Vol 4 | Issue 15 | 2023

Interesting cases NEUROLOGY

Neurology Case Series

Vol 4 | Issue 15 | 2023

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 rare case report of epilepsy-related open-lip schizencephaly with an arachnoid cyst.**
2. **Case report of synovial sarcoma of the tibial nerve, a rare tumour in an unusual site.**
3. **A case report of Cortical hemichorea-hemiballismus syndrome as a symptom of hypoglycemic encephalopathy.**
4. **A case report of a clinically silent gastrointestinal bleeding after dysautonomic spinal cord injury.**

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,

MICRO LABS SYNAPSE
Vol 4 | Issue 15 | 2023
Interesting cases NEUROLOGY

1. A rare case report of epilepsy-related open-lip schizencephaly with an arachnoid cyst

Introduction:

Schizencephaly is a rare congenital neuronal migration disorder characterised by the presence of a full-thickness cleft, lined with heterotopic grey matter and filled with cerebrospinal fluid (CSF), which connects the pial surface of the cerebral hemisphere with the ependymal surface of the lateral ventricle.[1] According to certain theories, the traction effect and splitting of the leptomeninges caused by schizencephaly are the causes of arachnoid cyst formation.[2] An electroencephalogram showed an epileptic form in the right temporal lobe in this case of an 18-year-old male who had a history of multiple complicated partial seizures during the previous two months.Right temporal fossa arachnoid cyst and a grey matter-lined right temporal cleft indicative of open-lip schizencephaly

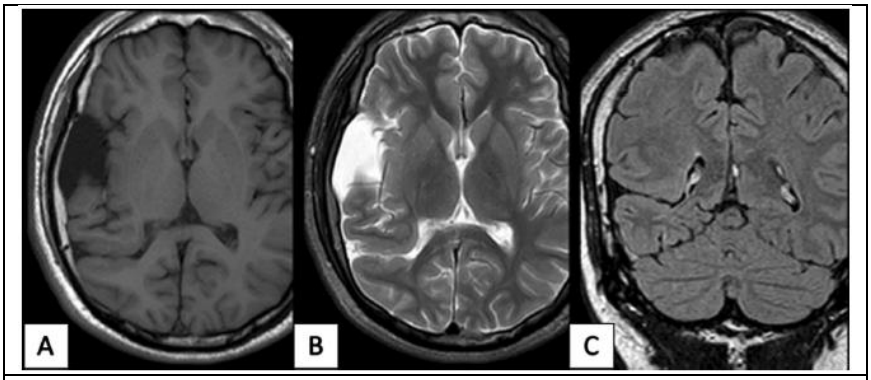
Schizencephaly with a new onset of seizures in an adult is extremely rare.

Case presentation:

A male patient, age 18, was referred to the hospital after experiencing two months of complex partial seizures. His initial seizure lasted a few seconds and was an absence seizure. He had fallen two weeks previously, hitting the ground with his right head. After then, he did not receive any hospital treatment. A focal clonic seizure in the left arm was the second event. The third episode was a tonic-clonic seizure with left arm convulsions lasting 10 minutes, followed by a period of unconsciousness and retrograde amnesia. He never experienced any headaches or nausea. He had never before exhibited these symptoms. Yet his father has a history of febrile seizures. He has no prior post-natal complications or widespread delays in achieving developmental milestones, and he is proportionately athletic. Has some degree of limitations on the Montréal Cognitive Assessment (MoCA) scoring during neurocognitive testing, with a total score of 22, interpreted as mild cognitive impairment. Antero-posterior transverse cranio-caudal brain magnetic resonance imaging (MRI) with contrast revealed a right temporal fossa extra-axial cystic lesion measuring ±8.17×3.46×6.27 cm that had a mass effect on the surrounding brain parenchyma and a slight leftward subfalcine herniation. Moreover, a right temporal cleft lined with GM and filled with CSF was visible on the MRI. It extended from the right lateral ventricle to the cortex. These results are consistent with schizencephaly and an arachnoid cyst. An epileptic form was indicated by electroencephalography (EEG) in the right temporal lobe. After performing a cystoperitoneal shunt , 750 mg of levepiracetam were administered twice daily. The patient had a right side facial focal seizure five days later. He had an absence seizure a month later and was treated with 1000 mg of levetiracetam twice a day and 2000 mg of sodium divalproex once daily.

MICRO LABS SYNAPSE
Vol 4 | Issue 15 | 2023
Interesting cases NEUROLOGY

Discussion:



Axial T1W (A), Axial T2W (B), and coronal FLAIR (C) Brain MRI showed grey matter-lined right temporal cleft extending from the right lateral ventricle to the cortex and filled with cerebrospinal fluid, these findings are consistent with open-lip schizencephaly

Arachnoid cysts, with the exception of open-lip schizencephaly, rarely cause symptoms and are often overlooked.[2] Arachnoid cysts, an intracranial lesion filled with CSF and surrounded by arachnoid membranes, make up just 1% of all intracranial space-occupying lesions (SOL), which are typically asymptomatic and discovered accidentally.[3] This case showed how schizencephaly could result in an arachnoid cyst that was very large and continued to pressure on the surrounding tissue, causing seizures.

Conclusion:

Schizencephaly with a new onset of seizures in an adult is extremely rare. In this case, enlarged arachnoid cysts caused epilepsy in the open-lip schizencephaly due to space-occupying lesions (SOL). Following a cystoperitoneal shunt to minimise the size of the arachnoid cyst and the administration of two anti-epileptic medications, the seizure episode was successfully resolved.

References:

1. Veerapaneni P, Veerapaneni KD, Yadala S. Schizencephaly. [Updated 2022 Oct 14]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; Jan 2022
2. Hapsari P, Nagpal C, Shen R. Epilepsy-associated open-lip schizencephaly with arachnoid cyst: a rare case report. The Egyptian Journal of Neurology, Psychiatry and Neurosurgery. 2023 Dec;59(1):1-5.
3. Mustansir F, Bashir S, Darbar A. Management of arachnoid cysts: a comprehensive review. Cureus. 2018;10: e2458.

2. Case report of synovial sarcoma of the tibial nerve, a rare tumour in an unusual site.

Introduction:

Synovial sarcomas (SS), which constitute 5–10% of all soft tissue sarcomas (STS), are a distinct subgroup of STS. The comparatively early onset of diagnosis and clinical presentation distinguish synovial sarcoma distinct from other STS. The pathognomonic t(X;18) chromosomal translocation and subsequent development of the SS18:SSX fusion oncogenes

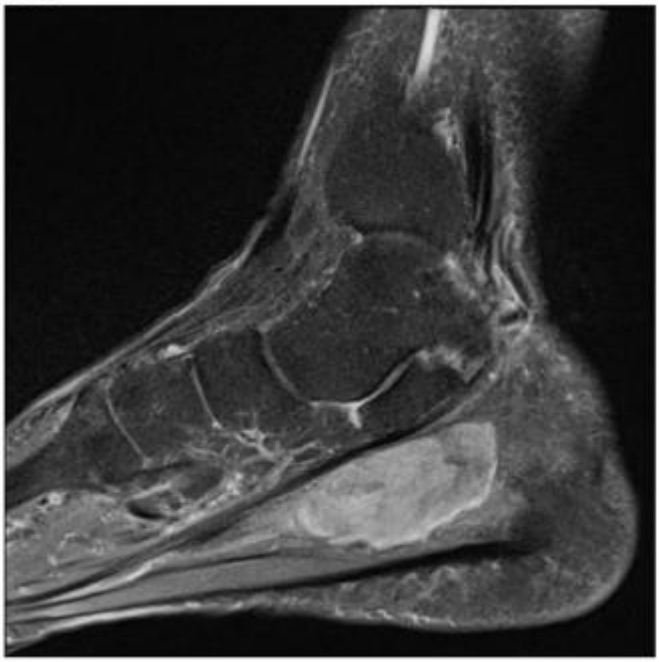
MICRO LABS SYNAPSE
Interesting cases NEUROLOGY
Vol 4 | Issue 15 | 2023

are responsible for the distinctive genomic features of synovial sarcomas.[1] There have only been 4 cases reported in the literature so far of patients with SS arising from the tibial nerve.[2] Here, researchers describe the instance of a patient whose limb was preserved following treatment for a rare synovial sarcoma (SS) of the tibial nerve.

This case report highlights a rare kind of malignant nerve tumour that might resemble benign peripheral nerve sheath tumours both clinically and radiologically

Case Presentation:

A 71-year-old woman complained of increasing pain under her left inner ankle, which corresponded to the sensitive innervation area of the tibial nerve and had a projection to her sole and fingers. She struggled to touch the region surrounding her ankle because of intense pain.I t was getting worse when she walked . The patient reported pain score was around 8 in a numerical scale (Numeric Rating Scale, NRS) with a range of 0- 10, or 0- 100, where "+" implies "no felt pain" and 10 (or 100) represents "worst conceivable suffering". The patient has so far received treatment for gastroduodenal ulcer illness, diabetes mellitus type II, bronchial asthma, arterial hypertension, and right nephrectomy for clear cell cancer in 1985. In 1999, she underwent a hysterectomy with an adnexectomy for myomatous uterus. The mother of the patient passed away from an unidentified gynaecological cancer. Other than that, the family history was cancer-free. Social history and work history were unimportant. Following the first neurological evaluation, the patient was referred for an ankle ultrasound, which revealed a lobular tumour measuring 50× 22 ×16 mm. The discovery after incorporating magnetic resonance imaging appeared to be a suspicious neurinoma of the tibial nerve.Without performing a biopsy first, the tumour was surgically removed. It was a 50 mm by 20 mm tumour that was removed from the distal part of the tarsal canal. It had grown through the tibial nerve's structure and in some places into the surrounding tissue, and it had intraoperatively taken the form of a neurofibroma. Radical resection of the tumour and liberation of the nerve fascicles from it. According to histology, it was a spindle cell mesenchymal affection with cells arranged in ill-defined fascicles and a variable collagenized stroma. The tumour had cells with a uniform appearance, nuclei with fine chromatin, tiny nucleoli, no substantial cytological atypia, and clearly discernible mitotic activity. This monophasic synovial sarcoma was identified based on the mitotic activity that was revealed in the tumour. The resection margin was approached by tumour structures, however the tumour sample was



Preoperative MR examination of the left ankle. Imaging in the sagittal plane (PD TSE sequence) with the finding of an oval-shaped soft tissue tumorous mass along the course of the tibial nerve

fragmented, making it impossible to accurately determine the resection margin. After that, a control MRI was done. No nodular solid tumour residual was discovered, but postoperative alterations, edoema, and scarring were visible on the medial side of the foot. The patient was advised to have a large reexcision of the skin with the subcutaneous tissue of size 91× 20 ×15 mm in January 2022 because to the histological results and potential microscopic residue. Histologically, the margins of the resection were free of the sarcoma formations. The body's CT revealed no evidence of disease spread. The patient's suffering was greatly reduced when the tumour was removed. An electromyographic test was completed 2 months after the second surgery, and a low summation muscle potential (CMAP) amplitude of the left tibial nerve (1 mV) was recorded. The results were otherwise normal, pointing to a partial axonal lesion of the tibial nerve. External radiation is now being used to treat the patient's tumour bed. The tibial paresis is not adequately expressed. The wounds from surgery recovered quickly.The patient is now attempting to walk using her feet.

Discussion:

Malignant mesenchymal tumours, or tumours originating from connective tissue, are synovial sarcomas. The term "synovial sarcoma" is misleading, nevertheless, as the tumour really develops from primitive mesenchymal cells rather than synovial cells.[3] The location surrounding the large joints on the limbs, more frequently on the lower ones, around the knee joints, is where the term most likely came from. A magnetic resonance imaging examination serves as the base for the diagnosis, and then biopsy should be carried out before to surgery.[2]

Conclusion:

This research sheds light on a rare kind of malignant nerve tumours that can mimic benign peripheral nerve sheath tumours both clinically and radiologically. In extremely painful resistances, which are often seen around the joints of the lower limbs and can expand gradually, synovial sarcoma should be taken into consideration. A prompt diagnosis utilising MR imaging and a biopsy with a histological evaluation is required in order to start therapy as soon as possible because the tumor's size is a poor prognostic indicator. To ensure that the tumour is sufficiently radical and does not require a second operation, as was the case with this patient, surgery should only be done after a biopsy with histological examination of the tumour.

References:

1. Gazendam AM, Popovic S, Munir S, Parasu N, Wilson D, Ghert M. Synovial Sarcoma: A Clinical Review. Curr Oncol. 2021 May 19;28(3):1909-1920.
2. Hemerková P, Matulová H, Vališ M, Soukup J, Kanta M, Jandura J. Synovial sarcoma of the tibial nerve - case report of a rare tumor in a rare location requiring early diagnosis. BMC Neurol. 2023 Feb 10;23(1):65.
3. WHO Classification of Tumours Editorial Board. Soft tissue and bone tumours. WHO classification of tumours series, vol. 3. 5th ed. Lyon: International Agency for Research on Cancer; 2020. p. 290–3.

3. A case report of Cortical hemichorea-hemiballismus syndrome as a symptom of hypoglycemic encephalopathy

Introduction:

MICRO LABS SYNAPSE
Vol 4 | Issue 15 | 2023
Interesting cases NEUROLOGY

Hemichorea and hemiballismus are examples of hyperkinetic motions that are well-known to be caused by hyperglycemia. Hypoglycemia can result in comparable clinical problems, despite its rare. The literature only includes a few cases of hypoglycemia hemichorea, most of which present with negative brain MRI results.[1] Hemichorrea-hemiballismus(HCHB) is an uncommon neurologic condition occurring in acutely decompensated type 2 diabetes mellitus (T2DM) . HCHB is most frequently linked to cerebral vascular accidents with an infarct or hemorrhagic aetiology. It is a disorder that can affect a diabetic patient.[2] The uncommon but well-known causes of chorea and ballismus, as well as hyper- and hypoglycemia conditions, are typically linked to brain lesions in the basal ganglia.[1] Researchers describe a case of hemichorea-hemiballismus (HCHB) syndrome in a 71-year-old man with poorly managed type 2 diabetes mellitus that followed a severe hypoglycemia episode.

This case may demonstrate that some individuals with cortical lesions have hypoglycemia as their underlying cause.

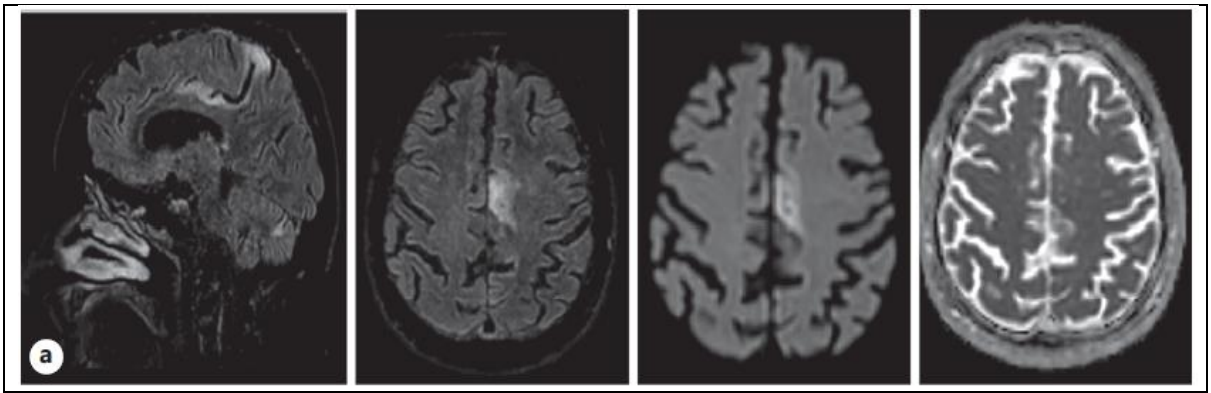
.

Case Presentation:

A 71-year-old man was discovered unconscious by his family and brought to the hospital's emergency room. His blood glucose level was very low and unmeasurable when we arrived. He received an intravenous infusion of a 33% glucose solution, which raised his blood sugar level to 200 mg/dL. He was able to regain consciousness three hours after being admitted. At the age of 58, the patient was diagnosed with a type 2 diabetes mellitus diagnosis. His hypoglycemic treatment had recently been changed by a diabetologist, who substituted slow-release insulin for metformin, which was poorly tolerated due to gastrointestinal side effects, and a regimen of fast-release insulin administered three times daily. For a minor supraaortic trunk vasculopathy, he was also on low-dose aspirin. He had no prior history of seizures, neurological illnesses, or movement abnormalities. During the patient's second day in the hospital, hyperkinesias of the right side of the body manifested as intermittent, ballistic movements of his right limbs, combined with infrequent distal choreic movements, according to the physicians. They were nearly constant, made worse by stress and activity, and subsided whenever slept. The patient was always aware of them and upset by them. Other than that, the neurological evaluation was normal, including the cognitive and cortical sensory tests. An EEG was recorded immediately after on the possibility of epileptic focal seizures, however it only revealed left-hemispheric background slowing and no epileptiform discharges. Nonetheless, the patient was administered levetiracetam 1,500 mg i.v. bolus, diazepam 10 mg i.v. bolus, diazepam 20 mg continuous 24 h infusion, and levetiracetam 1,000 mg three times a day. On the third day, levetiracetam was changed to valproate 400 mg three times a day and diazepam was stopped, but there was still no change in the patients' bowel motions. Two further EEGs were recorded in the days that followed, the second during a hypoglycemia episode (glycemia 42 mg/dL), with nonspecific results, and the antiepileptic medication was gradually tapered

MICRO LABS SYNAPSE
Interesting cases NEUROLOGY
Vol 4 | Issue 15 | 2023

down. Two further CT brain scans were taken on the second and third days of the patient's stay, and both again revealed no acute abnormalities. An MRI of the brain was done three days after the hyperkinesias first appeared, and it revealed a cortical-subcortical T2 and T2-FLAIR-hyperintense left frontal lesion that extended from the precentral gyrus to the cingulate gyrus. Moreover, a lesion in the superior parietal lobule's paramedian cortical region was shown to be ipsilateral. While the ADC sequence was somewhat lowered and the T1 sequence was marginally hypointense, both lesions had limited diffusion on the DWI sequence. There was no impact on the basal ganglia. Also noted were diffuse chronic cerebrovascular disease and a prior lacunar infarct in the superior cerebellar vermis. A second MRI taken 11 days after admission revealed that the frontal lesion, particularly in its cingulate region, had not altered. Nevertheless, the cortical T2- FLAIR left parietal lesion extension had. Both locations' DWI hyperintensity was significantly decreased. Parallel to this, the patient's clinical circumstances steadily improved: 12 days after admission, the choreic-hemiballistic involuntary movements had significantly decreased, and his glycemic control had improved. These results led to the conclusion that a lumbar puncture was not required because the clinical suspicion of a hypoglycemia encephalopathy was significant. Finally, 15 days after being admitted, the patient was transferred to a rehabilitation centre, where, concurrent with the patient's glycemic control improving, the choreic-hemiballistic involuntary movements totally disappeared within 20 days of their commencement. His neurological examination, which included cognitive testing, was normal four and twelve months later, respectively, and two brain MRIs revealed that the cerebral lesions had completely disappeared. The patient had adequate glycemic control, and he denied that the hyperkinesias having recur, demonstrating that the abnormal movements were a direct result of the hypoglycemic episode.



Serial patient’s brain MRIs are shown in the picture. Respectively, T2-FLAIR (first two pictures), DWI, and ADC sequences are reported. In the first MRI (a), a cortical-subcortical left frontal lesion, involving the cingulate gyrus, and an ipsilateral paramedian, parietal cortical lesion are evident. Both lesions are hyperintense on T2, T2-FLAIR, and DWI sequences, while ADC is mildly decreased. No lesions were detected in the basal ganglia on T1, T2-FLAIR, DWI, and ADC sequences

Discussion:

MICRO LABS SYNAPSE
Interesting cases NEUROLOGY
Vol 4 | Issue 15 | 2023

Acute strokes that primarily affect the subthalamic and lentiform nuclei of the basal ganglia have been associated with symptomatic unilateral chorea/ballism in the past, with a decreased activity of the "indirect pathway" in the traditional basal ganglia circuitry scheme being proposed as the pathophysiology.[1]This model, which centres on the classic basal ganglia circuitry dysfunction as the only cause of hemichorea and hemiballismus, is now being called into question by a growing body of literature reporting cases of such hyperkinetic movements arising from ischemic strokes affecting the frontoparietal cortex .[3,4] Uncertainty surrounds the process through which cortical lesions cause hemichorea. The disruption of the "hyperdirect pathway," a circuit limiting movement in which cortical neurons' axons directly stimulate the subthalamic nucleus, has been proposed as a potential cause of hemichorea through disinhibition of the thalamus, which is typically suppressed by the subthalamic nucleus.[1]

Conclusion:

Although cortical circuitries may independently contribute to the pathophysiology of chorea and ballismus, this is the first evidence of a hypoglycemic aetiology for cortical HCHB syndrome.Also, this is the first case of hypoglycemia encephalopathy involving the cingulate gyrus. Lastly, this instance could illustrate that a subset of patients who have cortical lesions as a result of hypoglycemia

References:

1. Pasini F, Karantzoulis A, Fanella G, Brovelli F, Iacobucci D, Aprea V, Storti B, Santangelo F, Canonico F, Remida P, Ferrarese C, Brighina L. Hypoglycemic Encephalopathy Manifesting with Cortical Hemichorea-Hemiballismus Syndrome: A Case Report. Case Rep Neurol. 2023 Feb 7;15(1):24-30.
2. Ticona J, Zaccone V, Zaman U, Kashani D, Chung Z, McFarlane IM. Hemichorea-Hemiballismus as an Unusual Presentation of Hyperosmolar Hyperglycemic Syndrome. Am J Med Case Rep. 2020;8(6):159-161.
3. Cotroneo M, Ciacciarelli A, Cosenza D, Casella C, Dell'Aera C, Grillo F, et al. Hemiballism: unusual clinical manifestation in three patients with frontoparietal infarct. Clin Neurol Neurosurg. 2020 Jan;188:105612.
4. Strauss S, Rafie D, Nimma A, Romero R, Hanna PA. Pure cortical stroke causing hemichorea-hemiballismus. J Stroke Cerebrovasc Dis. 2019 Oct;28(10):104287.

4 A case report of a clinically silent gastrointestinal bleeding after dysautonomic spinal cord injury.

Introduction:

A rare but serious consequence of spinal cord injury(SCI) is gastrointestinal bleeding (GIB) .[1] Patients with traumatic spinal cord injuries (tSCI) commonly experience gastrointestinal bleeding (GIB), which is often fatal, especially in the period following of the injury.[1,2] Many risk factors, some specific to this patient population, frequently increase the risk of bleeding. Typically, symptoms like coffee-ground emesis, hematemesis, melena, or even

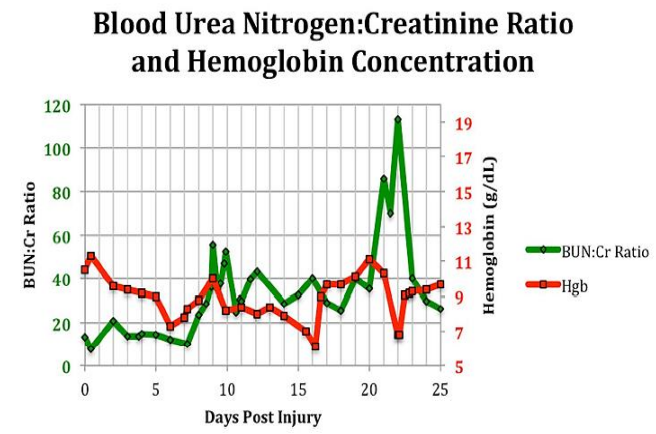
Dysautonomia can significantly delay the clinical presentation of GIB in the traumatic spinal cord injuries (tSCI) patients, making them more susceptible to disease.

MICRO LABS SYNAPSE
Interesting cases NEUROLOGY
Vol 4 | Issue 15 | 2023

hematochezia lead to a clinical suspicion of GIB. A decrease in haemoglobin may be a late sign. Nevertheless, patients with tSCI frequently develop dysautonomia and neurogenic bowels, which can delay the onset of symptoms and make it more difficult to diagnose GIB in a timely manner. [2] In the context of an acute cervical tSCI, researchers present a case of an almost clinically silent Gastrointestinal bleed.

Case Presentation:

After being involved in a high-speed car crash, a 21-year-old female who was previously healthy arrived at the hospital with acute cervical spinal cord transection at C7 level. She received early surgical decompression, which included a C6 laminectomy and a C4-T2 posterior spinal fusion, after being intubated upon arrival and being put on artificial ventilation. As per recommended standards of care for venous thrombotic event prevention, she started taking daily famotidine for gastrointestinal prophylaxis and was later given enoxaparin. Significantly, she tested positive for the COVID-19 strain of the SARS-CoV-2 virus at the time of admission, and a week later she had symptoms of COVID-19 pneumonia for which she was given a 10-day course of dexamethasone. A tracheostomy and a percutaneous endoscopic gastrotomy tube were both done two weeks after the incident. The latter surgery revealed normal duodenal and stomach anatomy. The patient's intermittent fever stopped 3 weeks after the injury, but she continued to be slightly anaemic for the duration of her time in the intensive care unit (ICU). The patient's BUN increased from 18 mg/dL to 43 mg/dL on day 21 of her



Line graph of patient BUN: Creatinine ratio alongside hemoglobin concentration, showing significant BUN: Cr spike

hospital stay, accompanied with a 2.5-fold rise in her BUN: Creatinine ratio, suggesting a potential GIB. Yet, there were no visible indications or symptoms of a recent or ongoing bleed. Following the BUN spike, haemoglobin initially remained steady for 25 hours, even on recheck, and no diaphoresis nor a change in stool colour were observed. General surgery examined the PEG tube and found no abnormal findings; stomach contents suctioning revealed only feeding formula that was the expected colour. Further confirming against a potentially significant GIB, the patient denied experiencing any stomach pain or a sensation of impending doom.The patient continued to be asymptomatic the next day, but a 3.5 g/dL decline in haemoglobin was detected, raising the possibility of GIB and necessitating a consultation with a gastroenterologist. After performing an esophagogastroduodenoscopy assessment, three duodenal ulcers which weren't bleeding and had clean bases in the duodenal bulb and sweep were found. During the surgery, a biopsy was taken for Helicobacter pylori, however the results were later interpreted as negative. At this moment, there are no visible indications of current ulcer bleeding. Omeprazole was suggested

MICRO LABS SYNAPSE
Interesting cases NEUROLOGY
Vol 4 | Issue 15 | 2023

for an eight-week treatment by gastroenterologist. Two units of packed red blood cells were given to her once as a transfusion (pRBCs). During afternoon, there were many bowel movements and significant volumes of melanotic faeces indicative of GIB. Her haemoglobin level increased at 9.1 g/dL by the evening, and she remained clinically stable. she was transferred to a rehabilitation facility of patients with spinal cord injuries. The patient was still ventilator-dependent when discharged.

Discussion:

This patient had a number of GIB risk factors. A recent procedure, the use of steroids, mechanical breathing, and prophylactic anticoagulation were among these. These risk factors are common among individuals with tSCI and are not specific to this patient. High dosage steroids have historically been used early in tSCI to protect neurological function. Dexamethasone was administered to this patient to treat COVID-19 pneumonia.[2] Nevertheless, a recent meta-analysis revealed that there may be a statistically insignificant link between the use of methylprednisolone and GIB. Generally, there is considerable discussion over the advantages and disadvantages of steroid treatment in tSCI.[3]

Conclusion:

Dysautonomia can considerably delay the clinical diagnosis of GIB in the majority of tSCI patients, making them more susceptible to disease. This case demonstrates the potential of a period during which the patient was bleeding, with a high BUN acting as the only warning sign. This case calls for increased awareness and monitoring of tSCI patients, as well as potential use of various strategies to lower the occurrence and improve early diagnosis of GIB in tSCI patients in order to ultimately lower the morbidity and mortality related to it.

Reference:

1. Cochrane P, Masri W, Silver J. The effects of steroids on the incidence of gastrointestinal haemorrhage after spinal cord injury: a case-controlled study. Spinal Cord. 2014 Jun;52(6):501.
2. Margo TE, McMullin PR, Kaddouh F. An interval of clinically silent gastrointestinal bleed in dysautonomic spinal cord injury: a case report. BMC neurology. 2023 Dec;23(1):1-5.
3. Sultan I, Lamba N, Liew A, Doung P, Tewarie I, Amamoo JJ, et al. The safety and efficacy of steroid treatment for acute spinal cord injury: A Systematic Review and meta-analysis. Heliyon. 2020;6(2):e03414.