

## Nail involvement in systemic sclerosis

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**Background:** Nail involvement has rarely been recognized in systemic sclerosis (SSc). Indeed, only a few small series have assessed nail changes in SSc, most of which are case reports.

**Objective:** The aims of the current case-control study were to: (1) determine the prevalence of fingernail changes in SSc; and (2) evaluate the correlation between fingernail changes and other features of SSc.

**Methods:** In all, 129 patients with SSc and 80 healthy control subjects underwent routine fingernail examination.

**Results:** The prevalence of fingernail changes was 80.6% in SSc. Patients with SSc more frequently exhibited: trachyonychia ( $P = .006$ ), scleronychia ( $P < .0001$ ), thickened nails ( $P < .0001$ ), brachyonychia ( $P = .0004$ ), parrot beaking ( $P < .0001$ ), pterygium inversum unguis ( $P < .0001$ ), splinter hemorrhages ( $P < .0001$ ), and cuticle abnormalities ( $P < .0001$ ) than healthy control subjects. The presence of fingernail changes was associated with digital ulcers ( $P < .0001$ ), calcinosis cutis ( $P = .004$ ), and higher values of mean nailfold videocapillaroscopy score ( $P = .0009$ ).

**Limitations:** The cohort originated from a single center.

**Conclusion:** This study underlines that fingernail changes are correlated with more severe forms of SSc characterized by digital microangiopathy, including digital ulcers and calcinosis cutis. Nail changes should be systematically checked in all patients with SSc, and may be included in the American College of Rheumatology/European League Against Rheumatism classification criteria for SSc. (J Am Acad Dermatol <http://dx.doi.org/10.1016/j.jaad.2016.11.024>.)

**Key words:** diagnosis; digital ulcers; nail involvement; predictive factor; systemic sclerosis.

Systemic sclerosis (SSc) is an inflammatory disorder, of unknown origin, affecting the skin and other organs.<sup>1-3</sup> Diffuse cutaneous SSc, interstitial lung disease, and pulmonary arterial hypertension have been associated with higher mortality in patients.<sup>4-9</sup>

Whereas skin abnormalities have been described extensively in SSc, nail involvement has rarely been recognized in these patients.<sup>10</sup> The aims of the current prospective case-control study were to: (1) determine the prevalence of fingernail changes in

unselected patients with SSc; and (2) evaluate the correlation between fingernail changes and other features of SSc.

### METHODS

#### Patients

Between January and December 2015, 129 consecutive patients with a definite diagnosis of SSc were included in the study. Criteria for diagnosis of SSc were based on the American College of Rheumatology/European League Against Rheumatism classification.<sup>11</sup>

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Ethical approval was obtained from the local ethical committee (Comité d'Éthique pour les Recherches Non Interventionnelles, Haute Normandie), and informed consent was obtained from all patients.

The cohort comprised 25 men and 104 women with a median age of 54 (range 21-82) years. The median duration of the disease, from onset of the first clinical manifestations of extra-Raynaud phenomenon, was 4.5 years. Patients were grouped into disease subset according to criteria by LeRoy et al<sup>12</sup>: diffuse cutaneous SSc (n = 46) and limited cutaneous SSc (n = 83). No patient had other connective tissue disorders.

Patients with SSc had pulmonary involvement as follows: interstitial lung disease (40.3%) and pulmonary arterial hypertension (7%). In all, 52 patients had digital ulcers/pitting scars (40.3%); digital ulcers were determined as denuded areas with a defined border, loss of epithelialization, and loss of epidermis and dermis on the volar aspect distal to the proximal interphalangeal joints.<sup>13</sup> Patients with SSc also had joint involvement (27.9%), severe esophageal motor impairment characterized by both aperistalsis and decreased low esophageal sphincter pressure on esophageal manometry (51.2%),<sup>2,14,15</sup> and history of renal crisis (2.3%).

Finally, 24 patients (18.6%) had calcinosis cutis, determined as ever evidence of subcutaneous calcium deposits, including history of calcium extruding from the skin as described by patients, on physical examination of the hand, on hand radiographs, or a combination of these.<sup>13</sup>

### Control subjects

The control group included 80 healthy subjects (15 men, 65 women) with a median age of 55 (range 23-80) years; they were age-matched ( $\pm 5$  years) control subjects. The control subjects were followed up in the department of internal medicine; none of them had connective tissue diseases, vasculitis, dermatoses, or endocrine disorders.

### Nail involvement

Fingernail examination was performed in all patients with SSc and healthy control subjects. Fingernail changes were classified as follows:

- Surface abnormalities: longitudinal ridging, transverse ridging, and pitting, which is determined by

pinpoint depressions within an otherwise normal nail plate;

- Consistency abnormalities: thickened nails, scleronychia, trachyonychia, and brittle nails, which are characterized by increased nail plate fragility;
- Coloration abnormalities: (1) leukonychia, determined by abnormal keratinization of the under-

lying nail matrix, resulting in white discoloration of the nail plate; (2) melanonychia, determined by a band of longitudinal black or brown pigmentation involving the nails; and (3) purple arch and perilunular redness;

- Curvature abnormalities: (1) digital clubbing; (2) parrot beaking,<sup>16</sup> which is determined by a peculiar, symmetric overcurvature of the free margin of the nails, with loss of substance of the nail plate

and the hyponychium<sup>10,17,18</sup>; and (3) koilonychia, presenting as transverse and longitudinal concavity of the nails;

- Other nail abnormalities: (1) hyponychium hyperkeratosis; (2) pterygium inversum unguis,<sup>19</sup> which consists of a forward extension of the hyponychium, anchoring to the undersurface of the nail plate and thus obliterating the distal nail groove<sup>10,18,20,21</sup>; and (3) brachyonychia, which is a sign of acro-osteolysis characterized by resorption of the distal phalangeal tufts<sup>17</sup>;
- Splinter hemorrhages: red-brown, longitudinal lines occurring in the nail bed;
- Cuticle abnormalities: irregularity, thickness, enlargement;
- Lunula abnormalities: absence of lunula, macrolunula, red or blue lunula.

### Nailfold videocapillaroscopy

Nailfold videocapillaroscopy (NVC) was performed in patients with SSc, using optical probe videocapillaroscopy equipped with a  $\times 200$  contact lens and connected to image analysis software. NVC was performed by the same operator, without knowledge of the patient's disease severity, in a blind manner. The following nailfold capillaroscopic parameters were considered, as previously described: presence of enlarged and giant capillaries, hemorrhages, loss of capillaries, disorganization of microvascular array, and capillary

### CAPSULE SUMMARY

- Fingernail changes are frequent in patients with systemic sclerosis (SSc).
- Some fingernail changes appear to be more specific of SSc; thorough fingernail examination may thus serve as a helpful diagnostic tool in SSc.
- Fingernail changes are correlated with more severe forms of SSc characterized by digital microangiopathy (digital ulcers and calcinosis cutis) and esophageal involvement.

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