MICRO LABS SYNAPSE

Interesting cases NEUROLOGY

Vol 3|Issue 9|2022

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. Immune thrombotic thrombocytopenia induced by covid-19 vaccine
2. Varicella Zoster Virus myelitis and polycranial neuritis presenting without rash: A Case Report
3. A case report on isolated abducens nerve palsy
4. A case study on eosinophilic granulomatosis with polyangiitis (churg-strauss syndrome) mimicking guillain-barre syndrome

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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Immune thrombotic thrombocytopenia induced by COVID-19 vaccine

Vaccine induced immune thrombocytopenia (VIIT) is a new syndrome which characterized by thrombocytopenia and thrombosis occurs at 4-30 days after the first dose of vaccination. This is seen in several COVID-19 vaccines.[1, 2] Many cases of this kind had thrombosis at rare sites including cerebral venous sinuses or in the portal, splanchnic or hepatic veins and some cases were seen with deep venous thrombi, pulmonary emboli or acute arterial thromboses.[1] This case demonstrates the rapid progression of the cerebrovascular thrombosis in VIIT involving both the both arterial and venous systems, requiring mechanical thrombectomy.

The initial presentation of VITT may be with mild symptoms of headache and fever, and with normal examination and investigations

Case Presentation: A woman aged around 54 years came to the hospital emergency department with complains of occipital headache, photophobia, fever and abdominal pain 7 days after receiving her first dose of the COVID vaccine. She is known case of Type 2 Diabetes mellitus and remote right nephrectomy, was on treatment with 1gm Metformin and 50mg Sitagliptin two times a day. Her Body Mass Index (BMI) was 31.5. Her examination and routine investigation looked normal (Table:1).

She was reassured and sent back and asked to revert to the hospital if the symptoms persist or worsen. After 4 day she reverted with the complaints of marked increase in headache associated with vomiting, diarrhoea and left calf pain. Patient was seen alert with normal neurological examinations and her blood tests revealed low platelet count 19×10^9/L, raised D-dimer >20 mg/L and CRP of 71 mg/L. The heparin/anti-PF4 antibody assay was strongly positive.

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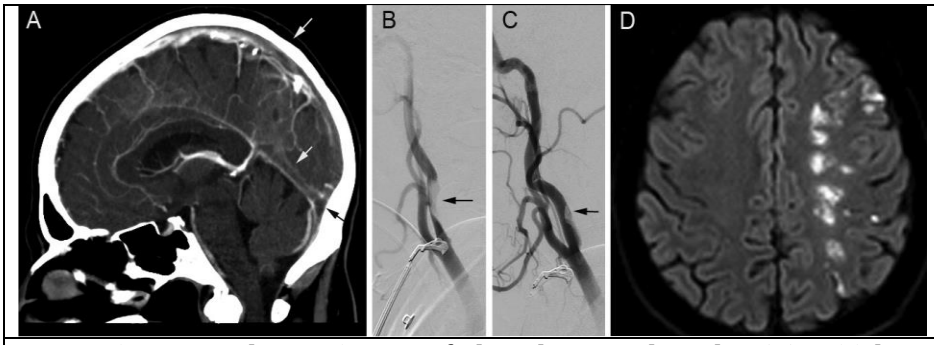

CT, angiogram and MRI images of thromboses and stroke. (A) Initial CT venogram demonstrating thromboses in superior sagittal and straight sinuses and torcula (arrows). (B, C) Angiograms during mechanical thrombectomy show near occlusive thrombus in the left internal carotid artery (B) and partially occlusive thrombus in the right internal carotid artery (C). (D) MRI (diffusion weighted image) shows an internal watershed infarct in the left hemisphere.

CT venogram showed widespread venous sinus thrombosis of the superior and inferior sagittal, bilateral transverse and left sigmoid sinues, and vein of Galen. She was diagnosed with VITT-related cerebral venous sinus thrombosis and was started on subcutaneous fondaparinux 7.5 mg daily and intravenous immunoglobulins 2 g/kg divided over 2 days.

The next day she developed an asymmetric quadriplegia and aphasia. CT head and CT angiogram illustrated new bilateral cervical internal carotid artery (ICA) thrombi near-occlusive on the left and partially occlusive on the right. She underwent stent-retriever mechanical thrombectomy of bilateral cervical ICA, and superior sagittal and transverse venous sinuses. Left hemispheric internal watershed infarcts and a small right cerebellar venous haemorrhage were seen in follow up MRI. She developed right hemiparesis and expressive dysphasia next day. Fondaparinux was replaced with intravenous bivalirudin infusion and she was put on a 5-day course of intravenous methylprednisolone, followed by a short tapering course of oral prednisolone. Platelet counts became normal and there was also improvement in the D-dimer after 5 days of her admission.

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		First presentation	Admission date	Thrombectomy	Post-thrombectomy	
Tests	Normal values	17/05/2021	20/05/2021	21/05/2021	22/05/2021	24/05/2021
Haemoglobin (g/L)	115–160	147	154	124	105	110
White cell count (10^9/L)	4.0–11.0	10.47	6.81	8.63	8.6	13.13
Platelets (10^9/L)	150–400	170	19	12	33	206
D-dimer (mg/L)	<0.5		>20			14.23
PT (s)	12–16.5		16.8	16.1	20.6	20.9
APTT (s)	27.5–38.5		31.7	28.3	52.9	53.3
Fibrinogen (g/L)	2.0–4.0		2.6	2.4	2	2.1
Sodium (mmol/L)	135–145	135	140	136	137	137
Potassium (mmol/L)	3.5–5.2	3.8	4.1	4.3	3.7	3.5
Urea (mmol/L)	3.0–8.0	6.5	5.3	6	6.2	6.6
Creatinine (µmol/L)	45–90	59	57	51	51	54
eGFR	>60 mL/min/1.732 m2	>90	>90	>90	>90	>90
CRP (mg/L)	<5		71			
Bilirubin (µmol/L)	<20	21	13			8
ALP (U/L)	30–110	76	83			61
PF4 antibodies ELISA			Strongly positive			
Lipase (U/L)	<60		11			

ALP, Alkaline Phosphatase; APTT, Activated Partial Thrmboplastin Time; CRP, C-Reactive Protein; eGFR, estimated Glomerular Filtration Rate; PF4, Platelet Factor 4; PT, Prothrombin Time.

Table 1: Hematological Laboratory Investigations

She underwent emergent stent retriever mechanical thrombectomy and in addition she received other treatment including intravenous immunoglobulin and intravenous and oral steroids. Outpatient rehabilitation including physiotherapy and speech therapy were given. One month after thrombectomy, her Dysphasia resolved but she had right hemiparesis and mild to moderate weakness of right upper and lower limbs .she was able to stand and walk with support and able to dress and feed herself. Her anticoagulation continued with apixaban 5 mg two times per day and no further complications seen.

The pathogenesis of the VIIT syndrome is not yet fully known, but patients with high levels of antibodies to platelet factor 4 (PF4)–polyanion complexes by ELISA should be identified. This serology pattern is similar to findings in patients with ‘atypical’ or ‘autoimmune’ heparin-induced thrombocytopenia, in whom thrombi develop in the absence of known previous exposure to heparin.

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This study tells that, the initial presentation of VITT may be with mild symptoms of headache and fever, and with normal examination and investigations. Thrombosis may progress rapidly even after institution of non-heparin anticoagulation and immunotherapy, mechanical thrombectomy should be considered urgently for significant cerebral arterial and venous thromboses.

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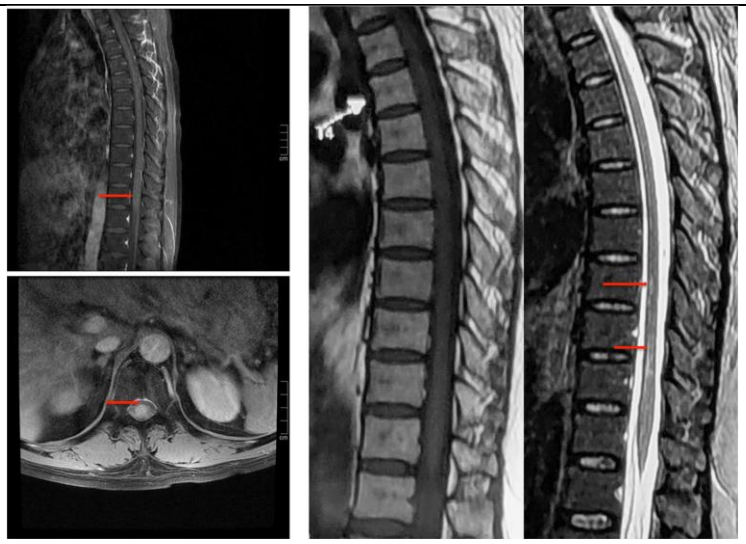
Varicella Zoster Virus myelitis and polycranial neuritis presenting without rash: A Case Report

Proper attention to be given towards various symptoms caused by VZV infection due to the clinical heterogeneity, especially in the absence of cutaneous lesions.

Varicella Zoster Virus (VZV) is neurotropic DNA virus that is responsible for causing chicken pox and shingles. VZV at the initial stages causes chicken pox and presumably becomes latent for many years in ganglionic neurons and adrenal glands.[1] Reactivation of VZV causes many complicated neurological diseases like meningitis, encephalitis, cerebral vasculopathies, cranial nerve inflammation, Guillain-Barré syndrome (GBS), other peripheral nervous system (PNS) diseases, and postherpetic neuralgia (PHN).[1] It is often difficult to connect neurological syndromes to VZV in the absence of the cutaneous lesions. There is no clear epidemiological data regarding VZV vasculopathy and myelopathy and is generally considered as <10%.[2] VZV myelopathy is a severe and commonly seen in immunocompromised patients. Here is a case of VZV myelitis and polycranial neuritis presenting without presence of any rash.



Spinal MRI revealed a hyperintense T2 lesion in the spinal cord extending from T8 to T11, and enhancement MRI indicated small patchy and strips hyperintensities in the spinal cord and meninges. The red arrow refer to the injuries

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Case study: A 52-year-old man presented to with complaints of headache, fever, right lower limb weakness, abdominal distension, and loss of hearing. Patient was healthy before without any medical history. Patient was physically examined; he was alert and oriented to time place and person .Bilateral peripheral facial paralysis and presence of hearing loss suggested that his bilateral facial nerves and vestibulocochlear nerves were affected. The medical research council (MRC) grade 3/5 indicated weakness of left lower limb. There was marked decrease in the puncture sensation on the left side of the body below T6 level. Patient was also suspected to have spinal cord involvement. Patient was suspected to have neurological infection due to the presence of multiple injuries of cranial nerves and spinal cord. CSF analysis showed increase in the lympochyte count and a slight increase in protein. MRI indicated mild changes in the white matter, Cranial MRI was normal and T2 weighted MRI showed the presence of a hyperintense lesion in the spinal cord extending from T8 to T11.

Metagenomic next-generation sequencing (mNGS) we done which showed significant increase in VZV DNA and IgG in cerebrospinal fluid (CSF). VZV-specific intrathecal antibodies test was also positive. Hence confirmed polyneuritis cranialis and myelitis were due VZV infection. The abdominal distention was secondary to segmental paresis caused by VZV infection. The patient was treated with combination of intravenous ganciclovir 5 mg/kg q12hr and 5 mg/d dexamethasone therapy. The patient's symptoms improved gradually except Urinary retention.

Many studies reported that the Reactivation of latent VZV on the ganglia can cause different neurological disorders like herpes zoster, meningitis or meningoencephalitis, cerebellum encephalitis, monocranial nerve palsy, polycranial nerves palsy, vascular disease, myelopathy and various other important ocular inflammatory diseases.[3] This case manifested a multiple neurological disorders without rash.

VZV infection may be severe, early as well as accurate diagnosis of VZV is important. Early diagnosis and treatment of VZV can reduce the disability and mortality. Proper attention to be given towards various symptoms caused by VZV infection due to presence of clinical heterogeneity, especially when there is absence of cutaneous lesions. Hence, complete and detailed examinations should be done to avoid misdiagnosis or missed diagnosis and provide suitable treatment to the patients.

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A case report on isolated abducens nerve palsy

The abducens nerve (Cranial VI) is responsible for ipsilateral eye abduction Dysfunction of the abducens nerve can occur at any point of its transit from the pons to the lateral rectus muscle, resulting in sixth nerve palsy. [1]The most common ocular motor nerve palsy is identified to be Abducens nerve palsy. In terms of isolated ocular palsy, abducens nerve is the most common nerve affected among cranial nerves. The etiologies of cranial nerve palsy include traumatic, vascular, neoplastic, inflammatory, infectious, demyelination and idiopathic. Subjects who are suffering from vascular comorbidities like diabetes and hypertension, microvascular ischemia is presumed to be the leading cause in adults above the age of 50. Incidence of idiopathic cases continues to rise despite its prevalence and progression made in neuroimaging.

The most common isolated ocular nerve palsy is the Abducens nerve palsy

A 31-year-old male patient presented to the emergency department with sudden onset diplopia and right sided frontal headache since 3 days. The patient was not a known case of any medical condition. Over the right frontal and retroorbital area the headache was observed to be constant which was radiating to the nasal side of the right eye, throbbing in nature and was not aggravated with sound, light and was not associated with redness or lacrimation. The subject did not have any history of fever, stiffness in the neck, phonophobia, pulsate tinnitus, seizures, nausea and vomiting. Upon right lateral gaze the horizontal diplopia was more prominent and was resolved on covering one eye. The left lateral vision was not affected. Pain was not present during the eye movement. For the first time headache and horizontal diplopia began simultaneously. Physical examination revealed right lateral rectus palsy and left sided horizontal nystagmus. There was no proptosis, ptosis or facial numbness present. Visual acuity, other cranial nerves and upper and lower motor neuron examinations were normal.

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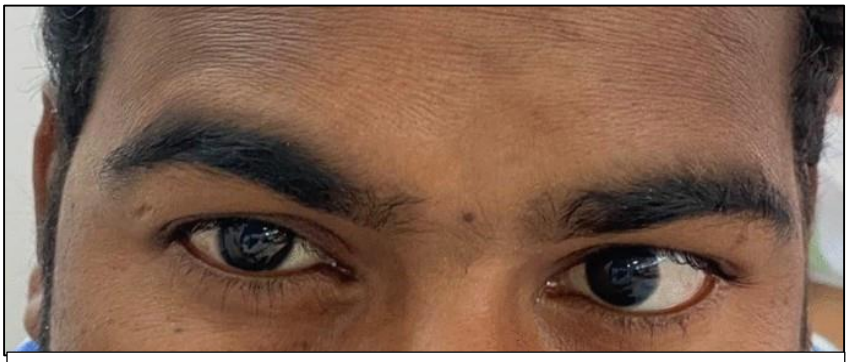

Figure 1 : Right Abducens nerve palsy on right lateral gaze.

Full blood count revealed raised WBC's (13.7 × 109/L) with a left sided neutrophil shift (80.4%), normal lymphocytic count (13.2%), normal hemoglobin (149 g/L) and normal platelets level (246). No elevated inflammatory markers. Creatinine kinase levels was 742 U/L and normal blood glucose levels (5.4 mmol/L). Rheumatological screening showed normal ANA, rheumatoid factor and complement levels. The subject was not sexually active and no other neurological signs were present. Therefore, infectious work up including Lyme and syphilis serologies and CSF analysis were not performed. A very small (5 × 6 mm) well-defined rounded peripherally hyperdense isolated lesion in the right frontal region, with central hypodensity was observed in Pre and post contrast CT brain.

Differential diagnosis revolved around space occupying pathologies compressing the sixth nerve, pseudotumor cerebri, inflammatory or infectious lesions because of the young age of the subject and sudden presentation of the headache and ipsilateral abducens nerve palsy. Physical examination was not suggestive of raised intracranial pressure, laboratory analysis was normal and brain imaging did not reveal any intracranial pathology apart from the incidental neurocysticercosis. This case was labelled as a case of idiopathic right abducens nerve palsy secondary to an undetermined cause. The subject was started on albendazole 400 mg and IV Dexamethasone 8 mg for presumed inflammatory causes for 5 days. Dexamethasone was tapered gradually and the subject was discharged on oral dexamethasone 4 mg (once/day) for 5 days. During hospital admission the headache had resolved but the palsy persisted on discharge.

This case describes about a young male patient who had no comorbidities, who presented with an acute episode of right sided headache and diplopia. An ipsilateral right abducens nerve palsy was revealed in cranial nerve examination. Laboratory tests and neuroimaging were inconclusive. Neoplastic, inflammatory, ischemic, autoimmune, traumatic and space occupying lesions were excluded as history. Hence this patient was diagnosed with an idiopathic abducens nerve palsy and managed him with steroids to which he responded well.

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The study concluded that the most common isolated ocular nerve palsy is the Abducens nerve palsy. The incidence of cases attributed to idiopathic causes are rising. Cases labelled as idiopathic could be attributed to a defined pathogenesis with detailed history and follow-up, due to recurrence of the illness and have a chance of changing the diagnosis if supported by the detailed history and physical examination findings.[2]

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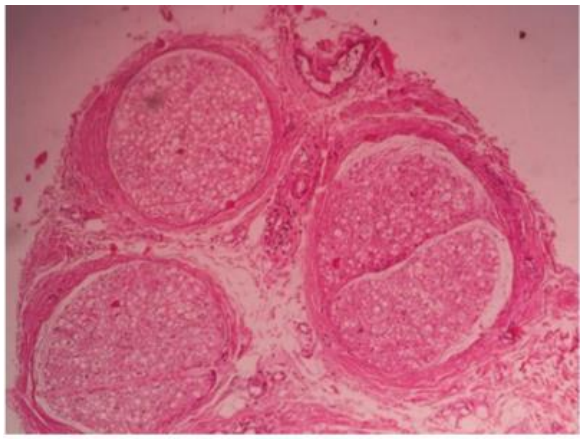
A case study on eosinophilic granulomatosis with polyangiitis (churg–strauss syndrome) mimicking guillain–barre syndrome

Eosinophilic granulomatosis characterized by eosinophil-rich granulomatous inflammation, small to medium-size vessel vasculitis associated with bronchial asthma and eosinophilia. It is a rare form of anti-neutrophil cytoplasm antibody (ANCA)-associated vasculitis[1]. A small-vessel vasculitis associated with antineutrophil cytoplasmic antibodies (ANCAs) is referred to as Churg–Strauss syndrome.

Mononeurities multiplex can mimic Guillain-Barre syndrome which is the most common presentation of vasculitic neuropathy of eosophillic granulomatosis with polyangiits and should always be considered in differential diagnosis

A 49-year-old female presented with a known case of bronchial asthma since 3 years. Twenty days back she developed severe pain in both lower limbs followed by weakness after 5 days of onset of pain which was associated with burning sensation in both feet. Within 48 hours the subject developed weakness in the right lower limb followed by left limb. She was found to have asymmetrical distal weakness in both lower limbs with intact power at hip and knee joint and normal upper limb examination. In lower limbs there was hyporeflexia in both lower limbs and diminished touch and pinprick sensation distally. No bladder, bowel or any cranial nerve involvement was observed. C-reactive protein (CRP), and leukocytosis, cerebrospinal fluid (CSF) study was normal.

Early motor demyelinating affection of bilateral tibial nerves, right peroneal nerve and motor demyelinating plus axonal affection of left peroneal nerve was disclosed by nerve conduction study (NCS). The subject received intravenous immunoglobulins (2 g/kg) along with analgesic keeping the possibility of acute inflammatory demyelinating polyneuropathy. The patient showed improvement and was discharged. On the 15th day, her weakness was worsened, progressing to both the limbs. On Examination, generalized areflexia, with 1/5 power at both ankle joint, 4/5 at knee joint and intact power at hip joint was



A Cross section of a nerve

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revealed. Leukocytosis with eosinophilia raised erythrocyte sedimentation rate {ESR} −96 mm/h, positive rheumatoid factor and C-reactive protein was revealed by laboratory data.

Vasculitic workup was sent which revealed high titers of perinuclear antineutrophil cytoplasmic antibodies. Sensorimotor axonal-demyelinating affection of all nerves was revealed by reporting nerve conduction study (NCS). Pan sinusitis was revealed contrast enhanced computed tomography (CECT) of paranasal sinuses. Bilateral hyper inflated lung fields with fattening of diaphragm with few subcentimetric mediastinal lymph nodes was revealed by computed tomography (CT) thorax. Diagnosis of Churg–Strauss syndrome was made after interpreting clinical and laboratory findings. Nerve biopsy revealed varying size nerve fascicles surrounded by thin perineurium; perivascular mononuclear inflammation and occasional eosinophils within the epineurium; suggestive of vasculitic changes.

Showing extravascular eosinophils

Intravenous methylprednisolone 1 g/day, for 5 days, followed by oral prednisone (1 mg/ kg per day) was given to the patient. For severe pain the subject also received non-steroidal anti-inflammatory drugs (NSAID's), opioids, gabapentin, topical analgesia. In view of long-term immunosuppression, the subject was also given intravenous rituximab (1 gm). Echocardiography (2-D echo) and routine urine study was normal. Follow-up revealed that pain got better and weakness also improved in right hand and foot (from 1/5 to 3/5) with improvement in appetite and body weight.

Peripheral neuropathy is quite common in EGPA and though not life threatening like myocardial or renal involvement, can lead to significant disability. In this case report the subject presented with the history of acute onset weakness of both lower limbs which was preceded by severe pain with burning sensation in glove and stocking pattern which gradually progressed to both upper limbs. The most common presentation of vasculitic neuropathy of eosinophilic granulomatosis with polyangiitis is Mononeuritis multiplex, but they can mimic Guillain– Barre syndrome and should always be considered in the differential diagnosis, since the treatment strategies for these conditions are radically different.[2]

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Eosinophilic granulomatosis with polyangiitis is a small-vessel vasculitis associated with antineutrophil cytoplasmic antibodies with significant paranasal sinuses involvement. Mononeuritis multiplex can mimic Guillain–Barre syndrome which is the most common presentation of vasculitic neuropathy of eosinophilic granulomatosis with polyangiitis and should always be considered in the differential diagnosis, since the treatment strategies for these conditions are radically different. [2]

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