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BASCULE syndrome after COVID -19 vaccination

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Introduction & Objectives: Bier anemic spots, cyanosis and urticaria-like eruption (BASCULE) syndrome is an underreported paediatric vascular disorder from the group of acrosyndromes. Most cases are adolescents between 12 and 19 years of age or newborns in the first few months of life. The skin lesions usually appear in the lower limbs after prolonged standing and resolve in a few minutes by lying down or walking. The course is unpredictable and the symptoms usually affect the patients 'quality of life.

Materials & Methods: A 14 year-old woman presented with a painful and pruritic erythematous and cyanotic rash, mainly located on her lower limbs. The lesions had appeared 8 months earlier, after receiving her second dose of Pfizer COVID-19 vaccine a few weeks ago. Past medical history included hereditary coagulation factor VII deficiency (normal hemostasis range). The lesions typically appeared on the lower extremities acutely after prolonged standing, resolving rapidly by lying down or walking. Upper extremities and hands were affected occasionally. Physical examination, after a few minutes in a standing position, showed multiple hypocromic macules and urticarial papules with a surrounding erythrocyanotic background on the bilateral legs and thighs.

Results: Laboratory tests, including a complete blood count, basic metabolic, hepatic and renal panel, C-reactive protein, thyroid studies, antinuclear antibody and IgA antitransglutaminase antibody, were unremarkable. The patient was referred to paediatric cardiology for an active standing test in order to exclude postural orthostatic tachycardia syndrome and other forms of orthostatic intolerance. Heart rate, blood pressure, electrocardiogram and echocardiogram were normal. Bilastine 20 mg twice daily was started and she achieved nearly complete resolution of pruritus while taking the drug, but recurred when the dose was halved. Additional preventive measures were recommended, such as physical exercise, healthy diet and elevating the legs when possible.

Conclusion: The pathogeneses of BASCULE syndrome remains unknown. An exaggerated response to decreased arterial oxygen saturation due to venous pooling induced by orthostatism appears to be the trigger of erythrocyanosis and Bier anemic spots, while urticaria-like eruption may result from mast-cell degranulation in response to hypoxia. SARS-COV2 infection has been associated with a variety of skin lesions, including pernio-like changes ("COVID-toes") and peripheral acrocyanosis. The pathophysiological mechanisms are not clearly elucidated, but it is well known that COVID-19 virus and COVID-19 vaccines promote a proinflammatory state that facilitates microvascular thrombosis and intussusceptive angiogenesis. Both may trigger Toll-like receptors 7/8 inducing type 1 interferons production and others proinflammatory cytokines. We present a case of acute-onset BASCULE syndrome following prior COVID -19 vaccination. Berrebi D et al published a case of acute-onset BASCULE syndrome following prior asymptomatic SARS-COV-2 infection. Although they assumed that this finding may very well be coincidental, this case uncovers a plausible biological mechanism by which post-COVID endothelial dysfunction and exaggerated arteriolar constriction trigger or exacerbate BASCULE syndrome in individuals with pre-existing vascular hypersensitivity. The same explanation may be attributed to our case.