GP65

A CASE REPORT ON NOROVIRUS RELATED URTICARIAL RASH IN PAEDIATRIC PATIENT

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There are several causes of infectious and non-infectious urticaria recognized. Reports described association with streptococcus, mycoplasma pneumoniae, parvovirus B19, enterovirus, hepatitis A or B and plasmodium falciparum and urticaria. Norovirus is a recognised albeit rare cause of urticaria. Leiste A et al. have published two cases identified on Norovirus-related urticaria in adult patients. There are no cases yet described in paediatric population on Norovirus related urticaria. Following is the unique case of norovirus related urticaria in paediatric patient.

A two year old, male patient was admitted to the paediatric ward with a sudden episode of generalised florid rash proceeding with multiple episodes of diarrhoea and vomiting. There was no involvement of mucous membranes and joints. The rash was erythematous, with widespread wheals spreading all over the body associated with intensive pruritus which left excoriation marks. The rash and pruritus was subsided with a course of antihistamines. There was no pyrexia documented during the episode and there was no history of atopies. He attends crèche and there was recent acute gastroenteritis in a crèche attendee. On physical examination, the only positive finding was mild to moderate dehydration with no evidence of shock thus intravenous fluid was commenced. His blood investigations done included full blood count, C-reactive protein, renal profile, blood cultures, extended viral panel on nasal swab were normal though stool culture was positive for norovirus. The rash was subsided completely within few hours. Patient was discharged home the following day.

Discussion Noroviruses are genetically diverse group of single-stranded positive -sense RNA, non-enveloped virus belonging to the family Caliciviridae. It primarily causes gastrointestinal symptoms. Patients usually feel very unwell with nausea, vomiting and diarrhoea. A headache and fever may occur. Norovirus is very contagious. It is mainly transmitted by direct person to person contact or through contaminated food/water. It is found in all age group and more prevalent in the winter months. Complications may include dehydration. The incubation period is up to 3 days. There were two cases described previously in adult patient which had an association with urticaria. Comparing the reports from adult's patients, the evidence strongly suggests association between norovirus and urticaria. There is need for further studies and case reports to justify the association.

GP66

FATAL CASE OF MULTIPLE EMBRYONAL TUMORS IN A CHILD WITH BECKWITH-WIEDEMANN SYNDROME

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Beckwith-Wiedemann syndrome (BWS) is the most common genetic cause of overgrowth in early childhood and also the most common syndrome caused by the loss of imprinting.

Depending on the underlying molecular genetics, it can increase the risk of development of embryonal tumours by up to 20%. Children with BWS benefit from radiographic and biochemical screening. In case of early detection of malignancy they are promptly treated with similar or better survival than general population. Multiple concomitant tumours remain rare.

We report a case of a 4-year-old boy diagnosed with BWS at birth. This was based on his clinical features and subsequently confirmed genetically as paternal uniparental disomy (11p15.5). His first malignancy was a hepatoblastoma at birth, treated by surgical resection and chemotherapy. Four months later, a suspected left sided nephroblastoma was identified and left sided nephrectomy was performed. Histology revealed nephroblastomosis. 2 years later, a nephroblastoma in the upper pole of right kidney was described. It responded well to chemotherapy and was resected using nephron sparing surgery technique. Three months into adjuvant chemotherapy, another tumor developed in the lower lobe of right kidney. This progressed despite an intensified chemotherapeutic treatment and a nephrectomy was carried out rendering the patient anephric. Radiation to the abdomen with a boost to the site of the right kidney was offered to the boy, while he was on dialysis. Within 2 months of completion of radiation, a new right sided mass was described on follow-up abdominal imaging. At that point all further oncologic treatment was deemed futile and the boy passed away 6 weeks later.

A review of our 30 year patient registry revealed 295 patients diagnosed with nephroblastoma between July 1988 and December 2018. There were 7 children with clinically suspected Beckwith-Widemann syndrome, 2 of those confirmed genetically. All but one were successfully treated without relapse, synchronous or metachronous malignancy.

Unlike other tumour predisposition syndromes, the increased risk of malignancy in BWS reduces sharply after 8 years of life. Synchronous or metachronous malignancy in this population remain exceedingly rare, making our patient an unfortunate exception. Patients with BWS rendered anephric following bilateral nephroblastoma resection have to remain in complete remission for 2 years post completion of oncological therapy before being considered for a renal transplant. Should they remain without evidence of disease, they are good candidates for kidney transplant, with similar rates of survival and graft rejection to other transplantees.

GP67

A CASE REPORT OF AN INFANT WITH GIANT CONGENITAL MELANOCYTES NAEVUS WITH NEUROCUTANEOUS MELANOSIS

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Background A term infant with features of Giant Congenital Melanocytic Naevus (GCMN) with Neurocutaneous Melanosis (NCM).

Case report A term, female infant, with an uncomplicated antenatal course, of non-consanguineous parents, was noted at birth to have extensive pigmentation of the body.

Examination demonstrated hyperpigmented, rugged, plaque-like lesions in a bathing suit distribution. Within the

pigmented naevus were irregularly shape macule, papules, and plaques of various colours.

Multiple pigmented satellite lesions of size 4–5 cm were present over the body, head, face and extremities. Tufts of coarse hair were present over the satellite lesions, with finer hair covering the abdominal areas of pigmentation. Three nodular lesions were present in the perianal region. Areas of excoriation were noted on the flank areas. No other physical abnormalities were present. Neurological examination was unremarkable.

MRI examination revealed extensive intracranial melanocytic infiltrate, confirming a diagnosis of CGMN with NCM.

Discussion CGMN is an extremely rare condition with incidence estimated at 1/500000 births. Lesions are caused by genetic mutations which lead to defective proliferation, differentiation and migration of melanoblasts. Risk of transformation of GCMN to malignant melanoma varies between 0 and 3.8%, with 80% of this number symptomatic by the age of seven.

CNN originates between the 5th–24th week of gestation and arises from gain of function mutations in either BRAF or NRAS. The protooncogenes c-met and c-kit have also been demonstrated to play a role in the formation of CMN. CMN are predominantly caused by sporadic de novo mutations.

Neurocutaneous melanosis is a rare complication of CMN with just over 100 cases reported. Most patients with NCM are asymptomatic at birth with sequelae appearing later in development.

Treatment of GCMN is both symptomatic and palliative, with surgical techniques including serial resection, excision and grafting and the use of tissue expanders. Non-excisional techniques include dermabrasion, laser ablation and curettage.

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GP68

MORE THAN A FEVER: A CASE SERIES OF ATYPICAL KAWASAKI DISEASE

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Introduction Kawasaki Disease is a vasculitis which tends to occur in children between the ages of 1 and 8 years. It is characterized by prolonged fever ($>38.5^{\circ}\text{C}$, >5 days), exanthem, non-exudative conjunctivitis, inflammation of mucous membranes and cervical lymphadenopathy. Coronary artery aneurysms may develop and rupture or cause myocardial infarction.

Diagnosis is by clinical criteria and treatment includes aspirin and IV immunoglobulin. Diagnostic criteria for Kawasaki disease are fever and at least four of five additional clinical signs.

Atypical (Incomplete) Kawasaki disease should be considered in all children with unexplained fever for more than 5 days, associated with 2 or 3 of the main clinical findings of

Kawasaki disease. Diagnosis of Atypical Kawasaki disease is based on echocardiographic findings indicating the involvement of the coronary arteries.

Case Series From February 2016 to October 2018, four patients presented to Sligo University Hospital with Atypical Kawasaki's Disease. The patients varied in age from seven months to four years old.

The classical signs of Kawasaki disease include persistent high grade pyrexia bilateral non-exudative conjunctivitis, bright red ('strawberry') tongue, cervical lymphadenopathy and skin desquamation. Less specific signs include rash and irritability.

All four patients developed persistent high-grade pyrexia ($>38.5^{\circ}\text{C}$, >5 days) and lymphadenopathy. None had all the typical signs of Kawasaki disease. Three were observed to have a widespread maculopapular rash. Two had bilateral exudative conjunctivitis, or irritability. One had a 'strawberry tongue' appearance. None had skin desquamation.

Only 50% of patients had Kawasaki disease in their initial differential diagnosis. Other differentials included upper respiratory tract infection, tonsillitis, scarlet fever, transient synovitis, cervical lymphadenitis and retropharyngeal abscess.

Management All four patients were treated with IVIG and aspirin. The mean duration of pyrexia prior to commencement of treatment was 7.5 days. The mean duration from hospital presentation to commencement of treatment was 3.5 days.

Outcome 50% of patients made a complete recovery. Despite treatment, 25% developed a mild fusiform dilatation of the left coronary artery. A further 25% developed a saccular aneurysm of the left circumflex coronary artery.

Discussion It is imperative to include Kawasaki and Atypical (Incomplete) Kawasaki disease in the differential diagnosis of a child with persistent high-grade pyrexia. Delays in treatment may lead to a greater risk of coronary aneurysm and mortality.

GP69

CEREBRAL VASCULAR ACCIDENT ASSOCIATION POST PRIMARY VARICELLA INFECTION

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Summary An 18 month old boy presented with right monoplegia and intermittent ataxia. This occurred five months post primary varicella infection. His CT brain revealed low dense lesion in the basal ganglia, a region associated with vasculopathy post varicella infection. The child showed motor recovery with physiotherapy, antiviral, steroid and aspirin. He developed chorea, a manifestation of basal ganglion dysfunction.

Case description An 18 month male presented to the emergency department, with inability to use his right hand since the morning and they noticed that he had difficulty crawling without history of trauma or fever. He had primary varicella infection five months earlier, and parents noticed intermittent unsteady gait for the preceding couple of weeks. He had measles mumps and rubella vaccination four months prior to his presentation and had hand foot and mouth disease a month later. He had a simple febrile convulsion four weeks beforehand and was treated as an outpatient in accordance with