Sudden onset of cyanosis due to systemic onset juvenil idiopathic arthritis

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A 16-year-old girl, with systemic onset juvenil idiopathic arthritis (SoJIA) of 1 year duration, was referred to the Underwater and Hyperbaric Medicine Department for the evaluation of persistent heavy cyanosis involving distal parts of the extremities (Figure 1, Panel A,B,C). The onset of cyanosis was noted following an acute arthritis attack with severe pain of the joints and a significant increase in body temperature which ultimately exacerbated and required admission to the intensive care unit for one week. Her initial physical examination records revealed a fever of 39 degrees, swelling of the joints, hepatomegaly and axillary, inguinal and cervical multiple lymphadenopathy. Additionally, her laboratory and radiological reports revealed increased acute phase reactants and pleural effusion on the pulmonary x ray. Following SoJIA diagnosis the patient was prescribed several drugs including a disease modifying anti-rheumatic agent (Methotrexate disodium PO 15mg/week) and Methylprednisolone 16mg PO BID combined with lansoprazole 30mg daily. The patient unfortunately did not regularly follow her medical appointments and frequently neglected her medication. After the onset of cyanosis she additionally received an anticoagulant (Enoxaparin 40mg/0.4 ml BID) a vasodilator (Pentoxifylline 600mg infusion within 500ml i.v. fluid a day) and a prostacyclin analog (Ilomedin 20 Mcg/ml 2cc/h, total duration 8h). During her one-month stay in our hospital, she also received a total of 20 sessions of hyperbaric oxygen (HBO) therapy, at a pressure of 2.4 ATA, daily and 2 hours each . Yet cyanosis progressed, particularly in the upper extremities, to a dry gangrene and became demarcated (Figure 1, Panel D,E). HBO treatment was terminated and the patient was referred for amputation of the involved extremities.

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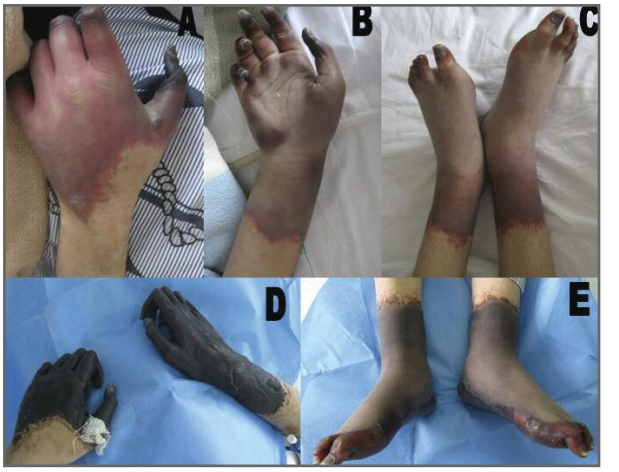

FIGURE 1.

The course of JIA is unpredictable, its consequences may be detrimental and exacerbations are not uncommon[1]. Many patients sustain a poor quality of life due to functional limitation caused from joint deformity and destruction, growth retardation, osteoporosis, etc[1]. SoJIA is the most severe and fatal form of JIA[2] and its pathogenesis still has not been fully elucidated[3]. Gangrene involving the four extremities, as described herein, has not been frequently reported so far. The diagnosis of SoJIA in our patient was established following detailed differential diagnosis excluding several other potential conditions such as systemic lupus erithematosus, dermatomyositis, systemic sclerosis and systemic vasculitis. The occurrence of cyanosis was attributed to vasculitis in SoJIA context after differential diagnosis comprising infective endocarditis and thrombotic events including atrial fibrillation. This illustrative case report clearly demonstrates the severity of SoJCI. Awareness and recognition of the disease should be improved through comprehensive education and the need for regular and close follow up visits to prevent or at least to take control of such disastrous complications should be vigorously explained to patients.

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