

above findings, elevated ferritin and tryglicerides, decreased NK cell activity, splenomegaly and enlarged lymph nodes were demonstrated. Bone marrow biopsy confirmed HLH. Homozygous mutation in the perforin gene was found. The patient was treated with antibiotics and immunochemotherapy. Bone marrow transplant was performed from a HLA identical brother.

#### CONCLUSIONS

Although the initial MRI was consistent with gliomatosis cerebri, presence of diffuse areas of enhancement are rarely seen in this condition. When they are present, they usually represent local malignant transformation. Response to steroids and development of pancytopenia were the key elements leading to the correct diagnosis. No patient with initial diagnosis of gliomatosis cerebri should be abandoned without adequate follow-up in order to exclude other pathologies.

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##### GRISCELLI SYNDROME: A CASE REPORT AND REVIEW OF THE LITERATURE

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#### PURPOSE

We present a rare case of Griscelli syndrome with involvement of the central nervous system and review the existing English-language literature. Typical and atypical neuroimaging findings on a series of CTs and MRIs will be discussed.

#### METHODS

A 3-and-a-half year old previously healthy boy was admitted to our institution after his third episode of fever in three month time. Extensive workup ensued including a bone marrow biopsy which revealed the presence of occasional hemophagocytic histiocytes. The presence of silvery hair was noticed and a presumptive diagnosis of Griscelli syndrome was made. During hospitalization, seizure activity was noticed which led to a series of brain MRIs included FLAIR, DWI, pre and post contrast T1W.

#### RESULTS

Bone marrow biopsy results showing occasional hemophagocytic histiocytes. Genetic test revealed mutations in the RAB27A gene. The first brain MRI obtained revealed numerous lesions at the gray-white matter junction in addition to brain stem involvement. A series of follow up MRI examinations revealed progressive involvement of the cerebral cortex and cerebellum ultimately involving the white matter while sparing the thalami and portions of the basal ganglia.

#### CONCLUSIONS

Griscelli syndrome, a rare autosomal recessive disorder is characterized by silvery hair, pigment abnormalities, severe immunologic dysfunction and central nervous system manifestations. Fulminant involvement of the brain results in a diffuse loss of cortical and deep hemispheric white matter with relative preservation of the deep gray matter, cerebellum and brain stem. While involvement of the basal ganglia has been mentioned in a prior published case report, the manifestations in the case above were far more extensive and cross at different stages with other more common pathologies.

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##### IN UTERO COCAINE INTOXICATION: PECULIAR POST NATAL MRI FINDINGS IN AN INFANT

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#### PURPOSE

Prenatal cocaine exposure causes hypertension, placental abruption, spontaneous abortion, poor pregnancy weight gain, and under-nutrition secondary to suppression of the appetite. The offspring can also show a variety of developmental behavioral, visual, hearing, and language disorders. The aim of our study is to describe Magnetic Resonance Imaging findings in an infant prenatally exposed to cocaine.

#### METHODS

Multiplanar and multiecho post natal MRI imaging of the infant's brain was performed emphasizing the short and long TR signal characteristics and morphological findings. Clinical evaluation of the patient and relevant history from the family was obtained. Hair testing was used to assess maternal consumption of drugs of abuse during pregnancy and consequent chronic fetal exposure. Maternal and neonatal hair testing was performed by a validated assay including keratin matrix digestion, extraction and analysis by gas chromatography-mass spectrometry.

#### RESULTS

A male newborn presented with hypoglycaemia and prolonged colestatic jaundice. At one month—follow-up, growth delay and visual impairment were detected. Sagittal T1 weighted image showed the absence of the posterior “bright spot” in its expected location within the posterior sella turcica cavity associated with pituitary stalk hypoplasia. Findings included optic nerve atrophy bilaterally. Prenatal cocaine intoxication was demonstrated by the presence of cocaine and its metabolite benzoylecgonine both in maternal hair segments corresponding to the three gestation trimesters (mean concentration: cocaine: 7.5 ng/mg hair, benzoylecgonine: 1.2 ng/mg hair) and in neonatal hair (cocaine: 5.7 ng/mg hair and benzoylecgonine 1.1 ng/mg hair).

#### CONCLUSION

Recent reports suggest significant visual and developmental anomalies in infants prenatally exposed to cocaine. In the present study MR images correlated with clinical symptoms as demonstrated by atrophy of the optic nerves and pituitary axis anomalies. The role of MRI in depicting cocaine related Central Nervous System anomalies is discussed.

#### Spinal Interventions

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##### SPINE INTERVENTIONAL PROCEDURES UNDER IMAGING-GUIDANCE: COMBINING-COMPARING FLUOROSCOPIC VIEWS WITH 3D AND CROSS-SECTIONAL CT ANATOMY

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