LETTER TO EDITOR (VIEWERS' CHOICE)

DIGITAL GANGRENE COULD BE CUTANEOUS POLYARTERITIS NODOSA

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A four years old female child was referred to our hospital with continuous fever and rash over extremities since 15 days. She also had pain in limbs and bilateral pedal swelling since 15 days. She developed multiple painful nodular swelling over both upper and lower limbs on day 5 of fever. Her left ring finger and right 4th toe became black on day 10 of fever. There were no respiratory, cardiac, neurological, renal or gastrointestinal symptoms. The diagnosis of referring pediatrician was septicemia. After admission, she clinically deteriorated. She had fever almost daily and pain in limbs was increasing. She developed palmar erythema and erythematous papules on the dorsum of both feet. Her vitals including blood pressure were normal. Skin examination revealed multiple patchy, maculopapular rash, livido reticularis, subcutaneous tender nodules and evolving gangrene over left ring finger and right 4th toe. Systemic examination was unremarkable. Initial tests revealed hemoglobin of 8.9 g/dl, total leucocyte count of 40,600 cells/cumm with 74% polymorphs, platelet counts of 6,60,000 cells/cumm and erythrocyte sedimentation rate of 24 mm at end of 1 hour. Serum creatine phosphokinase was 22 IU/L, lactate dehydrogenase was 127 IU/L. C-reactive protein and mantoux tests were negative. Urine, serum electrolytes, liver and renal functions tests were normal. Antinuclear antibody (ANA), anticardiolipin antibody (IgG, IgM), ANCA, antistreptolysin O titre, HIV ELISA, Hepatitis B surface antigen and VDRL titres were negative. Blood culture grew coagulase negative staphylococci. Ultrasonography of abdomen and echocardiography were normal. Doppler study of both the lower limbs was normal.

Based on acute onset of gangrene in periphery, livido reticularis, tender subcutaneous nodules, leukocytosis, thrombocytosis, high ESR, negative autoantibodies and raised C-reactive protein, a diagnosis of cutaneous polyarteritis nodosa (c-PAN) was made. Skin histology from lesions showed leukocytoclastic vasculitis with fibrinoid necrosis in the dermal vessels consistent with clinical diagnosis of polyarteritis nodosa (PAN). She was treated with high-dose corticosteroids and cyclophosphamide. Gangrene led to autoamputation of left ring finger whereas toe gangrene which was mild got healed in next few days. She had five relapses in the initial 2 years that were managed by cyclophosphamide and prednisolone. Presently she is on regular follow up and on low dose azathioprine.

Polyarteritis Nodosa (PAN) is a rare autoimmune disease of children. Kussmaul and Maier first described it in 1866. (1) It is characterized by necrotizing vasculitis of small and medium-sized arteries of any organ of the body resulting in ischemia and infarction of affected organ. (2) It can present either in form of cutaneous lesions (c-PAN) or systemic involvement (classic PAN). Classic PAN always presents with involvement of visceral organ (renal, gastro-intestinal system, central nervous system, cardiovascular system)

with poor prognosis whereas c-PAN has been defined as a chronic, benign, relapsing disorder characterized by the involvement of skin, muscles and joint. Diagnosis requires the presence of cutaneous lesions typical of c-PAN (nodules, livedo, ulceration, gangrene, purpura) and on skin biopsy demonstration of a leucocytoclastic vasculitis in the arteries of the deep dermis or hypodermis, with or without associated fibrinoid necrosis; and the absence of visceral involvement at the time of diagnosis. (3) The criteria for diagnosis of c-PAN has now been validated, it requires a systemic inflammatory disease with evidence of necrotising vasculitis OR angiographic abnormalities of medium-/small-sized arteries (mandatory criterion) plus one of five criteria: (1) skin involvement; (2) myalgia/ muscle tenderness; (3) hypertension; (4) peripheral neuropathy; (5) renal involvement .

Remission can be achieved with corticosteroids. The exact duration of treatment is uncertain. (4,5) Immunosuppressive agents such as cyclophosphamide, azathioprine or methotrexate can be used in cases unresponsive to steroid therapy. All children of c-PAN should be followed-up closely for evolution of systemic symptoms. c-PAN has the potential to evolve into systemic PAN although there are only few reported cases. (6-7) Rarity of vasculitic syndrome allows us to disregard polyarterits nodosa as a differential diagnosis but little awareness can help us in diagnosing such a rare disease.

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DOI: 10.7199/ped.oncall.2014.22

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