Pediatric Rheumatology

Poster presentation

Chronic granulomatous disease in an infant with sweet syndrome

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Sweet syndrome is a rare and recurrent febrile neutrophilic dermatosis that is characterized by pyrexia, leukocytosis, painful erythamatous plagues of skin and neutrophile infiltration of the dermis.

An 1.5 – month old boy presented with the complaints of fever and rash. He was pale and had widespread lymphadenopathy. He had oral candidiasis and eruption characterized by macules and plaques on an erythematous basis. He was anemic and trombocytopenic. His ESR and CRP levels were high. Antibiotherapy was started with the possible daignosis of sepsis. But there was no growth in his cultures. Viral serology was also negative. After a course of antibiotics he was still febrile and his skin lesions did not diminish. Skin biopsy was performed and the histopathological examination confirmed neutrophilic infiltration of the dermis. According to the diagnostic criteria developed by von den Diesch, the diagnosis of Sweet syndrome was made. He was evaluated for a possible accompanying immunodeficiency and a diagnosis of chronic granulomatous disease was confirmed with NBT test. He was evaluated for the presence of malignancies and collagen vascular diaseases, but none was present. His clinical and laboratory findings improved after systemic steroid treatment.

Here, a rare syndrome accompanied by a concurrent rare immunodeficiency is presented. In the literature there was 2 cases reported with these associations. But still any relationship between them remains to be described.