

# Yellow Nail Syndrome: An Unusual Case Report

## Sarı Tırnak Sendromu: Nadir Bir Olgunun Sunumu

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**ABSTRACT** The yellow nail syndrome (YNS), a triad of yellow discolored, dystrophic nails, lymphedema, and pleural effusions, is a rare clinical condition. Most patients develop yellow nail syndrome in early middle age, and the overall male: female ratio is 1:1.6. The etiology of YNS is obscure, while the pathogenesis seems to involve impaired lymphatic drainage. A 17-year-old patient with yellow dystrophic nails, sinus retention cyst, and lower-limb lymphedema was presented with a review of the current medical literature.

**Key Words:** Malformed nails; lymphedema; maxillary sinus

**ÖZET** Tırnaklarda sarı renkli trofik değişiklikler, lenfödem ve plevral effüzyondan oluşan bir triad olan sarı tırnak sendromu (STS) nadir bir klinik durumdur. Bu hastaların çoğunda STS erken orta yaşlarda gelişir ve erkek: kadın oranı 1:1.6'dır. Patogenez bozulmuş lenfatik drenaja bağlı gibi gözükse de STS'nin etiolojisi karanlıktır. Biz mevcut tıbbi literatürü gözden geçirerek alt ekstremitelerde lenfödemi, sinüzal kist ve distrofik, sarı tırnaklarla başvuran 17 yaşında bir hastayı sunduk.

**Anahtar Kelimeler:** Malforme tırnaklar; lenfödem; maksiler sinüs

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The yellow nail syndrome (YNS) is an unusual lymphangitic disorder classically characterized by the presence of nail discoloration, lymphedema (80%) and pleural effusion (36%). A review of the literature, including over 150 patients, was presented. Most patients developed YNS in early middle age, and the overall male: female ratio was 1:1.6. Some etiologic aspects of this disorder remain poorly defined.<sup>1,2</sup> In this paper we describe a case with YNS and review the literature.

### CASE REPORT

A 17-year-old girl was referred with right leg swelling and pain. There was no positive familial history. She has realised her yellow nails for 5 years; initially the nails grew slowly; later they became thick and brittle and developed a yellow discoloration. Therefore she had gone to a lot of dermatologist; but any fungal infection had not been confirmed in nail exams and the appearance of her nails had not changed after antimycotic therapy. Ankle swelling had extended gradually to the knee. At this time she had not noticed any periorbital swelling. During the physical examination, we noted

that she has yellow dystrophic nails of fingers and toes, and edema of the right ankle without evidence of cardiac failure (Figure 1). Pulmonary auscultation was normal.

Her hemoglobin concentration, leukocyte and differential counts were within the normal range. Serum immunoglobulin concentrations were normal. Results of blood biochemistry and liver function tests and serum thyroxine concentration were normal but parathyroid hormone (PTH) was 8.6 pg/mL (ref. 10-65). Nail clippings contained no evidence of fungi.

Magnetic resonance lymphangiography with gadolinium and  $^{99m}\text{Tc}$  nanocolloid lymphoscintigraphy showed a lymphatic obstruction (Figure 2).

She did not give any history of chronic respiratory symptoms or chronic rhinorrhea, and her physical exam was normal; nevermore a CT scan was obtained because of the possibility of YNS. In fact, it showed a left maxillary sinus mucous retention cyst in spite of normal thoracic findings (Figure 3).

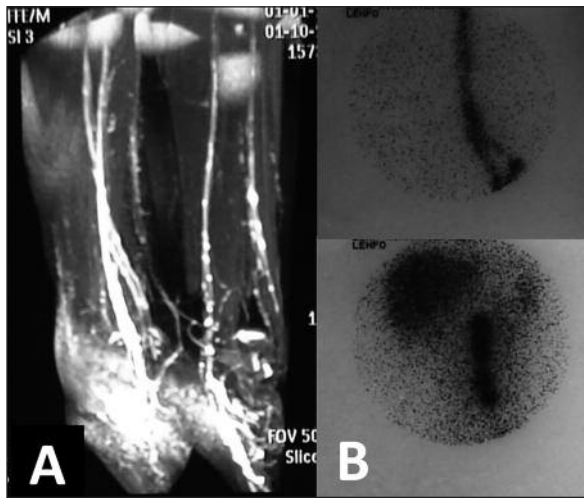
The patient was administered graduated elastic compression stocking, venoactive drug (MPFF), zinc, biotin and vitamine E and A. She was also referred to an otolaryngologist for her sinusual cyst.

## DISCUSSION

The “yellow nail syndrome”, the association of slow-growing, yellow, discolored nails with primary lymphedema, was firstly reported in 1964.<sup>3</sup>



**FIGURE 1:** Photograph showing lymphedema of right lower-leg (A) and symmetrical thickened, brown-yellowish dystrophic nails of fingers (B) and toes (C). (See for colored form <http://cardiovascular.turkiyeklinikleri.com/>)



**FIGURE 2:** (A) MR lymphangiography demonstrating dilated lymphatic channels in the right ankle and lateral calf; (B) Lymphoscintigraphy showing lymphatic obstruction in right lower extremity; note that activity accumulation in related to lymph nodes in popliteal and inguinal area can not seen.



**FIGURE 3:** Axial CT scan demonstrating an asymptomatic mucous retention cyst in the left maxillary sinus.

Samman and White hypothesized that the nail changes and lymphedema (most frequently of the lower extremity) may result from dysfunctional lymphatic drainage.<sup>3</sup> The respiratory component, idiopathic pleural effusion, is added to the syndrome after two years.<sup>4</sup> Although YNS has been defined as the complete triad of slow growing yellow nails, lymphedema, and pleural effusions, today it

is accepted that the presence of two of the three symptoms is sufficient to establish the diagnosis. The yellow nails, is not necessary for a diagnosis, are thickened and smooth and may show transverse ridging; they are excessively curved from side to side. Onycholysis is frequently present, and culture of the nails for fungi is always negative.

The cause of YNS is unknown. The underlying abnormality is presumed to be anatomical and/or functional defects in the lymphatic vessels, but so far this has not been demonstrated to be the cause of the nail changes, the pathogenesis of which remains obscure.<sup>1,2,5</sup> However, it is seen in patients with chronic bronchiectasis, recurrent pleural/pericardial effusions, chronic sinus infections, internal malignancies, breast carcinoma, immunologic abnormalities, immunodeficiency syndromes, macroglobulinemia, lymphopenia, and rheumatoid arthritis.<sup>2,5-9</sup> Nothing is known of the mechanism of production of the nail changes; there is no proof so far that they result from impaired function of the lymphatics.<sup>2,3</sup> Thyroid abnormalities including Hashimoto's thyroiditis, thyroid enlargement, hypothyroidism, thyrotoxicosis have been reported.<sup>6,10</sup> Low PTH level has been found in our patient. When YNS is suspected on the basis of the examination of the yellow nails, an extensive dermatologic, pneumologic, lymphographic, and biologic investigation has to be launched to exclude underlying neoplasma, or autoimmune disease, frequently associated with this syndrome.

The earliest age at which nail changes have been noted was 10 years and the latest, 82 years.<sup>2,5,11,12</sup> When our patient realised her nail changes, she was 12 years old. The diagnosis of YNS is based entirely on a combination of clinical findings for which all other possible causes have been excluded. No confirmatory tests exist and the pathogenesis of the condition is poorly understood. It has been suggested that this syndrome is basically the result of a congenital abnormality of the lymphatics and therefore could be related to primary lymphedema. In both primary lymphedema and the YNS females predominate. In our review of chronic limb lymphedema, of the patients with primary lymphedema, 80% were female; the ratio

of YNS was 2.5%. The various combinations of the three major features of the YNS may find: yellow nails alone; yellow nails and lymphedema; yellow nails, lymphedema and pleural effusions; lymphedema and pleural effusions; and yellow nails and pleural effusions. The individual features of YNS may occur at wide intervals, so that absence of the classic yellow nails does not preclude the diagnosis, provided all other possible causes of lymphedema and pleural effusions have been excluded.<sup>6</sup> Familial occurrence of YNS has been reported.<sup>13</sup> However, the lack of a positive family history in the majority of patients in a study, the late onset of the disease and recovery of nail changes in the patients suggest that YNS may not be primarily a genetic disease as it is currently classified.<sup>14</sup>

The treatment is controversial and poorly effective. Vitamin A, vitamin E (topical/oral, 1 200 mg daily), Zn sulfate, and biotin are drugs the most often advised.<sup>5,7,12,15</sup> Itraconazole cannot be recommended in patients with YNS.<sup>7</sup> Spontaneous healing is reported in 30% of the cases.<sup>3</sup> In most YNS cases, associated respiratory manifestations are generally manageable with a regimen of medical and surgical treatments.<sup>2,5</sup> When the sinusal mucosa is involved, endoscopic endonasal surgery in such cases is controversial and its outcome is not yet documented.<sup>15</sup> It also may achieve an im-

provement in lymphatic function and partial resolution of nails after complex decongestive physiotherapy in YNS.<sup>16</sup> The YNS patients may be treated with conservative management, including a low-fat diet supplemented with medium-chain triglycerides.<sup>17</sup> If endotracheal intubation is performed, the patients with YNS should be observed for 48 h. because acute laryngeal edema might develop.<sup>18</sup> Yellow nails improve in about one half of patients, often without specific therapy.<sup>5</sup> In various malignancies associated with YNS, the yellow nails, "paraneoplastic manifestation", improved dramatically after an effective cancer treatment such as surgery and/or chemotherapy.<sup>8,9</sup> There are some possible explanations such as direct involvement by tumor of already stressed and dysfunctional lymphatics or the elaboration of mediators such as peptide hormones that inhibit lymphatic function. The existence of YNS should raise the index of suspicion for malignancy and other associated diseases.

The yellow nail syndrome is a rare disorder in middle-aged and elderly individuals, which is typically characterized by pleural effusions, peripheral lymphedema, and slowly growing, dystrophic yellow nails. To our knowledge this is the first young case of yellow nail syndrome associated with an asymptomatic sinus retention cyst.

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