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LETTERS TO THE EDITOR

Dear Editor,

THROMBOCYTOPAENIA IN SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS: NOT ALWAYS MACROPHAGE ACTIVATION SYNDROME!

Pseudothrombocytopaenia is one of the silently puzzling nuisance to the treating physician.[1] EDTA (ethylene diaminetetraacetic acid) is a favoured anticoagulant for laboratory use as it does not distort the blood cells, but it is also known to be notorious to cause clumping of platelets.

A 5-year-old boy presented with high-grade fever for 1 month and arthralgia of both knees and ankles for 5 days duration. He also had evanescent rash over trunk and neck that used to appear at the spike of fever and disappear once the fever subsides. Outside investigation records showed polymorphonuclear leukocytosis (21.5×10^9/L) and thrombocytosis (750×10^9/L). He also had elevated erythrocyte sedimentation rate and C-reactive protein levels. Biochemical profile, serum urea, creatinine, bilirubin, alanine transaminase, aspartate transaminase, albumin levels were normal. Chest radiograph and ultrasound abdomen were normal. Microbiology work-up including blood cultures, urine cultures, hepatitis B surface antigen, brucella serology were negative. Viral serologies including cytomegalovirus, Epstein–Barr virus, human immunodeficiency virus, Hepatitis-C virus were negative. Work-up for tuberculosis and lupus was non-contributory. Bone marrow examination showed reactive changes. Serum ferritin was elevated (1642 ng/mL). In view of quotidian fever and arthritis along with evanescent rash diagnosis of systemic juvenile idiopathic arthritis was proffered after excluding other causes of fever. He was started on oral naproxen 15 mg/kg/day. However, he continued to mount high-grade fever spikes and he was also noted have thromocytopaenia (69×10^9/L). On the background of systemic juvenile idiopathic arthritis, persistence of fever, anaemia, thrombocytopaenia, high ferritin and triglyceride values, macrophage activation syndrome was suspected and child was started on prednisolone 2 mg/kg/day.

On prednisolone, fever and arthralgia showed prompt response, but thrombocytopaenia persisted. A repeat serum ferritin level value was lower (1095 ng/mL), and serum triglycerides, fibrinogen levels were normal. A repeat bone marrow examination was performed which showed non-specific changes and mild increase in histiocytes. Despite clinical improvement, thrombocytopaenia persisted which was not explained with any obvious cause. A peripheral smear showed marked clumping of platelets. Blood counts done with citrated and heparinized samples showed normal platelet counts (4.29×10^9/L, 4.05×10^9/L, respectively).

EDTA occasionally causes clumping of platelets possibly because of alteration of surface glycoproteins. EDTA induced thrombocytopaenia must be considered in patients with systemic juvenile idiopathic arthritis who has persistent thrombocytopaenia despite having improving trend. Clinicians must be aware of this benign entity, as it might avoid multiple other investigations done to rule out causes for thrombocytopaenia. A peripheral smear examination is a valuable tool to identify the presence of platelet clumps due to EDTA.[2] Presence of normal platelet counts in the citrated and heparinized samples will confirm the fact that thrombocytopaenia is EDTA induced and not the real one.[3,4]

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Dear Editor,

PRIMARY EBSTEIN-BARR VIRUS INFECTION PRESENTING WITH PROMINENT LIP AND TONGUE SWELLING

A 7-year-old boy presented to the hospital with a 5-day history of fevers associated with painful lip and tongue swelling. The child was vaccinated and previously well.

The illness initially started with mild bilateral conjunctivitis (spontaneously resolved) and rhinorrhoea 5 days before the onset of the fevers and lip swelling. No other skin changes or rashes were noted by the family. As the swelling of the lips and tongue became more significant, the child began to have increasing difficulty tolerating oral fluids prompting presentation to the emergency department.

On presentation, the child was alert with normal cardiorespiratory vitals, although febrile to 38°C.

His lips were painfully swollen with associated blistering and shallow ulceration (Fig. 1). His tongue was also swollen with

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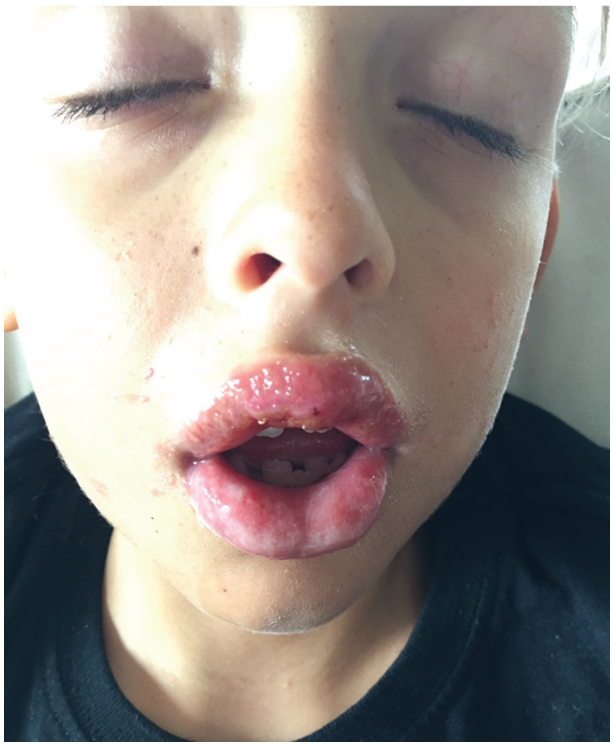

Fig. 1 Lip swelling on the day of presentation to hospital.

small ulcers along the periphery. There were no other oral lesions observed. Bilateral mildly enlarged exudative tonsils were noted. He had mild submandibular lymphadenopathy (<1 cm).

No other skin or mucous membrane changes were appreciated on examination. Cardiorespiratory and abdominal exam was unremarkable.

Investigations included – full blood count, electrolytes and liver enzymes, C-reactive protein, viral serology (Cytomegalovirus, Ebstein-Barr virus (EBV), mycoplasma), lactate dehydrogenase, urine microscopy culture sensitivities, blood culture and oral swabs for Herpes simplex virus and enterovirus. The full blood count was unremarkable, C-reactive protein was <2, while there was a very mild transaminitis noted on liver function tests. Swabs for HSV and enterovirus were negative. Urine MCS was normal and the blood culture remained negative. EBV serology was positive for IgG and IgM suggestive of acute infection.

The patient was managed with supportive measures receiving analgaesia and intravenous fluids. He was also given a total of three doses of oral dexamethasone with a resultant clinical improvement in the mucosal swelling 4 days after presentation to hospital.

Manifestations of EBV infection in children can be heterogeneous.[1] Mucocutaneous presentations are thought to occur in 3–15% of patients with primary infection, with this most commonly manifesting as a morbilliform rash or as a hypersensitivity reaction to antibiotics such as penicillins.[1,2] Other mucocutaneous manifestations may include transient urticaria, erythema nodosum or Gianotti-Crosti syndrome/papular acrodermatitis of childhood.[1,3] Mucous membrane involvement has been known to occur in the form of non-sexually related acute genital ulcers, generally occurring in adolescent females.[4]

No cases of prominent lip or tongue swelling could be found in the literature as a presentation of primary EBV infection. We therefore suggest this case as a unique presentation of EBV infection in childhood currently not represented in the medical literature.

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Dear Editor,

SURVEY ON CLINICAL PRACTICE OF PARENTERAL NUTRITION IN NEONATES IN AUSTRALASIA

Following the formation of the Neonatal Parental Nutrition Consensus Group in November 2010 and publication of consensus guidelines, many neonatal intensive care units (NICUs) across Australia have begun to implement the use of standardised parenteral nutrition (PN) formulations.[1] These formulations have been shown to provide improved nutritional support and promote optimal weight gain in extremely low birthweight infants.[2] A recent systematic review of practice surveys on PN for preterm infants demonstrated significant differences in PN delivery within individual surveys and between surveys, highlighting that there are multiple infrastructural or logistic challenges that need to be addressed to ensure consistent delivery of optimal nutrition across networks of NICUs.[3]

We conducted a survey (SurveyMonkey Inc., San Mateo, CA, USA) among all the representative units in the consensus group to review current practice, with respect to the use of PN, in each individual unit. The main objective was to use the results of the survey to assist in developing an acceptable and practical consensus guideline for the use of PN. The survey was conducted over a

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