CASE REPORT

Severe digital necrosis in a 4-year-old boy: primary Raynaud's or jellyfish sting

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SUMMARY

Raynaud's phenomena is a common disorder which may be primary or secondary to some connective tissue disorders such as systemic sclerosis and systemic lupus erythematosus. Jellyfish sting is a rare but life-threatening cause of Raynaud's phenomena. Digital gangrene is reported in 3% of children with secondary Raynaud's phenomena but does not occur in children with primary Raynaud's phenomena. We report a case of a 4-year-old boy who initially presented with episodes of pain and bluish to blackish discoloration and necrosis affecting the fingers on both hands after a jellyfish sting without any sign of connective tissue disorder.

BACKGROUND

Raynaud's phenomenon (RP) is a clinical manifestation which may be primary or secondary to underlying disorders such as connective tissue diseases. The rare causes of secondary RP are infections, some drugs and jellyfish stings.^{1–3} Digital gangrene is encountered in 3% of children with secondary RP and was not reported in children with primary RP.⁴ We report a case of a 4-year-old boy who initially presented with episodes of pain and bluish to blackish discoloration and necrosis affecting the fingers on both hands after a jellyfish sting.

CASE PRESENTATION

The boy was referred to paediatric cardiology department for the evaluation of peripheral cyanosis located in his hands and fingers. His family did not declare any trauma or insect bite. He had contact with jellyfish 10 days before the first admission to the outpatient clinic. He had fever and rashes on the hands, feet and mouth 3 days after contact with jellyfish. This was diagnosed as hand-foot-mouth disease. After recovery from this disease, bilateral

Figure 1 Bluish to blackish discoloration of fingers.

pain and cyanosis of the fingers had occurred. At first admission to our department he was afebrile. Physical examination showed bilateral bluish to blackish discoloration of the fingers (figure 1). There were small areas of necrosis over the pulps of the left and right index fingers (figure 2). The peripheral and central pulses were equal and regular bilaterally. There was no rash on the skin. Results of the laboratory tests were as follows: white cell count 16 000/mm³ (71% segmented neutrophils, 23% band forms), Hb 12.2 g/dL, platelet count 313 000/mm³, erythrocyte sedimentation rate 35 mm/h,

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Figure 2 (A and B) Areas of necrosis over the pulps of the index finger.

C reactive protein 0.55 mg/dL (normal value <0.8). Renal and liver functions were within normal limits. His coagulation tests (PT and aPTT) were normal, antiphospholipid antibodies and antinuclear antibody (ANA) were negative. Transthoracic echocardiography revealed normal cardiac anatomy and did not show any intracardiac mass, thrombus or vegetation suggestive of an embolic process. The Doppler ultrasound of the upper extremities showed bilateral monophasic flow without any sign of thromboembolism. This result suggested to us peripheral digital vasospasm. We started once daily subcutaneous dose of 100 IU/kg nadroparin, 4 mg/kg/day aspirin, 1 mg/kg/day nifedipine and 1 mg/kg/day sildenafil. At the end of the fifth day of treatment no improvement was observed. Cold agglutinins, ANAs, pANCA, cANCA, Factor V Leiden mutation were negative. Serum C3, C4 and C3a, anticardiolipin antibodies, protein S, protein C, antithrombin III were normal. Owing to the rapid progression of necrosis, intravenous iloprost 2 ng/kg/min, intravenous steroid and hyperbaric oxygen were started. Iloprost was continued for 6 h/day for 4 weeks. At the end of the first month of treatment, the necrotic tips separated and the fingers healed. We stopped intravenous iloprost and added azathioprine and bosentan. Two months later there was a small area of ulceration on the pulp of the right finger of the hand (figure 3). Steroid treatment was stopped gradually but azathioprine and bosentan were continued with reduced doses. No adverse effects of mentioned drugs (eg, endocrinological, haematological and hepatotoxic adverse effects) were observed. He is still being followed up on as an out-patient with nearly normal findings.

DISCUSSION

RP refers to transient vasospasm of peripheral arteries and arterioles.⁵ In primary RP, vasospasm does not have any association with other illnesses. Secondary RP has association with other conditions, most commonly autoimmune diseases such as systemic sclerosis, systemic lupus erythematosus and polyarteritis nodosa.^{1–3} Some drugs such as ergotamine, β -blockers, clonidine, cocaine and some other systemic disorders such as hypothyroidism, cold agglutinin syndrome can cause RP. There are reports of infectious diseases causing RP in the literature.¹ Emotional stress and cold can aggravate vasospasm by releasing endothelin 1, catecholamines and other vasoconstrictors. In some patients with severe RP, endothelin 1 and tumour necrosis factor- α can increase the activation of neutrophils and platelets which contribute to the damage of endothelium.

A rare but serious cause of secondary RP is stings of some jellyfish types. *Physalia physalis* is a name of a species of jellyfish encountered in the Atlantic and Mediterranean waters. Stings of

Physalia physalis are more painful and severe than those caused by other jellyfish species. The contact areas may progress to vesicular, haemorrhagic, necrotic or ulcerative stages. Severe localised complications of *P physalis* stings may also include arterial spasm in the sting site that can result in distal digital gangrene.⁶

Digital gangrene is an important complication of secondary RP.⁷ Digital gangrene is reported in 3% of children with secondary RP but do not occur in children with primary RP in the largest paediatric series. In some studies 13% of patients with primary RP are finally diagnosed as secondary RP.⁴ It is a clinical dilemma as to which of the patients with primary RP will finally be diagnosed as secondary RP. Determination of avascular fields, giant capillaries and irregular architecture of nailfold capillaries are important predictors of the development of secondary RP due to some connective tissue disorders. Normal nailfold capillary and the absence of ANA are suggestive of primary RP.

The treatment of mild RP is usually non-pharmacological and depends on precise underlying conditions. Avoidance of exposure to emotional stress or cold is a basic precaution. But in moderate-to-severe cases, vasodilator and antithrombotic agents are useful. Iloprost, endothelin-1 receptor antagonists and phosphodiesterase inhibitors are preferred for severe forms.^{1 8 9} Severe necrotising vasculitis associated with connective tissue disorders can be treated with steroids and cytotoxic agents such as cyclophosphamide and azathioprine.¹⁰

All the laboratory tests were negative in our patient which were performed to show underlying conditions. He only had leukocytosis and elevated sedimentation rate. Interestingly he had the history of red papules or erythematous rashes on the hands after the jellyfish sting which was initially diagnosed as hand-foot-mouth disease. Although the absence of ANA was suggestive of primary RP but as mentioned before 13% of patients with primary RP are finally diagnosed as secondary RP. Jellyfish sting was also recognised as an important triggering factor. Owing to progressions of ischaemic lesions, we urgently used vasodilators, immunosuppressive agents as well as hyperbaric oxygen. The relatively rapid recovery after immunosuppressive drugs suggested that this is real RP. Additionally, the patient is closely monitored for the possibility of the development of secondary RP as mentioned before.

In conclusion, this case report emphasises that the diagnosis of RP in children is confusing. We suggest that children with digital necrosis should be managed urgently to avoid serious complications and should be followed up closely to ensure an accurate diagnosis. Jellyfish stings should also be recognised as a rare trigger of RP.

Figure 3 Recovered fingers after treatment.

Learning points

- ▶ Raynaud's phenomena due to a jellyfish sting is a challenging diagnosis in a child.
- ▶ Jellyfish stings are also the cause of Raynaud in children.
- ▶ Digital necrosis of a child is a severe condition which requires prompt therapeutic approach.
- ▶ This condition also needs a careful follow-up in order to avoid infection or even amputation of a finger.

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