MICRO LABS SYNAPSE

Interesting cases NEUROLOGY

Vol 1|Issue 2|2020

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 of adenomyosis presenting with Acute cerebral infarction.
2. Dentate nucleus as a target for stereotactic thermolesion in central post stroke pain –A case report.
3. Rasmussen's Encephalitis- A case report.

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 of adenomyosis presenting with Acute cerebral infarction.

Introduction:

Adenomyosis occurs in women of childbearing age and mainly manifested as dysmenorrhea, menorrhea and infertility, characterized by presence of ectopic endometrial tissue (endometrial glands and/or stroma) in the myometrium, surrounded by proliferation and hypertrophic smooth muscle. Acute cerebral infarction with adenomyosis is associated with elevated D-Dimer, elevated CA125, anemia and menstruation. Current is a case presentation of adenomysis patient presenting with acute cerebral infarction and fever.

Case presentation:

A 38-year-old female patient presented headache and fever for 4 days and left limb weakness for 1 day. Patient had fever during menstruation 4 days back, temperature was 38 °C went upto 39.1°C , paroxysmal headache, lower abdominal pain, muscle soreness, intermittent cough, sputum was noted. Pulse 80 beats per minute, Blood pressure 120/70 mmHg and oxygen saturation was 97%. Patient did not present dizziness, nausea and vomiting. Just a day before admission, patient had left limb weakness, left mouth angle deviation and vague speech. Patient had history of adenomyosis for 5 years, treated with triprillin acetate, estradiol valerate, dydrogesterone and aspirin. But treatment was stopped 4 months before due to the poor treatment effect. Patient also had undergone bilateral thyroidectomy for thyroid cancer 2 years back. Currently she is taking 2 tablets of Levothyroxine orally every day. Patient did not have history of smoking, hypertension, diabetes, hyperlipidemia, coronary heart disease and family history of cerebrovascular disease. Examination of nervous system revealed sleepiness, vague speech, left central facial-lingual paralysis, other cranial nerve examinations being normal, left limb muscle strength IV, right limb muscle strength V, limb muscle tension being normal, limb tendon reflex symmetry. Bilateral needling sensations were normal. Left Babinski's and Pussep's signs were positive. Meningeal irritation sign was negative. NIHSS score was 3 points. On auscultation, harsh breath sounds and crepitations were heard in bilateral lower lung area. Chest X-ray raised the suspicion of bilateral lower pneumonia. Patient was diagnosed with pulmonary infection after admission. Blood routine indicated the total number of white blood cells (12.06 × 109/L), Neutrophil percentage (84.2 %), HGB (112 g/L). Elevated CRP (149 mg/L, normal: 0–8 mg/L), D-Dimer (27.4 mg/L, normal: 0–1.5 mg/L), Fibrinogen degradation products (FDP) (69.20 mg/L, normal: 0–5 mg/L), fibrinogen (Fbg) (4.79 g/L, normal: 1.7–4 g/L), CA125 (937.7 U/ml, normal: 0–35 U/ml), NSE (39.51 ng/ml,

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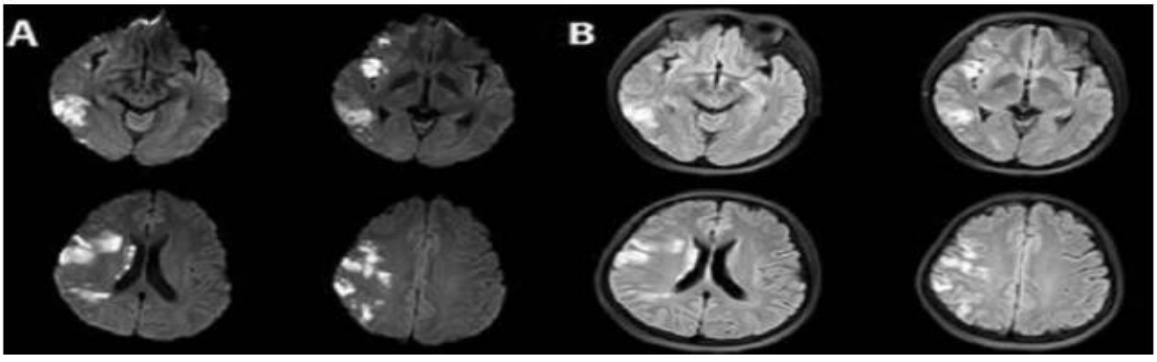
normal: 0–18 ng/ml), CYFRA211 (3.65 ng/ml, normal: 0–3.3 ng/ml). Myocardial enzyme (CK, CK-MB, TNI, TNT) levels were in normal range. ESR, Anti chain "O", Rheumatoid factor, ANCA, ANA, ENA, Immunoglobulin and complement were in normal range. Anticardiolipin antibody, Protein S and protein C were negative. Thyroid profile was normal . Filaria antibody, Lyme-IgG antibody, Cysticercosis-IgG antibody, Leishmania spp-IgG antibody in blood were negative. Result of rose bengal plate test was negative, tuberculosis infection T cell detection was negative. HIV antibody, HCV antibody, HBsAg, HBeAg and Anti-HBc, Treponema pallidum antibody were negative. Cerebrospinal fluid analysis showed normal cerebrospinal fluid pressure, cell number, glucose and chloride of cerebrospinal fluid. Amphiphysin, CV2, PNMA2 in cerebrospinal fluid and blood were negative. Magnetic resonance imaging (MRI) indicated acute cerebral infarction in right basal ganglia and subcortical region of right frontotemporal lobe. Computed tomography angiography (CTA) of head and showed stenosis in the M1 segment of right middle cerebral artery and the distal branches of right middle cerebral artery were significantly less than those on the opposite side, neck showed no carotid stenosis. Transabdominal gynecological ultrasound suggested adenomyosis. Pelvic MRI revealed adenomyosis. Patient was treated with anti-infective therapy (meropenem for injection) for 1 week and using anticoagulant therapy with low molecular weight heparin for 2 weeks. Anticoagulant therapy was discontinued and replaced by antiplatelet therapy with clopidogrel when D-Dimer was normal. CA125 was 202.5u/ml on the 20th day after onset. Transvaginal Gynecological Ultrasound suggested adenomyosis on the third month after onset. Patient was followed up for 4 months, and no recurrence of cerebral infarction was found.



a Diffusion weighted imaging (DWI). b Fluid-attenuated inversion recovery (FLAIR)

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Discussion:

Anemia is a hyperkinetic state which disturbs endothelial adhesion molecule genes that may lead to thrombus formation. Coagulation disorder can also occur during menstruation. Infection can increase the incidence of acute cerebral infarction. Cause of acute cerebral infarction with infection relates to infection load, changes in lipid metabolism, increased plasma fibrinogen, platelet activation or aggregation, platelet lysis, hypercoagulability, a Diffusion weighted imaging (DWI). b Fluid-attenuated inversion recovery (FLAIR) endothelial dysfunction, vascular smooth muscle spasm, unstable atherosclerosis and subsequent plaque rupture. Patient with adenomyosis during non-menstrual period has normal coagulation system. During menstrual period, FDP can be increased, APTT and PT can be prolonged In current case acute cerebral infarction with adenomyosis was accompanied by fever, anemia, menstruation, elevated levels of CA125 and D-Dimer. Studies have shown increased CA125 and D-dimer in patients with adenomyosis during menstruation.

Conclusion:

A case of Adenomysis with acute cerebral infarction was successfully treated. Infection is a potential risk factor for developing acute cerebral infarction with adenomyosis.

Reference:

Zhao Y et al. Acute cerebral infarction with adenomyosis in a patient with fever: a case report. BMC Neurology; 2020 :20:210.

2. Dentate nucleus as a target for stereotactic thermolesion in central post stroke pain –A case report.

Introduction:

Dejerine Roussy syndrome (Central post stroke pain) is a type of neuropathic pain that occurs due to previous stroke in the thalamic region. Central post stroke pain (CPSP) usually suffer from dysesthesia, allodynia and hyperalgesia. It can present with headache, painful spasticity or shoulder pain. It may be due to the involvement of spinothalamic tract. Treatment involves antidepressants, anticonvulsants, opioids or N-methyl D-aspartate Receptor. Other modalities of treatment involves repetitive transcranial magnetic stimulation, motor cortex stimulation, transcutaneous electrical nerve stimulation. Below is a presentation of unusual case of a patient

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suffering from CPSP with a spastic component, pain relief interestingly occurred after the stereotactic thermolesion of the right dentate nucleus (DN).

Case presentation:

60 year-old man with CPSP presented with complaints of intractable spastic pain on the right side of his body after an ischaemic stroke in the thalamic region of the left side. Neurological examination revealed degree 2 of spasticity according to the Modified Ashworth Scale (MAS), pain was evaluated by Visual Analog Scale (VAS) as 9-10. Patient underwent a medial thalamotomy (CM/Pf complex of the left Side; thermolesion 60 °C, 70 °C, 80 °C for 60 s) as drug treatment failed for his intractable pain 1 ½ years after the stroke as shown in figure. Pain disappeared during the procedure,Complete pain relief lasted only 1 month after surgery. After 1 month patient was suffering from the same spastic and burning pain as before the procedure (MAS 2; VAS 9–10) A second ablative procedure in the Dentate nucleus was planned for right side eight months after the thalamotomy. He underwent a classic thermolesion under local anaesthesia using a Leksell stereotactic frame. The right DN was identified directly according to magnetic resonance imaging (MRI) and trajectory planning with 3D FLASH T1W and T2W sequences was performed. The target was localised 14 mm lateral to the midline, 6 mm behind the fastigium and 3 mm ventral to a line connecting the posterior commissure and fastigium. In semi-sitting position patient underwent the procedure with Leksell frame. A small burr hole was made in the right occipital area for the transtentorial approach. The tentorium was perforated by electrocoagulation using a monopolar electrode. Neurological status and vital signs were controlled during the lesioning, procedure lasted one hour. The accuracy of thermolesion was confirmed by a control MRI performed next day A relief from the spasm of the right hand, swallowing improvement and pain relief immediately after the surgery. Follow up after one month of stereotactic treatment, the spastic pain had totally disappeared (VAS 0) and the MAS score was reduced to 1. CPSP was significantly reduced. At the follow-up session 18 months after the surgery, the patient reported a partial recurrence of the pain (VAS 3) that still remained after 3 years.

Discussion:

CPSP condition management requires a multidisciplinary approach and appropriate choice of treatment. In case of r severe CPSP, surgical therapy may be the best option. Dentatotomy ameliorates the spastic pain that improves quality of life of these patients. Hyperexcitability is significantly reduced after dentatotomy of the motor cortex, and its effect on spasticity is as well

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demonstrated by the F-wave reduction. After 2 months of second surgery, the spasticity and pain disappeared and the CPSP was significantly reduced.

Conclusion:

Stereotactic thermolesion of Dentate nucleus is an effective procedure that significantly reduces spastic pain and improves the quality of life in patients with CPSP.

Reference:

Hanuska J et al. Dentate nucleus as a suitable target for stereotactic thermolesion in central poststroke pain: Case report. Clinical Neurology and Neurosurgery; 2020; 195 : 105850.

2. Relapsing Case of Susac's Syndrome treated with Infliximab- A case report.

Susac's syndrome is a potentially debilitating and rare disease that affects small cerebral and retinal arteries and the cochlea, resulting in the triad of encephalopathy, branch retinal artery occlusions, and sensorineural hearing loss. Pathogenesis may be due to cytotoxic CD8+ cell-mediated endotheliopathy with consequent microinfarctions. Immunosuppressive therapy is often required to reduce disease sequelae. Current is a case presentation of effect of infliximab upon acute and chronic disease activity in this case of Susac's syndrome which was refractory to other immunosuppressive and immunomodulatory agents.[1]

Case presentation:

A 25-year-old presented with a four-week history of progressive right-sided hearing loss, wide based gait, and cognitive disturbance characterized by short term memory loss, impaired attention span, and verbal fluency. Patient did not have any personal or family history of any medical problems, and he did not smoke, drink excessive alcohol, or take illicit drugs. On examination, he had dysdiadochokinesis and cerebellar ataxic gait. MRI brain demonstrated multiple hyperintense lesions in the corpus callosum, periventricular white matter, cerebellar hemispheres, and leptomeninges CSF showed pleocytosis with normal cytology and an elevated protein 3.06 g/L, an absence of oligoclonal bands in both CSF and serum was seen. Microbiology of CSF revealed a Gram-negative stain, syphilis serology, and cryptococcal antigen. Triphasic waves consistent with encephalopathy but with no focal or generalised epileptic activity were demonstrable on an electroencephalogram (EEG). Evaluation for autoimmune profile did not reveal any data. Differential diagnosis included acute disseminated encephalomyelitis, aquaporin-4 negative

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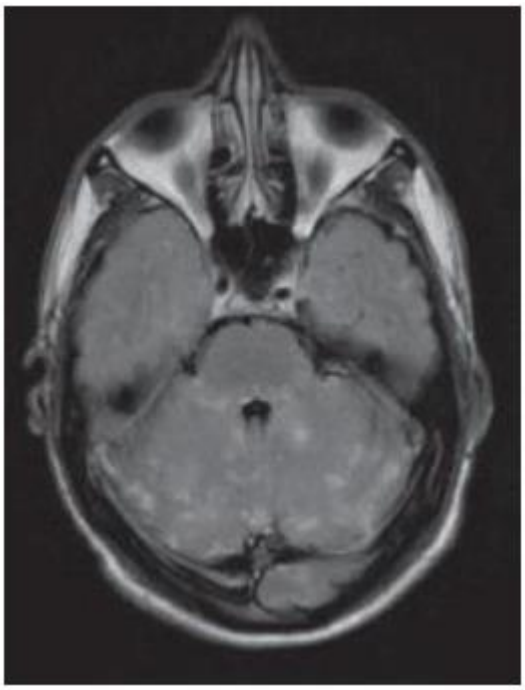
neuromyelitis optica, neurosarcoidosis, and primary angiitis of the CNS. Even after treatment with intravenous methylprednisolone 1 g daily for 3 days followed by a tapering course commenced at 1 mg/kg daily, patient suffered progressive bilateral hearing loss over the following four weeks and impaired vision from the right eye. Audiometry demonstrated low frequency sensorineural hearing loss bilaterally. Neuropsychometric assessment revealed severe global cognitive impairment including difficulties with verbal fluency, as well as deficits in short-term memory and executive function. Biopsy of the cerebellum and meninges was performed due to disease progression and lack of response to the high dose corticosteroid therapy. Histopathology showed mild, nonspecific perivascular inflammation and a diffuse pial infiltrate dominated by CD8 T lymphocytes and macrophages with microinfarctions in the territory of the small pial arteries. No evidence of vasculitis or malignancy were seen. Culture and PCR analysis of the specimen excluded mycobacterial or fungal infection. Patient developed painless scotomata, and a branch retinal artery occlusion (BRAO) of the right temporal retina, as identified on fundal photography and retinal fluorescein angiography. Susac's syndrome was diagnosed based on the triad of encephalopathy, right BRAO, and bilateral sensorineural hearing loss. Methylprednisolone 1 g daily for 3 days was subsequently readministered followed by oral prednisolone 1 mg/kg daily concurrently with intravenous cyclophosphamide 1 g (15 mg/kg) monthly for 6 months and intravenous immunoglobulin (IVIg) 2 g/kg loading and 0.4 g/kg per month for 6 months. Co treatments included cotrimoxazole prophylaxis against Pneumocystis jiroveci, risedronate for osteoprotection, and aspirin to aid vessel patency. Clinical remission achieved by 6 weeks, but no improvement in the sensorineural deafness on the right side. Maintenance of remission was attempted with methotrexate 20mg (0.3 mg/kg) weekly following cyclophosphamide. Despite, another clinical relapse, occurred 3 months after the maintenance therapy with methotrexate was started, characterized by recurrent bilateral hearing loss, bilateral BRAOs, and new T2 hyperintensities shown by MRI of the brain. Despite treatment, the patient relapsed again two months later, suffering a generalised tonic-clonic convulsion, encephalopathy, recurrent bilateral



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hearing loss, fresh BRAOs, and new MRI lesions. MRI brain with MR venogram demonstrated right transverse venous sinus thrombosis. A procoagulant screen was positive for the lupus anticoagulant. Infliximab 5 mg/kg was started using a loading regimen at 0, 2, and 6 weeks for induction and then 8 weekly treatments for maintenance. During course of 4 weeks, both clinical and radiological remission of disease was seen. Prednisone dose was tapered completely from 12.5 mg daily over 4 months, thrombus resolved, and the patient returned to work. Patient was on maintenance infliximab therapy for two years , patient has been free of relapses, the visual acuity is normal, and there are no cognitive deficits on follow-up neuropsychological testing.

Discussion:

Susac's syndrome is rare condition with female preponderance with peak age at onset occurring in the third and fourth decades of life. Disease is autoimmune characterized by cerebral microinfarction, hyalinised vessel walls due to collagen deposition, CD8+ lymphocytic inflammation, and perivascular lymphocytic cuffing in the leptomeninges, with enlarged and reactive endothelial cells leading to near occlusion of the vessel.[1]

Conclusion:

Infliximab may be effective therapy in aggressive treatment of refractory Susac's syndrome to improve prognosis.

Reference:

1)Fernando SL et al. The Successful Use of Infliximab in a Relapsing Case of Susac's Syndrome. Case Reports in Neurological Medicine. 2020; 9317232: 1-6.

3. Rasmussen's Encephalitis- A case report.

Rasmussen's encephalitis is a rare neurological disease of childhood characterized by unilateral hemispheric atrophy, pharmacoresistant focal seizures, and progressive neurological deficits. Etiopathogenesis is not clear. MRI Imaging brain plays a pivotal role in diagnosis and control of disease progression. Cases with atypical MRI features of RE represented by improvement and reoccurrence of signal abnormalities are reported previously. Current is a case presentation of Rasmussen's Encephalitis.

Case presentation:

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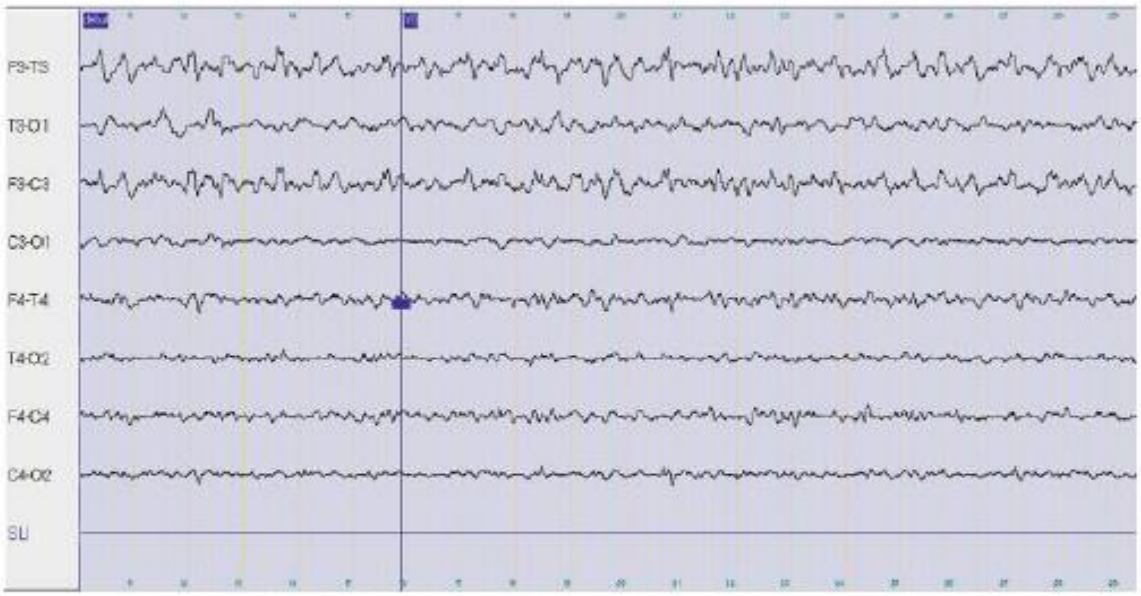
A 12-year-old boy born to nonconsanguineous parents, had a family history of febrile seizures in his sister, epilepsy in 2 cousins, and ulcerative colitis in his mother. He did not have significant antenatal and perinatal history. Psychomotor development was normal. Patient was earlier treated for adrenal insufficiency and dysthyroidism.Patient presented with focal clonic right-sided seizures, preceded by gastroenteritis 1 month ago. Neurological examination showed right hemidystonia, myoclonia, right pyramidal syndrome, and right hemihypoesthesia. Electroencephalogram done inter ictally showed left frontotemporal discharge persisting during sleep. Magnetic resonance imaging (MRI) showed cortical and subcortical hyperintensity on T2-weighted (T2) and fluid-attenuated inversion recovery (FLAIR) images in the left frontoinsular



EEG

region, homolateral lenticular, and caudate nuclei. Spine MRI was normal.Diagnosis of acute disseminated encephalomyelitis (ADEM) was suspected initially. Routine blood and Infectious serologies were negative. Patient received a pulse of steroids (1 g/ day) during 5 days and, then, relay per os at the dose of 1 mg/kg/day for 10 days.With clinical improvement and partial seizure control, Valproic acid and clobazam were prescribed. Follow up MRI performed after 3 weeks was normal.He presented again with intractable focal right seizures, progressive impairment of language abilities, and behavioral disorders with irritability, eating disorders (polyphagia), and worsening school performance with working memory problems.Neurological examination showed

right hemiparesis and dystonia of the right upper limb. With the fluctuating subacute course, seizures, behavioral disturbances, and progressive cognitive impairment, autoimmune encephalitis was suspected.MRI brain showed cortical hyperintensity on T2 and FLAIR images in left frontoinsular, left frontoinsular cortical atrophy with homolateral striatum atrophy, and dilatation of the ipsilateral ventricular system. Diagnosis of RE was done.Numerous regimens of antiepileptic drugs were prescribed (valproic acid 2 g/day, carbamazepine 1400 mg/day, levetiracetam 2500 mg/day, clonazepam 4 mg/day,and piracetam 1600 mg/day).Steroid pulse at adose of 1 g/day for 3 days was administered every month during 12 months. Azathioprine was prescribed at the dose of 100 mg/day. Partial control of seizures was obtained.Patient again presented with several status epilepticus concomitant to infectious episodes. Motor function improved mildly. Brain MRI on follow up showed an increase of the left hemispheric atrophy

Discussion:

RE is a progressive chronic inflammatory disease of the central nervous system. It is rare and affects mostly children. Median age of onset is 6 years. Both sexes are equally affected. characterized by focal intractable seizures, progressive neurological deficit, and cognitive decline, with unihemispheric brain atrophy. Etiologies involves an autoimmune phenomenon involving circulating antibodies against glutamate receptors, and cytotoxic T cells. Many patients show unilateral enlargement of the ventricular system which is accentuated in the insular and periinsular regions.

Conclusion:

Rasmussen's encephalitis is disease of one cerebral hemisphere that is progressive and characterized by frequent focal seizures, hemiparesis, and mental deterioration.

Reference:

Klaa H et al. Rasmussen's Encephalitis: A Report of a Tunisian Pediatric Case and Literature Review. Case Reports in Neurological Medicine. 2020; 6810237: 1-5 pages

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