phototherapy in the ward. Her BP stabilized. She was given physiologic oral hydrocortisone replacement (6 mg/m^2/day). Six months later, her weight reduced with resumption of linear growth, and improved metabolic control.

CONCLUSION

Exogenous Cushing syndrome resulting from topical medications has been described well especially among young infants. Potent topical steroids particularly for young children should ideally be administered with doctor's prescription.

EP_P007

STEROID-RESPONSIVE ENCEPHALOPATHY ASSOCIATED WITH AUTOIMMUNE THYROIDITIS: THE OUTCOME OF NEUROLOGICAL AND THYROID STATUS

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INTRODUCTION/BACKGROUND

Steroid-Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT), also termed as Hashimoto's Encephalopathy is a neuroendocrine disorder characterized by a triad of subacute onset of encephalopathy, elevated anti-thyroid antibodies, and neurological improvement following steroid therapy. It is a rare but more likely under-diagnosed condition in patients presenting with encephalopathy.

CASE

A 7-year-old Chinese female was referred for headache, seizures, and mood changes for 2 months. She was found to have hyperthyroidism when she presented with frequent hunger and weight loss 4 months ago. She was started on carbimazole 2.5 mg twice a day and she became euthyroid clinically and biochemically. She was otherwise a brilliant child with no other medical illness. She had no family history of thyroid or autoimmune disorders. Examination revealed an irritable child with upper motor neuron signs. Her cerebrospinal fluid analysis for viral PCR and neuronal antibodies were negative. Her cranial MRI and EEG were reported as normal. Her thyroid function was normal (TSH 3.65 uIU/mL, T4 8.5 pmol/L). Her thyroid antibody levels were all significantly elevated (thyroid stimulating immunoglobulins 3.1 IU/L, anti-thyroid peroxidase antibodies 731.8 IU/ml, anti-thyroglobulin antibodies 642.9 IU/ml). A diagnosis of SREAT was made. She received intravenous methylprednisolone 30 mg/kg/day for 5 days followed by a course of prednisolone for a month. She made a complete recovery. She remained clinically and biochemically euthyroid without medication. A year later, her symptoms recurred, and she was treated similarly as her previous presentation. She recovered but had persistent seizures and needed anti-seizure medication. Her seizure frequency was once a month until 4 years later wherein her seizure frequency increased to weekly without other symptoms. She was given a course of oral prednisolone for 3 months. She was seizure-free for 2 months before her seizures resumed monthly. She was also found to have hypothyroidism (TSH 28.69 uIU/ml, T4 6.65 pmol/L) on screening. She was started on L-thyroxine and became euthyroid after 2 months of treatment.

CONCLUSION

This case report illustrates that epilepsy is a clinical sequela of SREAT despite being a steroid-responsive condition. Thyroid status does not determine seizure control; hence it reflects an association rather than causation of the encephalopathy.

EP_P008

TRIPLE-A SYNDROME: A RARE PRESENTATION OF ADRENAL INSUFFICIENCY

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INTRODUCTION/BACKGROUND

Triple-A syndrome or Allgrove syndrome is a rare autosomal recessive congenital disorder. It is characterized by Addisonianism, achalasia and alacrima. It is a progressive disorder that can take years to develop the full-blown clinical picture.

CASE

We report 2 individuals with Triple-A syndrome who initially presented with recurrent hypoglycemic seizures at about 4 years old. They also had faltering growth with short stature. Both had significant hyperpigmentation, without ambiguous genitalia or neurological abnormality. Hormonal assay confirmed glucocorticoid deficiency, with sparing of mineralocorticoid involvement. Both were subsequently started on hydrocortisone replacement.

In the subsequent years, both developed new symptoms of recurrent vomiting and dysphagia, suggesting achalasia. Subsequently, patient 2 had barium swallow study done which confirmed an esophageal dysmotility disorder. Retrospectively, both patients were also found to have absent tear production from a very young age, signifying alacrima.

Patient 1 managed to undergo Whole Exome Sequencing (WES) study and a homozygous variant of uncertain significance (VUS) was identified in the AAAS gene (cDNA: NM_015665.6:c.1087G>A). In view this is the gene of interest that fits into the clinical picture, the mutation was likely pathological.

CONCLUSION

High index of suspicion is required to diagnose this rare entity. Unexplained cases of adrenal insufficiency should be carefully evaluated for signs and symptoms of alacrima and achalasia.

EP_P009

AUTOSOMAL DOMINANT, NON-AUTOIMMUNE, CONGENITAL HYPERTHYROIDISM

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INTRODUCTION/BACKGROUND

Primary non-autoimmune hyperthyroidism is a rare cause of neonatal hyperthyroidism. This results from an activating mutation in the thyrotropin-receptor (TSHR). TSHR is a G-protein coupled receptor and is found primarily in the thyroid gland. It is also present in adipocytes, fibroblasts and bone cells. It can be inherited in an autosomal dominant manner or occur sporadically as a de novo mutation. Affected individuals display a wide phenotype from severe neonatal to mild subclinical hyperthyroidism. The severity of the hyperthyroid symptoms is variable and phenotype differences have been described in subjects harbouring the same mutation.

CASE

A 10-month-old female was born late preterm at 36 weeks via spontaneous vertex delivery with birth weight of 2.5 kg. Antenatally, the mother was asymptomatic for hyperthyroidism. There was no family history of thyroid or autoimmune diseases. The baby was referred at birth with deranged cord TSH of 0.05 miu/L. Repeat thyroid function tests at day 7 of life demonstrated an elevated T4 of 27.6 pmol/L and low TSH of 0.01 miu/L. She had persistently high free T4, hence, was started on carbimazole at 3 months old. She was also asymptomatic for hyperthyroidism. Her growth parameters were appropriate for age. She does not have dysmorphism. She had normal heart rate, blood pressure, and tone. She did not have a goiter.

Her thyrotropin receptor autoantibodies were negative. Her neck ultrasound revealed a normal thyroid gland with normal vascularity, with no focal thyroid lesion seen. Genetic test revealed a heterozygous pathogenic variant in TSHR NM_000369.5:c.1891T>G (NP_000360.2:p.Phe631Val). During the course of follow-up, she was well and remained asymptomatic.

CONCLUSION

Although rare, TSHR gene mutation should be considered in an infant who presents with hyperthyroidism with a negative autoimmune screen and a negative maternal history of thyroid disorders. Early diagnosis is important so as to rapidly initiate anti-thyroid therapy and manage the thyrotoxicosis, to potentially avoid an enlarging goiter, and to prevent neurocognitive delays in young children with nonautoimmune hyperthyroidism.

EP_P010

DIABETIC KETOACIDOSIS WITH MELIOIDOSIS: A CASE REPORT

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INTRODUCTION/BACKGROUND

Paediatric melioidosis is uncommon yet is associated with high morbidity and mortality in severe disease particularly in immunocompromised patients. Reports of melioidosis in paediatric diabetes are scarce. We present two patients with melioidosis who presented with diabetic ketoacidosis (DKA) in our centre.

CASE

Patient A is a 15 year-9-month-old female with underlying type 1 diabetes (T1D). She presented with mild DKA (serum glucose: 25 mmol/L, serum ketone: 3.9 mmol/L venous pH: 7.30, HCO_3- : 13 mmol/L) associated with fever and symptoms of upper respiratory tract infection (URTI) for