MICRO LABS SYNAPSE

Interesting cases NEUROLOGY

Vol 1|Issue 3|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. Neurosarcoidosis Presenting as Adipsic Diabetes Insipidus Secondary to a Pituitary Stalk Lesion- A case report.
2. Rasmussen's Encephalitis- A case report.
3. Neurosarcoidosis Presenting as Adipsic Diabetes Insipidus Secondary to a Pituitary Stalk Lesion- 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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Neurosarcoidosis Presenting as Adipsic Diabetes Insipidus Secondary to a Pituitary Stalk Lesion- A case report.

Sarcoidosis is one of the multisystemic inflammatory disease , whose primary feature is the presence of noncaseating granulomas. It usually involves lung involvement in 95% of the cases , however, neurosarcoidosis is present only in 5–15% of the cases, of which in 50% is the initial manifestation. Isolated neurosarcoidosis is very rare, and up to a third of the patients will develop systemic features in the future.Current is a case of adipsic diabetes insipidus secondary to a pituitary stalk lesion, with the final diagnosis of neurosarcoidosis. Patient had neuropsychiatric symptoms and anti-NMDA antibodies detected in cerebrospinal fluid.

Case presentation:

A 20-year-old woman presented with a two-month clinical picture of headache, incoherent language, aggressive behavior, inappropriate conducts of disinhibition, and visual-auditory hallucinations). The patient had gradually developed a diminished state of awareness. Medical history was unremarkable. Patient had two uncomplicated previous pregnancies. No history of animal contact, recent traveling, ingestion of drugs, or exposure to toxics. On examination, patient had tachycardia with a weak pulse, blood pressure normal, dry mucous membranes, and prolonged capillary refill time. Patient was disoriented and presented with retrograde amnesia, impairment in judgment, and loss of abstract thinking. Her chest X-ray and head computed tomography (CT) were normal. Laboratory results revealed severe hypernatremia (Na 180 mEq/L) and acute kidney injury (AKI) (measured creatinine 2 mg/dl–baseline creatinine 0.6 mg/dl). Serum electrolytes, liver tests, blood count, urinalysis, and toxicologic screen were normal. Urinary electrolytes showed a urinary sodium of 20 mEq/L, with a fractional excretion of sodium (FeNa) of 0.5% suggesting hypovolemia. calculated water deficit was 7.5 liters. Calculated electrolyte-free water excretion was of 805 ml within a 24-hour urinary volume of 1,000 ml, a value that was inappropriately elevated. Initiated treatment with enteral administration of free water and hypotonic intravenous solutions for restitution of intravascular volume. Patient presented a partial clinical response, with optimization of volemia and resolution of (AKI). A new electrolyte profile showed persistence of hypernatremia (Na 157 mEq/L) and mild hypokalemia (K 3.2 mEq/L). Patient had a urinary sodium level of 8 mEq/L, with a calculated urine osmolarity of 85 mEq/L, in the context of a 24-hour urinary volume of 4 liters and electrolyte-free water excretion of 3.3 liters. Documented aqueous polyuria and hypernatremia suggested diabetes insipidus. water deprivation test was contraindicated. Brain magnetic resonance imaging (MRI) showed a pituitary stalk lesion

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with chiasmatic and hypothalamic extension. Desmopressin was initiated for treatment with progressive normalization of her sodium level, urinary volume, and volemia status. Hormonal profile was compatible with panhypopituitarism, hence initiated treatment with thyroid and glucocorticoid hormones. Cerebrospinal fluid (CSF) analysis showed an elevated protein level of 75 mg/dl, normal glucose, a cell count of zero, and negative Gram and India ink stains. An electroencephalogram showed diffuse cerebral dysfunction without the epileptiform activity. A stereotactic biopsy of the pituitary mass revealed noncaseating granulomatous inflammation and gliosis. Based on the pathology findings and the 2018 consensus diagnostic criteria a final diagnosis of isolated neurosarcoidosis was made. Patient received treatment with prednisone and showed significant improvement of the neuropsychiatric symptoms. Patient was discharged with continue ambulatory treatment.

Discussion:

Neurosarcoidosis infiltrates the CNS. Most frequent manifestation is cranial neuropathy, aseptic meningitis, cauda equina syndrome, diffuse encephalopathy, focal neurological symptoms, hypothalamic and pituitary dysfunction, intracranial hypertension, peripheral neuropathy, and vascular syndromes. Polyuria-polydipsia syndrome is usually multifactorial and can be secondary to central diabetes insipidus, primary polydipsia, nephrogenic diabetes insipidus secondary to hypercalcemia, or less frequently dysfunction of the thirst control mechanism.

Conclusion:

A case of rare case of Isolated neurosarcoidosis was successfully managed.

Reference:

Solis JG et al. Case Report - Neurosarcoidosis Presentation as Adipsic Diabetes Insipidus Secondary to a Pituitary Stalk Lesion and Association with Anti-NMDA Receptor Antibodies. Case Reports in Neurological Medicine. 2020; 7956350: 1-5 pages.

2. 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

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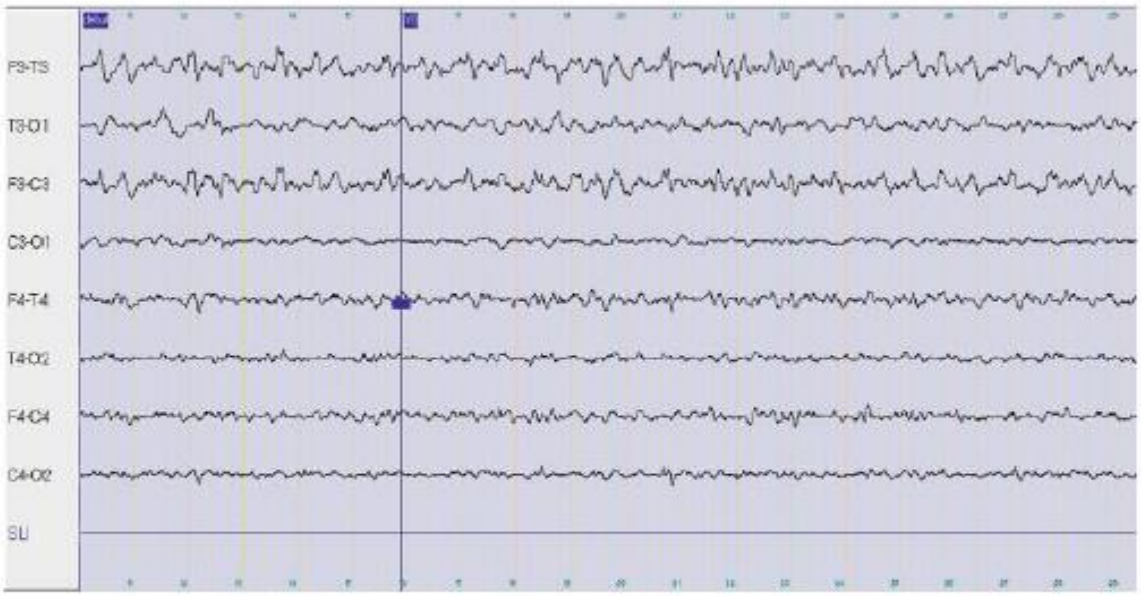
reoccurrence of signal abnormalities are reported previously. Current is a case presentation of Rasmussen's Encephalitis.

Case presentation:

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



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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

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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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3. Neurosarcoidosis Presenting as Adipsic Diabetes Insipidus Secondary to a Pituitary Stalk Lesion- A case report.

Introduction:

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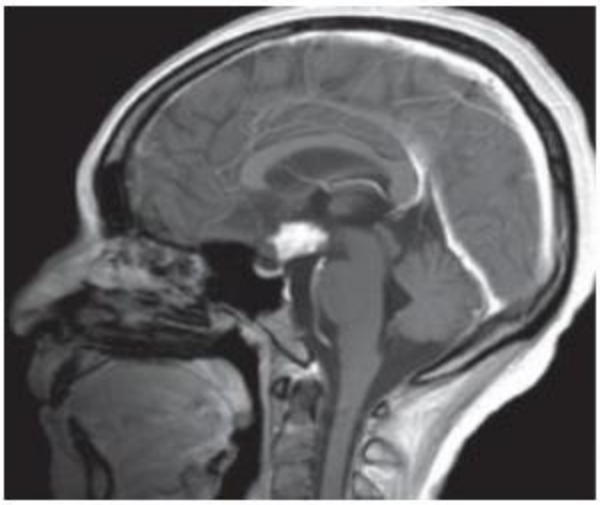

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