

## Roentgenographic and Clinical Signs in Yellow Nail Syndrome

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### Summary

The yellow nail syndrome is a rare condition consisting of thickened opaque yellow nails, lymphedema and respiratory symptoms such as bronchitis or pleural effusions.

In a 25 year-old woman, presenting all clinical signs of this syndrome, lymphangiography revealed impaired lymphdrainage of both legs. Both, number and size of the lymph vessels and regional lymph nodes were found to be reduced and several extravasations of the contrast medium were noted.

Lymphangiography may help to establish the diagnosis of yellow nail syndrome, especially if the association of clinical anomalies is incomplete.

The yellow nail syndrome includes yellow, slow-growing nails, lymphedema, and respiratory manifestations such as sinusitis, bronchitis or pleural effusions. The syndrome was first described by *Samman* and *White* (10) in 1964. The etiology is unknown. So far, 38 cases have been reported. The syndrome predominantly occurs in middle-aged patients of both sexes (8, 10). The nail changes involve both fingers and toes and usually precede the other signs and symptoms of the syndrome by many years.

The nails are thickened, opaque, yellow or brownish in color, and the lunula are no more visible. They show an excessive curvature on their long axis. Their growth rate is extremely slow.

Lymphedema usually appears a few years after the onset of nail changes (9, 10). In some cases, however, congenital lymphedema (7) or familial primary hypoplasia of the lymphatics (11) were present. The edema is most often located around the ankles, but the arms and the face may also be involved.

Respiratory disorders were found in most cases. They consist in recurrent infections of the upper respiratory tract, often associated with bronchopneumonia, bronchiectasis and secondary pulmonary fibrosis (1, 2, 6, 9, 12). Chest X-ray often reveals pleural effusions that are asymptomatic in spite of their extent (3, 4, 5, 6, 8, 9).

### Report of a case

The 25 year-old woman had noticed her nail changes since 1966. Her toe nails, and later on her finger nails, showed a yellowish-brown color and were growing less rapidly. Since childhood she had frequent attacks of bronchitis and sinusitis, often associated with fever. Since 1969 she suffered from recurrent edema of the eyelids, the cheeks and the legs. Her family history was non-contributory. When she was first seen at the Department of Dermatology in December 1975, her finger and toe nails were thickened, opaque and yellow. They were excessively curved on their long axis and showed transversal ridging. The first diagnosis was onychomycosis, since *Candida albicans* had been cultured from nail scrapings. Therefore, all of the toe nails and seven of her finger nails were extracted, and local antimycotic treatment was performed over a period of one year. Despite of this treatment the nail regrew in the same manner (Fig. 1). Now, the diagnosis of yellow nail syndrome was suspected. Further examination revealed that the patient suffered from chronic bronchitis. Coarse, crepitant rales were heard over both lungs. Moreover, edema of both cheeks, but more marked on the right side, and a mild



Fig. 1 Characteristic onychodystrophy of the yellow nail syndrome

edema of the ankles and feet were noted. X-ray examination of the chest showed no abnormalities.

In January 1977, a lymphography was performed. After subcutaneous injection of Pa-

tent-Blue-Violet into the first and fourth interdigital folds of both feet, a typical dermal backflow was noticed. Fifteen minutes later, the cutaneous lymphatics showed an intense coloration about ten centimeters above the ankles of the right foot. On the left side, a



Fig. 2a and b Lymphangiogram of the right leg



Fig. 3a and b Lymphangiogram of the left leg

less intensely colored area of cutaneous lymphatics was noted in the middle of the lower leg.

The skin of the dorsum of both feet was thickened. For cannulating we found very thin lymph vessels with rigid walls. A few minutes after starting the injection of Lipodol-UF, we saw fluoroscopically the contrast medium in the cutaneous lymph fissures. An extravasation of Lipiodol-UF occurred in the same area previously colored by Patent-Blue-Violet. Four lymph vessels could be seen on the right side and two on the left side. They were found to be very thin and tortuous, showed several small extravasations, and had a beaded appearance (Fig. 2 and 3). The number and size of the iliac and aortal lymph vessels were also reduced.

On lymphadenograms performed 48 hours later, we found fewer and smaller lymph nodes than normal. The lymph nodes stored, however, the contrast medium very intensely. Even one week later, the contrast medium could be found in the above mentioned cutaneous regions (Fig. 4).

#### Discussion

The lymphographic findings of this patient are consisting with those reported in other cases of yellow nail syndrome. *Samman* and *White* (10) observed in their patients a dermal backflow after the interdigital subcutaneous injection of Patent-Blue-Violet. Occasionally, they observed only one colored lymph vessel on the dorsum of the foot. These authors also described

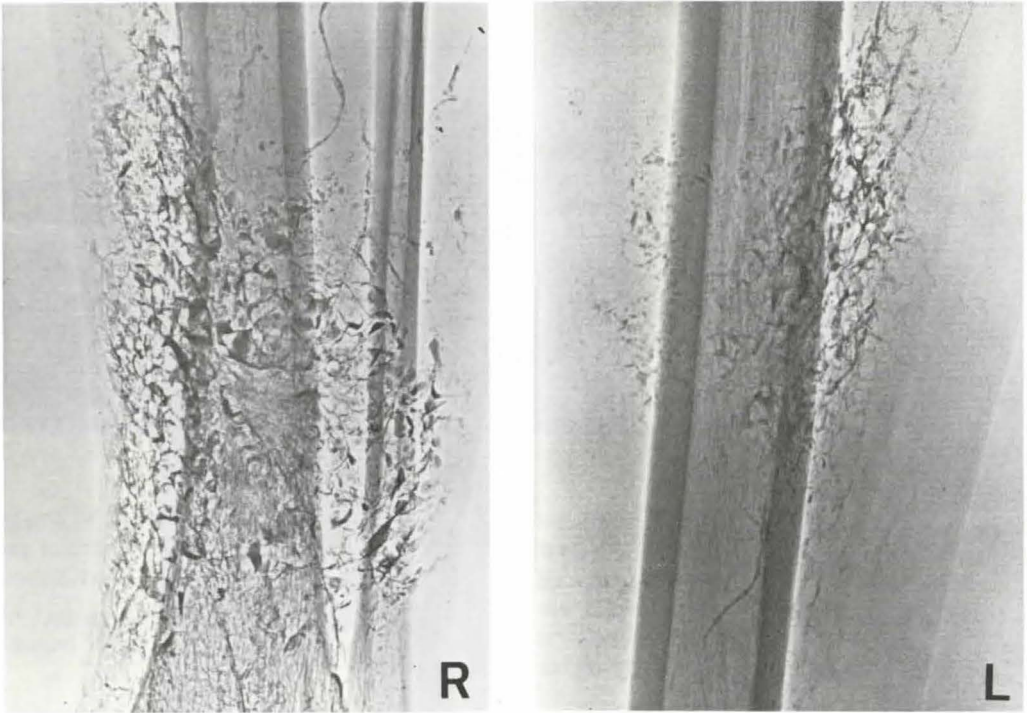


Fig. 4a and b Xeroradiograms of both lower legs one week after lymphography. Stasis of contrast medium in the cutaneous lymph fissures and the lymphatics

the rigid and thickened skin, presumably resulting from the edema in this region. Meiers et al. (8) reported that the dissected lymphatics had rigid walls and extremely small lumina. Cannulation was difficult or even impossible.

The visualisation of only tiny lymph collector in the calf with interspersed ectatic portions has already been reported by several other authors (3, 5, 10). They also observed a reduced number of collectors on the thigh and fewer lymph nodes than usual in the iliac and aortal regions (9).

The extravasation of Lipiodol-UF is most likely due to the injection pressure causing ruptures of the rigid lymphatics (8, 10).

Histologically, the walls of the lymphatics are thickened; they may obliterate the lumen (8).

The lymphographic findings may constitute

important diagnostic criteria of this syndrome, especially if the association of the other signs and symptoms is less complete as in the case presented here.

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