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YELLOW NAIL SYNDROME IN A 10-YEAR-OLD GIRL*

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A 10-year-old girl with yellow dystrophic nails, bronchiectasis, chronic sinusitis and lower-limb lymphedema is presented. The underlying mechanism remains unkown although it has been postulated to be associated with lymphatic abnormalities. To date no causative treatment exists. Our patient was treated with conservative management, including a low-fat diet supplemented with medium-chain triglycerides. Moderate improvement in the lymphedema of the lower extremities was observed. To our knowledge this is the first case of yellow nail syndrome to be treated with diet.

Key words: bronchiectasis, chronic sinusitis, dystrophic nails, yellow nails, lymphedema, special diet.

Yellow nail syndrome is characterized by yellow dystrophic nails in association with lymphedema and respiratory findings, including pleural effusion, bronchiectasis and chronic sinusitis[1-5]. The etiology of this syndrome still remains obscure[6]. Hiller[7] and Beer[8] postulated that the cause of the pulmonary manifestations and lymphedema was hypoplastic or deficient lymphatic vessels. Most published cases have been adults[1-4,7,8]; children with yellow nail syndrome have rarely been reported[5,6]. We present a 10-year-old girl with yellow dystrophic nails, lymphedema, bronchiectasis and chronic sinusitis.

Case Report

A six-year-old girl presented with recurrent respiratory symptoms. Physical examination demonstrated nail changes comprising yellowish discoloration of the nail plate, thickening and exaggeration of the lateral curvature. The remainder of the physical examination was unremarkable. Laboratory investigation revealed a normal hemogram, alpha-1 antitrypsin level, quantitative immunoglobulins and sweat chloride test. Chest radiograph and computed tomography of the thorax showed mild bronchiectasis of the lower left lobe (Fig. 1, 2). Sinus roentgenograms showed a bilateral maxillary sinusitis.

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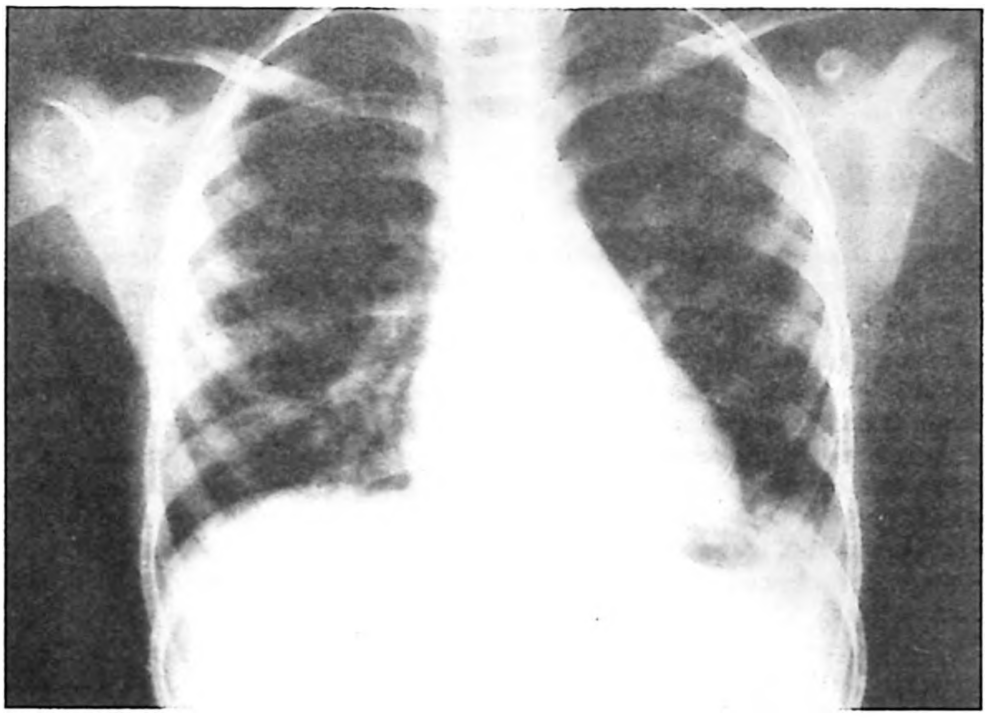

Fig. 1: Chest roentgenogram showing bronchiectasis in the lower left lobe.

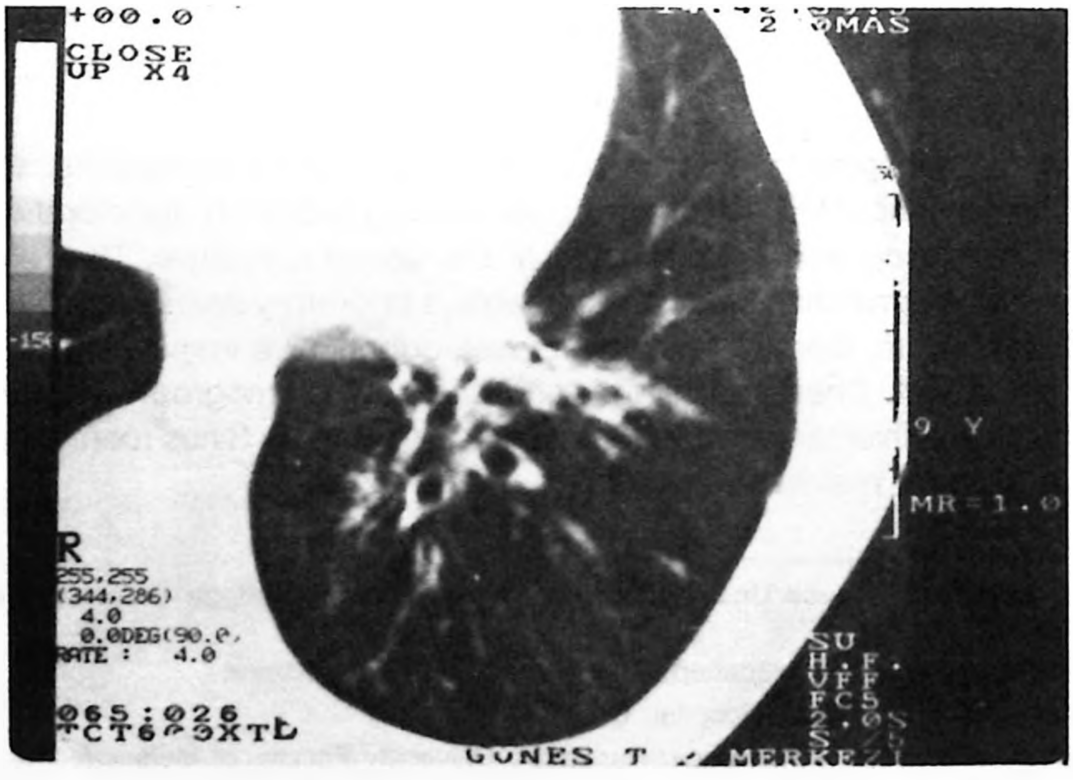

Fig. 2: Computed tomography of the thorax showing bronchiectasis in the lower left lobe.

The nail changes were diagnosed as tinea unguinum. While receiving symptomatic therapy for mild bronchiectasis and sinusitis, the yellow, dystrophic nails were remarkable (Fig. 3). All fingernails and toenails had a yellowish, thickened appearance with increased lateral curvature and loss of the culticles. Repeated nail cultures and smears for bacteria and fungus were negative; thus the diagnosis of tinea unguinum was ruled out.

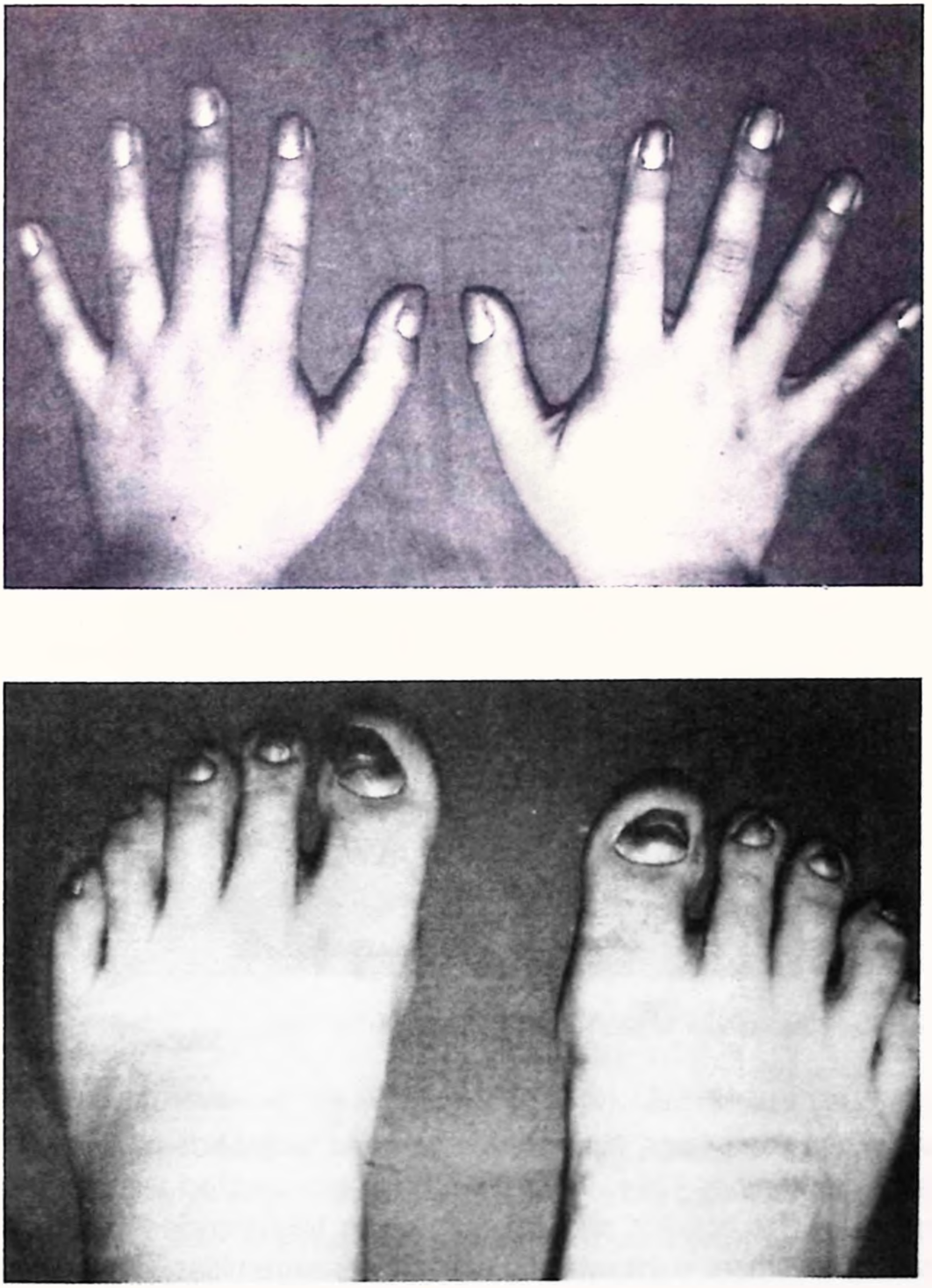

Fig. 3: Fingernails and toenails are yellowish, thickened and opaque, with increased lateral curvature and no visible lunulae.

Three years later, the patient complained of swelling of the lower extremities, and lymphedema was diagnosed (Fig. 4). Doppler ultrasonographic examination revealed normal venous and arterial systems. Lymphangiograms of the lower extremities were abnormal with hypoplastic lymphatic vessels; together with the characteristic nail changes, bronchiectasis and sinusitis, the diagnosis of yellow nail syndrome was established.

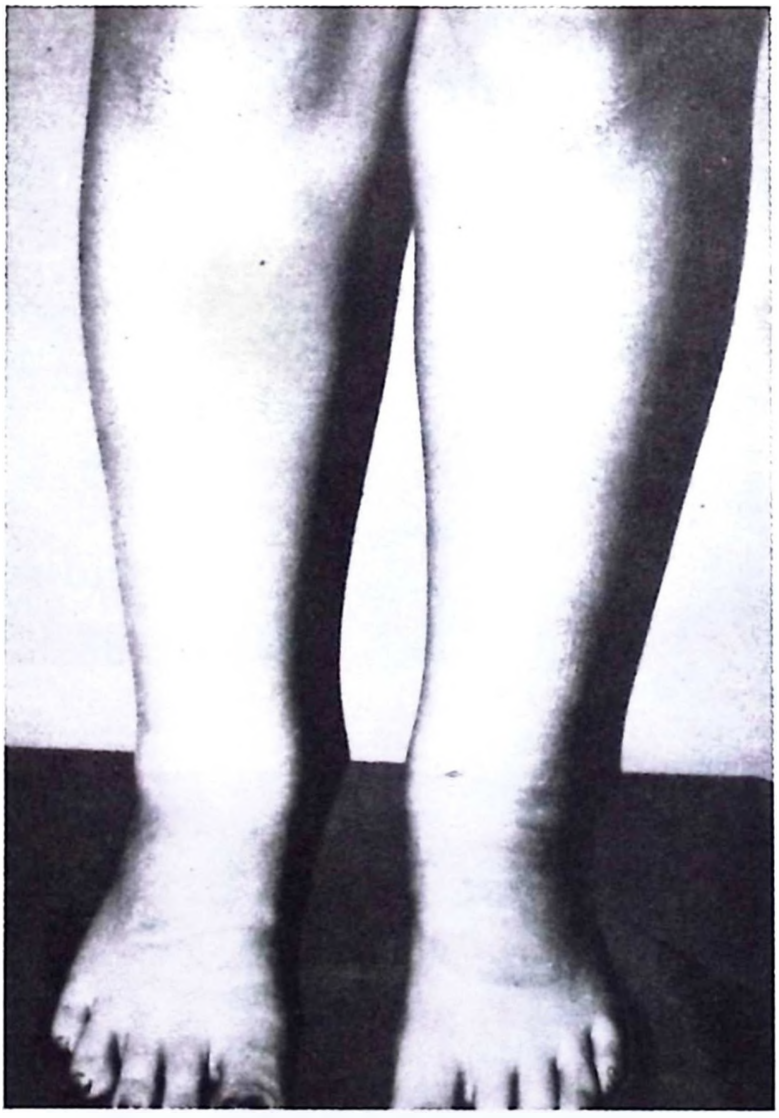

Fig. 4: Lymphedema of the legs.

At follow-up two years later, the nail changes and lymphedema were worse than ever. Since September 1994, the patient has been treated with a specific diet, including a low fat content and medium-chain triglycerides. There has been moderate improvement in the swelling of the lower extremities, but the nail findings have remained unchanged. Because of the improvement in the lymphedema, the diet is being continued.

Discussion

Although yellow nail syndrome was first described in 1964 by Samman and White[1], children with uncommon findings including dystrophic nails and lymphedema had been reported in 1960[5-6]. The classical triad of yellow nail syndrome are slow-growing yellow nails, lymphedema and respiratory manifestations, including pleural effusions, bronchiectasis and bilateral chronic maxillary sinusitis. The main cause of the disease is unknown. The age at onset of symptoms is variable, ranging from birth to the eight decade[2, 3]. Lymphedema is usually the first clinical manifestation, but in some cases nail findings appear first. Eastwood et al.[3] reported a patient with lymphedema beginning in childhood who developed classic nail changes in the eighth decade and pleural effusion in the nineth decade. When any one manifestation of the triad is present, the others should be sought, and the patient should be followed for further developments. The pathogenesis of the pulmonary manifestations is unknown. Airways are richly supplied with lymphatic vessels. It is possible that the hypoplastic lymphatics play a great role in the pathogenesis of bronchial and paranasal sinus infections[7-8]. In some patients bronchiectasis is the only manifestation of pulmonary involvement[7]. No specific treatment is available at the present time.

Conservative management, including a low fat intake supplemented with medium chain triglyceride, which are absorbed directly into the portal venous system, has been suggested in children with cylothorax. We initiated the same diet, including a low fat intake supplemented with medium-chain triglycerides, considering the hypoplastic lymphatic vessels to be the possible etiopathogenesis of yellow nail syndrome. To our knowledge there has been no report regarding therapy for this uncommon disease. This is the first case of yellow nail syndrome that has been treated with a special diet.

REFERENCES

1. Samman PD, White WF. The "yellow nail" syndrome. Br J Dermatol 1964; 76: 153-157.
2. Govaert P, Leroy JG, Pauwels R, et al. Perinatal manifestations of maternal yellow nail syndrome. Pediatrics 1992; 89: 1016-1018.
3. Eastwood HD, Williams TJ. Pleural effusions and yellow nails of late onset. Postgrad Med J 1973; 49: 364-365.
4. DeCoste SD, Imber MJ, Baden HP. Yellow nail syndrome. J Am Acad Dermatol 1990; 22: 608-611.
5. Lorber J, Illingworth RS. Generalised oedema of obscure origin. Acta Paediatr 1960; 49: 748-751.
6. Jones HE. Symmetrical peripheral oedema in infants. Arch Dis Child 1990; 35: 192-196.
7. Hiller E, Rosenow EC, Olsen AM. Pulmonary manifestations of the yellow nail syndrome. Chest 1978; 61: 452-458.
8. Beer DJ, Pereira W Jr, Snider GL. Pleural effusion associated with primary lymphedema: a perspective on the yellow nail syndrome. Am Rev Respir Dis 1978; 117: 595-598.