The importance of nailfold capillaroscopy in children with rheumatic diseases

Sirs,

We read with interest the article published in Clin Exp Rheumatol by Ingegnoli et al. (1) describing nailfold capillaroscopy (NFC) abnormalities in childhood-onset rheumatic diseases. The authors found the presence of a “scleroderma pattern” in children with systemic sclerosis (jSSc), dermatomyositis (jDM) and primary Raynaud’s phenomenon (PRP), “Normal” or “non-specific pattern” was found in systemic lupus erythematosus (jSLE), mixed connective tissue disease (jMCTD), juvenile-idiopathic arthritis (JIA) patients and healthy children (HC). Major abnormalities were also found in jSLE, jMCTD and systemic onset-JIA, although minor abnormalities were frequent (both ‘elongated <10%’ and/or ‘tortuous <50%’ capillaries). No other clinical or immunological features were evaluated. We aimed to assess NFC patterns in children with PRP (Raynaud’s Phenomenon with negative ANA testing) and juvenile-onset CTD patients with and without Raynaud’s phenomenon (RP). Socio-demographics, complete or incomplete RP presentation and anti-nuclear antibodies (ANA) data were collected. A NFC with DINOLite 300x®-USB camera was performed following recommended procedures. NFC patterns were established based on specific literature-suggested definitions (1, 2). A total of 115 NFC (72.5% girls) were performed, mean age 13.8 ±3.19 years old. Disease distribution was: 61 patients with PRP, 16 jDM, 11 jSLE, 10 jMCTD, 3 systemic vasculitis, 1 jSSc, 1 juvenile-anti-phospholipid syndrome, and 12 HC. Among CTD patients, a) 58.8% did not showed RP, 27.4% of patients showed complete RP and 13.7% incomplete RP; b) 50.9% showed positive ANA with following titres: 1/80 in 25.5%, 1/160 in 13.9%, 1/320 in 16.2%, 1/640 in 37.2% and 1/2560 in 6.9% of patients; c) 60.7% showed “normal” NFC pattern, 36.2% “non-specific pattern” and 2.9% “scleroderma pattern”. NFC patterns from all participants according to ANA result are shown in Figure 1. No statistical differences in NFC patterns were observed regarding with the assessed variables. However, ANA negative patients more frequently showed “normal” NFC pattern compared to ANA positive patients (71% vs. 48%, respectively; p=NS). None of ANA negative patients showed “scleroderma pattern” on NFC. CTD that showed abnormal NFC patterns were jSSc, jDM and jSLE. Limitations of our study were the observational nature design. When ANA tests are positive, NFC might help as a diagnostic and prognostic tool for juvenile-onset CTD diagnosis within six months, as suggested by Pain et al. (2). This algorithm would benefit patients’ journey to the paediatric rheumatologist, including costs, both parents’ and patients’ expectations and appropriate counselling. Moreover, Pavlov-Dolijanović et al. (3) have suggested that those patients who present RP would develop a connective tissue disease within 6 months after a “scleroderma pattern” detection on NFC. Therefore, ANA positive become a prognostic factor to develop CTD alongside with NFC (4).

As suggested by Ingegnoli et al. (1, 5), interpretation of NFC is dependant of the observer experience and might provide significant information in diseases rich as jDM and jSSc, mainly. However, other different CTD would not significantly benefit from NFC assessment. Recently, a ‘fast track algorithm’ has been applied to help to clarify clinically significant NFC patterns that are associated to adult CTD (6). Interestingly, a combination of ANA testing, NFC and a paediatric version of the ‘fast track algorithm’ application in children with RP would help paediatric rheumatologists to properly address these patients.

In conclusion, we suggest performing NFC in children with CTD suspicion with ANA positive, irrespective of the presence of complete or incomplete RP.

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References