Raynaud's phenomenon in paediatric age

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DESCRIPTION

A 9-year-old female patient, with irrelevant personal and family history, was brought to the paediatrician's office because of vasospastic phenomena in hands and feet, bilateral and symmetrical, with 6 months of progress ([figure 1](#)). They usually last from 10 to 15 min with spontaneous resolution. Phenomena occurs once or twice a week, sometimes fortnightly and are triggered by cold, not by stress or pain. She denied systemic symptoms (asthenia, arthralgias, myalgias, fever, weight loss and so on). The examination showed acrocyanosis and sweating in hands and feet. No oedema, arthritis, fever or other changes. Modified Rodnan total skin thickness score was zero. Laboratory investigation showed positive rheumatoid factor, antinuclear antibodies (ANA) and anticentromere antibodies. Nailfold capillaroscopy revealed heterogeneous capillaries, observing niches of fine capillaries alternating with areas of rarefaction (ischaemia) with megacapillaries and giant capillaries, with spontaneous rupture and periungual haemorrhage ([figure 2](#)). The findings are compatible with secondary Raynaud's phenomenon (RP), with active scleroderma pattern. The two dimensional and M-mode echocardiogram showed no signs of pulmonary arterial hypertension. The chest radiography and the lung function tests did not reveal any changes. Pulmonary CT scan was not performed. The child was referred for ophthalmology (no alterations) and rheumatology. She started treatment with nifedipine and nitroglycerin in SOS on colder days. She continues to be followed in rheumatology consultation every 4 months or earlier if new symptoms or interurrences appear because of eventual scleroderma (score 8 in 2013 American College of Rheumatology - European League Against Rheumatism (ACR-EULAR) classification criteria).¹⁻³

RP is caused by episodic spasm of the small arteries and arterioles of the fingers, in more severe cases it may also affect the nose, ears or tongue.⁴⁻⁹ It is triggered by several stimuli including exposure to

Figure 2 Videocapillaroscopy images.

cold or a stressful event.¹⁻⁸ The afflicted area goes through a three-colour variation, from white, to purple and red (some cases only two colours).^{4-6 8-10} Its prevalence in paediatric age is unknown, but it is believed to be a rare and predominantly female entity, with most cases described in children aged 12-15 years.^{6 8 10 11}

According to the aetiology, RP can be primary/idiopathic (80% of cases) if symptoms occur in the absence of any associated disorder. Secondary RP is attributed to an underlying condition, most commonly connective tissue diseases including systemic lupus erythematosus or systemic scleroderma and so on.⁴⁻⁸

Diagnosis can be made using clinical history, physical observation, laboratorial and imaging studies.^{1 9-11} ANA's positivity and changes in capillary bed are suggestive of secondary RP.^{1 5 10-12} Differential diagnosis includes: excessive cold sensitivity, external compression of blood vessels, peripheral neuropathy, complex regional pain syndrome, occlusive vascular disease, acrocyanosis, erythromelalgia and acute idiopathic blue

Figure 1 Raynaud's phenomenon.

Learning points

- ▶ Raynaud's phenomenon (RP) is a rare entity in paediatric age, and every patient should be carefully evaluated to distinguish primary from the secondary RP.
- ▶ In patients with symptoms or signs suggestive of systemic disease, laboratory tests should include antinuclear antibodies and other specific autoantibodies (like anticentromere), depending on the suspected underlying disease.
- ▶ Most cases are primary RP and the treatment is supportive.

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finger.^{4,8} Pharmaceutical agents including beta-blockers, cisplatin and bleomycin can also cause RP.⁵ Treatment includes patient education and eviction of stimulators.^{4,5,8,9} In the secondary RP cases or with intense symptomatology, pharmacological therapy is recommended (may include calcium antagonists, alpha-blockers, nitrates, prostaglandins, selective serotonin receptor antagonists, selective serotonin reuptake inhibitors, endothelin receptors antagonists, ACE, pentoxifylline and acetylsalicylic acid).^{4,5,8,9} Surgery is reserved for cases resistant to pharmacological treatment.⁸

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