958 FATAL HUMAN BOCAVIRUS INFECTION IN A BOY WITH IPEX-LIKE SYNDROME AND VACCINE-ACQUIRED ROTAVIRUS ENTERITIS AWAITING STEM CELL TRANSPLANTATION

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We report about a 21-month-old boy presenting with chronic diarrhea and obstructive lung disease since infancy. His older brother, the first male child of non-consanguine Egyptian parents, had died at the age of 20 months suffering from BCCHs and severe CMV infection, suggesting a severe primary immunodeficiency syndrome.

We found immunological dysregulation, endocrine dysfunction and enteropathy compatible with IPEX syndrome, therefore the child was listed for stem cell transplantation (SCT). FOXP3 gene showed no IPEX associated mutations, but sequencing IL2RG we found an mutation in Exon 2 associated with x-linked IPEX-like SCID (c.252C>A, p.N84K).

The pulmonary condition of the boy deteriorated and he was admitted on the ICU, where he was mechanically ventilated ever since. After common respiratory infections were excluded, chronic human bocavirus (hBoV) was identified by multiplex-PCR as the primary causative pulmonary agent causing his respiratory failure. hBoV was found in decreasing quantity in respiratory material, blood and stool specimen.

Interestingly, chronic rotavirus shedding was notified in repetitive stool specimen. As the boy had been vaccinated against rotavirus, vaccine-aquired chronic infection was suspected and confirmed by typing of a vaccine virus specific gene variant.

The boy died before SCT could be performed.

Since its discovery, there is an ongoing discussion if hBoV can cause serious infections or represents only a harmless "bystander". This case report shows that hBoV can result in lethal respiratory infections in immunocompromised children.

959 MYCOPlASMA PNEUMONIAE ASSOCIATED WITH STEVENS JOHNSON SYNDROME IN TWO CHILDREN

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Background Mycoplasm pneumoniae (MP) infections are often asymptomatic but can involve multiple organ systems. Secondary skin reactions are common, although few patients infected develop Stevens Johnson syndrome (SJS).

Results We describe 2 cases of Mycoplasma pneumoniae chest infection associated with Stevens Johnson syndrome. The two patients had prodromal symptoms of an upper respiratory tract infection before the onset of the eruption. The patients had extensive epidermal bullous vesicles, oropharyngeal and genital ulceration, injected conjunctivae and sclera and swollen lips with flaccid bullae. The mycoplasma IgG and IgM titers returned positive, and blood cultures and other titers were negative. They were successfully treated with macrolides and glucocorticoids.

Conclusion Although the clinical course may be severe and prolonged, the prognosis is uniformly good with complete recovery.

960 SECONDARY PSEUdohypoalDosteronism due to pyelonephritis: two cases report

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Secondary pseudohypoaldosteronism occurs due to transient resistance in renal tubules causing renal sodium loss, hyponatremia and hyperkalemia. This may resemble congenital adrenal hyperplasia. This is to be considered specially when a urinary tract infection is present as treatment and prognosis varies.

We report two infants who developed pseudohypoaldosteronism secondary to acute tract urinary infection and urologic malformations.

Patient 1 27 days old infant, weight: 4056 gr. He suffers severe dehydration, hynoponatremia 119 mmol/L, K 7 mmol/L and metabolic acydosis, normal serum creatinine and normal 17 OH progesterone. Pathological urine analysis and high levels of serum rennin and aldosterone. Urine culture: E.Coli. We can also apreciate left Ultra sonography hydronephrosis IV/V and primary meaurerat. He was treated with Ampiciline + gentamicin and ions stabilisation within the following 36 hours after hospital admission.

Patient 2 14 days old infant, 2510 gr. Admitted at emergencies with a severe dehydration. Hyponatremia and hyperpotasemia that needed urgent treatment and severe metabolic acydosis. Electrolytic disorders were amended, then a urine culture was done with positive results in E. Coli test. He was treated with ampicicilne + gentamicin. The renal scan evinced a bilateral ureteral hydronephrosis. The infant showed a recovery thanks to antibiotics treatment.

Conclusions An endocrinological evaluation is necessary to a diagnosis but we musn’t forget the urinary cultures.

Most probable cause is the immature renal responsiveness to aldosterone in three first months of life when exist urinary tract anomalies and/or urinary tract infection.

This secondary pseudohypoaldosteronism is reversible with early treatment.

961 THE DIAGNOSTIC VALUE OF NEUROIMAGING IN RECOgnIZING AND THERAPY MONITORING OF ENCEPHALITIS AND MENINGITIS IN CHILDREN

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