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Editorial

Neutrophilic dermatoses: from bench to bedside

Sweet's syndrome and pyoderma gangrenosum are both considered to be 'classical' neutrophilic dermatoses that are uncommon but important as both conditions are associated with underlying disease in approximately 50% of cases, such as malignancies, infections and rheumatological diseases. Sweet's syndrome was first described in 1964 by Dr. Robert Douglas Sweet as an 'acute febrile neutrophilic dermatosis' which was 'a distinctive and fairly severe illness', characterised by fever, neutrophilia, acute onset of painful, erythematous lesions and histological findings of a dense neutrophilic infiltrate without evidence of primary vasculitis. Prompt improvement of both systemic symptoms and skin lesions after the initiation of treatment with systemic corticosteroids is the characteristic.

The pathogenesis of Sweet's syndrome has not been fully elucidated. The theory of cytokine dysregulation leading to activation of neutrophils and release of toxic metabolites has been proposed.[1] In this issue, Drs Toholka and Su, paediatric dermatologists, share with us a clear and concise account on some of the basic concepts of autoinflammatory syndromes presented with neutrophilic dermatoses in the paediatric setting. To most of us, autoinflammatory syndrome is a relatively new area. This article illustrates that autoinflammatory syndrome, classified as interleukin-1β activation disorder can present with a wide spectrum of cutaneous manifestations such as pyoderma gangrenosum and acne in PAPA syndrome.

Sweet's syndrome is rare in childhood and has a wide range of extracutaneous manifestations. In a systematic reveiw of paediatric Sweet's syndrome, associated diseases were reported in 58% of the cases and 33% were categorised as "parainflammatory" Sweet's syndrome including cases of associated chronic recurrent multifocal osteomyelitis, vasculitis with aortitis, recurrent infections due to immunodeficiency, arthritis, and systemic lupus erythematosus.[2] Twenty five percent of the patients were categorised as paraneoplastic, comprising both malignant and premalignant diseases like leukaemia, aplastic anaemia and Fanconi anaemia. Su et al proposed that Takayasu's arteritis should be considered one of the disease associations of Sweet's syndrome when complicated by postinflammatory elastolysis and early referral for cardiovascular screening should be considered in this group of patients.[3] Hence, clinicians should be aware of the possible underlying inflammatory diseases, such as arthritis, vasculitis and inflammatory bowel diseases, especially in children.

Sweet's syndrome has recently been reported to be associated with a new immunodeficiency syndrome mainly found in Southeast Asia. Among the reactive skin conditions, Sweet's syndrome was the most commonly observed entity in patients with anti-IFN-γ autoantibodies and was found to be statistically significantly associated with the immunodeficiency syndrome.[4] Sweet's syndrome is usually preceded by or coexists with opportunistic infections especially due to atypical mycobacteria. The initial manifestation of an undiagnosed case, or an undetected disseminated infection in a susceptible case, may be a reactive skin change encountered in daily clinical practice. It is important for

clinicians to recognise the association with this emerging immunodeficiency syndrome.

In our local setting, patients often present to hospitals with alarming skin lesions and systemic upset under the care of surgeons, physicians or paediatricians. A multidisciplinary approach in managing these cases is desirable as early recognition of distinct inflammatory skin lesions, exclusion of active infections and investigation of the possible underlying medical diseases are of utmost importance. This makes the dermatologist's role in the management of these patients especially relevant. I have seen cases of pyoderma gangrenosum with pathergy being treated as post-operative suppurative wound infection with repeated debridement, leading to extensive tissue loss and severe disfigurement. In summary, Sweet's syndrome and pyoderma gangrenosum should be considered in the differential diagnosis of prolonged fever with cutaneous involvement and rapidly enlarging suppurative and ulcerative lesions, respectively. As most cases of neutrophilic dermatoses are associated with underlying serious diseases particularly malignant/premalignant diseases, immunodeficiency and autoimmune diseases, careful screening and monitoring of these patients are essential.

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