduction:

A temporary loss of motor and often sensory function in the arms and legs is known as transient quadriplegia. The motor symptomscan range from paralysis of arm and leg weakness to full paralysis. Numbness, tingling, and burning pain are examples of sensory symptoms. A hit to the head or a neck injury are common causes of this illness .[1] This is an uncommon combination of conditions in the case of a relatively common accident with potentially fatal implications.[2] The need of maintaining spinal perfusion pressure in the acute therapy of traumatic spinal injury is highlighted in this instance.

The need of maintaining spinal perfusion pressure in the acute therapy of severe spinal injury is highlighted in this case.

Case Presentation:

While body surfing, a man in his late 60s had a violent hyperflexion cervical injury. His medical history includes osteoarthritis, which necessitated bilateral knee replacements, as well as a single incident of sciatica, which was completely healed after a paraspinal corticosteroid injection. Apart from osteoarthritic, degenerative abnormalities, there was no history of hypertension or signs of any particular spinal condition. In a Rugby scrum event 40 years ago, a younger sibling sustained a bifacet C4-5 fracture dislocation without any neurological damage. There was no indication of any underlying spinal illness. The patient was able to self-extricate following the acute injury in the surf, but he felt extreme neck pain and was found to have marked cervical muscular spasm. Patient couldn't lie flat due to pain, so he was gently supported in a semi-sitting posture while waiting for an ambulance to pick him up from the beach. In partial beachside evaluation, the patient seemed to be totally functioning, with normal motor and sensory function. Patient could move all of his limbs on request and had no sensory issues, however this was not systematically examined. His intercostal muscle function was found to be normal, and he had a regular breathing pattern. He experienced quickly progressing quadriplegia over the next 15–30 minutes. The patient felt as if he was' shutting down,' and he got quite worried, as if he was about to die. Although no full neurological examination was performed, there was no voluntary movement or subjective sensation in any of the limbs. He was suffering from dyspnoea. With the commencement of paradoxical breathing and intercostal recession, intercostal muscle function was now impaired.



MRI scan of the cervical spine indicating anterior dislocation of C3 on C4 with marked canal narrowing and displacement of CSF but without any direct injury or compression to the spinal cord.

With continued manual neck support, he could finally be laid flat. Dyspnoea was reduced as a result of this. The blood pressure was 70 mm Hg systolic with a heart rate of 60 beats per minute when the ambulance arrived more than 60 minutes after the initial injury. The lack of cerebral function below C4 was established by basic questions and observation, rather than a full neurological test. The presence of relative bradycardia rather than reflex tachycardia implies a neurogenic mechanism for the hypotension, which is compatible with the tetraplegia. Fluids were administered using intravenous line. Metaraminol was given by the emergency doctor, and the patient's blood pressure was normalised. . A rigid cervical collar was placed around the patient's neck. The patient was gently lifted onto a palisade and carried away from the beach by hand. One toe and then one foot were seen moving just before being taken into the ambulance. The patient was sent to a regional spinal trauma centre for treatment. during transport, a dose of metaraminol was given. Neurological function had nearly totally restored by the time the patient arrived at the tertiary centre, with only some residual hand weakness and minimal sensory impairment visible. C3–5 fixation and anterior reduction and C3–C6 surgical posterior were performed. The same evening, the patient made an uncomplicated recovery after the fixation three days later. Following surgery, he was discharged from hospital and returned home on Day 8 after the injury. Examination of the nervous system during the recovery period, no objective abnormality was discovered despite some subjective weakness. The patient experienced a change in sensation in his right hand, as well as some neuralgic pain. Approximately 4 months after the incident, Subjective hand sensations vanished all of a sudden. The neuralgic pain in the head continues, however has greatly decreased.

Discussion:

This case demonstrates the value of well-established principles of acute spinal injury therapy, such as field support and neck stabilisation, as well as early surgical fixation. The unique path, on the other hand, emphasises the necessity of spinal cord perfusion in acute spinal cord injury post-injury care.[3] Maintaining perfusion pressure in spinal injuries has become a part of standard clinical guidelines.[2]

Conclusion:

Although the exact pathogenesis of the transient quadriplegia observed in this case cannot be established, the personal and societal effect of spinal cord injury is so great that any initiative that could reduce or even prevent the clinical impact of spinal injury, such as in the case described, deserves to be considered. Blood pressure management for instance, should be considered "standard of care" since it has a low cost and a low risk of complications.

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3. A case on rare co-occurrence of multiple sclerosis and Wilson's disease

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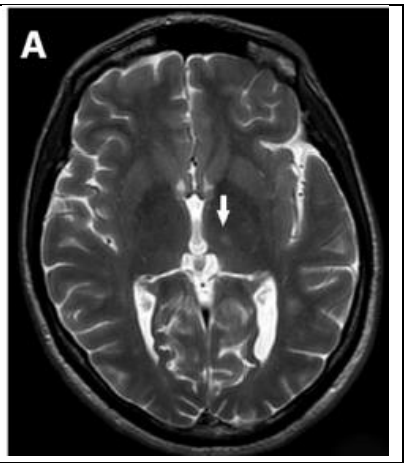
Introduction:

Wilson disease, also known as hepatolenticular degeneration, is an autosomal recessive condition in which the body accumulates too much copper. It predominantly affects the liver and the brain's basal ganglia, although it can also impact other organ systems.[1] Multiple sclerosis (MS) is a central nervous system (CNS) autoimmune disease marked by inflammation, demyelination, gliosis and neuronal death. Perivascular lymphocytic infiltrates and macrophages degrade the myelin sheaths that cover neurons.[2] The co-occurrence of these two is nevertheless regarded extremely unusual and can cause diagnostic challenges. In addition, comorbidity has a significant impact on MS quality of life and disease progression, making timely detection and treatment of these illnesses crucial.[3]

The co-occurrence of the two diseases appears to be more of a coincidence than a shared pathophysiology of common factors.

Case Presentation:

The patient is a 33-year-old woman previously in 2013. She had been diagnosed with hyperprolactinaemia and was treated with bromocriptine. There were no significant immunological or genetic problems found during investigations on family history. She had limb ataxia and tactile sensory loss in both legs, as well as hand and foot paresthesia, anxiety and dysthymia. Multiple periventricular white matter lesions were seen on cranial MRI, including a gadolinium-enhancing lesion in the left occipital region, a juxtacortical lesion in the left frontal lobe, and a lesion in the left thalamus. High IgG index (2.31; elevated when > 0.67) and oligoclonal bands (OCB) were seen in the cerebrospinal fluid (CSF). The values of cobalamine and folate were within normal limits. Retrospectively, the CSF neurofilament light chain level was 1904 pg/ml. The use of parenteral methyl-prednisolone treatment was started, and the neurological symptoms improved. The diagnosis of multiple sclerosis was made based on the clinical presentation and diagnostic results, and the criteria for dissemination in space and time were met in line with the McDonald Criteria of 2010. A follow-up MRI exam in 2014 revealed left optic neuritis with no evidence of white matter lesions progressing. Without clinical indications, visual evoked potentials (VEP) exhibited greater delay on both sides. Antibody levels suggestive of systemic immunological disorders with possible CNS involvement were evaluated in 2015 and determined to be normal. A possible infectious aetiology was ruled out by serologic studies. A follow-up MRI exam in 2014 revealed left optic neuritis with no evidence of white matter lesions progressing. Without clinical indications,



Brain MRI scan upon first admission. Axial T2 scan shows a lesion in the left thalamus (full white arrow).

visual evoked potentials (VEP) exhibited greater delay on both sides. Antibody levels suggestive of systemic immunological disorders with possible CNS involvement were evaluated in 2015 and determined to be normal. A possible infectious aetiology was ruled out by serologic studies. In 2018, a recurrence appeared, creating new, fluctuating sensory issues in both legs, and MRI scans revealed many unique lesions in the spinal cord and one in the right parietal lobe, after 5 years without clinical symptoms of disease activity. In 2019, immunomodulatory therapy was started, and it was shown to be effective in avoiding relapses with no concerns with adherence or tolerance. Due to constipation and flatulence, a full gastrointestinal examination was undertaken in 2013. Anxiety, avoiding behaviour, and eating issues were also present. Because of the elevated liver enzymes in the blood, additional tests were undertaken to rule out an infectious or immunological aetiology, all of which came out negative. The possibility of Wilson's disease was raised during 2014 gastrointestinal follow-up. She was admitted to the hospital in 2015 with significant lower limb edoema, hypoproteinemia, anaemia, and ascites. The findings of laboratory tests indicated hepatic decompensation. Hepatomegaly and ascites were seen on an abdominal ultrasonography. Extended virological testing came up negative. Chronic hepatic cirrhosis with copper accumulation was discovered by a liver biopsy. Heterozygosity for the H1069Q gene was discovered through genetic testing. ATP7B gene mutations R778Q and R778Q, both of which cause Missense point mutations are considered pathogenic. Wilson's disease was diagnosed based on these findings. During this period, severe hepatic decompensation, blood transfusions and parenteral nutrition oral diuretic therapy and albumin supplements were required. Penicillamine was then taken orally. The patient was started on therapy, which she is continued until the present day. This therapy resulted in the patient's liver function slowly returning to normal and a considerable improvement in her symptoms, including the edoema and ascites, as well as an increase in her attitude and appetite. A hepatologist was on hand to keep an eye on the patient. liver function, blood testing, and abdominal ultrasound, as well as clinically by a neurologist. In 2021, the patient showed MRI progression (new lesions), as well as a recurrence with loss of vibration in both lower limbs, requiring cladribine medication intensification.

Discussion:

This research looked at a patient who had both MS and Wilson's illness. Wilson's illness is known to induce a variety of CNS abnormalities, the most prevalent of which include involvement of the basal ganglia, thalami, and brainstem, as well as the cerebral white matter, especially the corpus callosum. Wilson's disease may have caused some of the white matter lesions in this case, although gadolinium enhancement in the left occipital area on admission, as well as spinal cord involvement after the second relapse, are not characteristics of Wilson's disease.[3] Yetkin et al. described a case of 44-year-old man who was first diagnosed with MS

and then developed WD, which included pathological abnormalities in the basal ganglia that are characteristic of Wilson's disease.[4]

Conclusion:

This incident is similar to those previously reported. The two disorders' co-occurrence appears to be more of a coincidence than a shared pathophysiology of common causes, however they are considered to impact each other. When it comes to rare co-occurrences like this one, each new case is crucial because it allows for a more thorough analysis and understanding of the clinical symptoms that are more prevalent in these cases, aiding in the prediction of disease course as well as treatment possibilities.

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4. A Case Report of Delayed Hydrocephalus Detection in Corticobasal Degeneration Following Mildly Traumatic Subarachnoid Hemorrhage

Introduction:

Neurodegenerative illnesses presenting with parkinsonism have become a worldwide concern as life expectancies have increased. CBD (corticobasal degeneration) is a tauopathy. that is an uncommon neurological illness. Parkinsonism, apraxia, aphasia, and cognitive impairment are some of the neurological symptoms that patients with CBD experience.[1] Blood accumulates between the arachnoid and the pia mater, causing subarachnoid haemorrhages, which can be fatal.[2] Due to frequent falls, CBD patients may be at a higher risk of traumatic SAH-induced hydrocephalus. This case emphasises the need of being cautious about traumatic SAH-related hydrocephalus in patients with parkinsonism who are at high risk of falling and having delayed identification of posttraumatic sequelae, especially if they have aphasia like CBD.[1]

Even in patients with moderate traumatic SAH, clinicians must pay close attention to brain injury in patients with parkinsonism who are at a high risk of repeated falls

Case Presentation:

When she was 60 years old, a 65-year-old woman started having trouble speaking. The motor aphasia symptoms become more severe with time. At the age of 64, the patient had difficulties understanding discussions. She went to a clinic where she was given haloperidol and biperiden. She had gait disturbance at the same time. She had a fall three months after beginning her therapy and suffered brain injuries as a result. The following day, the patient fell for the second time. The patient was taken to the hospital two days following the second fall for gait abnormalities and repeated falls. She had akinesia, axial and left-side dominant limb stiffness, and postural abnormalities, all of which were symptoms of parkinsonism. A computed tomography scan of the brain indicated a subdural hygroma and moderate intraventricular bleeding. Mild SAH with intraventricular haemorrhage was discovered using a susceptibility-weighted picture of brain magnetic resonance imaging. The size of the ventricles was normal. Her brain MRI data revealed that her brain damage was minor. Despite the termination of antipsychotic medicines and the start of therapy, gait disturbance and hypo-responsiveness progressively worsened. She was moved to hospital a month later. A physical examination performed upon admission indicated no untoward results. The patient had significant motor aphasia on neurological testing, and her Glasgow Coma Scale (GCS) score was E4VAM5. She was unable to follow verbal directions to the letter. The patient has a little restriction in vertical eye movement. There was no sign of paralysis. The symptoms of Parkinson's disease, such as limb and trunk muscle stiffness and akinesia, were discovered. There were pyramidal signs such as hyperreflexia and abnormal reflexes, as well as a positive grasp reflex. She couldn't stand up or walk. There were no abnormalities found in the blood tests. Bacteria, fungus, and acid-fast bacilli cultures in the CSF were all negative . The EEG revealed diffuse slow waves with no paroxysmal activity. The presence of communicating hydrocephalus was detected by brain magnetic resonance imaging. The patient's neurological status did not improve after conservative therapy with oral L-DOPA at a maximal dose of 450 mg/day. On the 23rd day following admission to hospital, a ventriculoperitoneal (VP) shunt was implanted. Brain biopsy from the right parietal lobe at a place where a VP shunt tube is inserted to obtain a histological diagnosis of underlying disorders producing aphasia and parkinsonism. The ventricle's size was decreased. The neurological status of the patient did not significantly improve. Hematoxylin and eosin staining revealed inflated neurons in the brain disease. Gallyas staining revealed neuropil threads and coiled bodies. The neuropil threads and coiled bodies, as well as the cytoplasm of certain neurons, were positive for phosphorylated-tau immunostaining. The diagnosis of CBD as an underlying illness presenting with aphasia and parkinsonism was validated by these pathology results. She was sent to a long-term care facility . Her neurological state did not improve six months after the VP shunt was implanted.

Discussion:

Traumatic SAH-related hydrocephalus in this patient would be mostly linked with subacute neurological impairment following a fall that resulted in brain damage. In individuals with head

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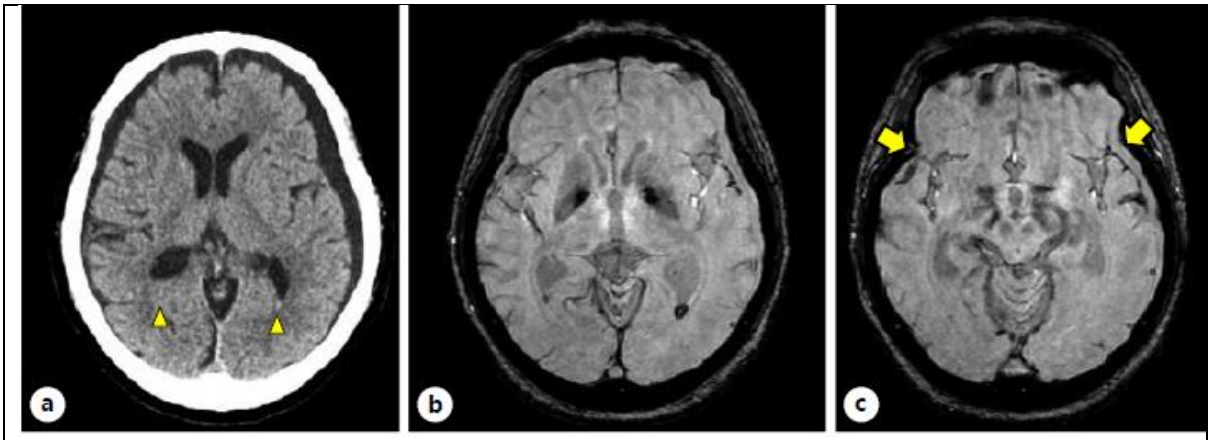
trauma, posttraumatic hydrocephalus is a common consequence.[1] Individuals with traumatic SAH have a greater chance of developing posttraumatic hydrocephalus than patients without traumatic SAH.[3] In individuals with traumatic SAH-related hydrocephalus, underlying parkinsonism may be a risk factor for poor response to VP shunts.[1]



Brain images. Brain CT 3 days after brain trauma. Arrowheads show mild intraventricular hemorrhage (**a**). SWI of brain MRI 5 days after brain trauma reveals mild SAH with intraventricular hemorrhage (**b**). Arrows show a small amount of SAH around the Sylvian fissure (**c**). FLAIR sequence image detects normal ventricular size

Conclusion:

After a traumatic SAH with intraventricular haemorrhage, a CBD patient developed hydrocephalus and decreased neurological function, according to the researchers. Even in patients with moderate traumatic SAH, clinicians must pay close attention to brain injury in patients with parkinsonism who are at a high risk of repeated falls. In CBD patients, not only parkinsonism but also aphasia may cause a delay in identifying brain trauma-related issues.

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