BRIEF COMMUNICATION

YELLOW NAIL SYNDROME OR DIFFUSE LYMPHATIC NETWORK DISEASE

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Summary: We report a man aged 68 years old with pneumothorax and chronic bilateral pleural effusion in association with a history of yellow nails. The diagnosis of yellow nail syndrome based on yellow nails, lymphedema, chronic pleural effusion and intestinal lymphangiectasia.

Key words: Yellow nail syndrome; Pneumothorax; Chronic pleural effusion

The yellow nail syndrome (YNS), a combination of yellow discolored nails, lymphedema and pleural effusions, is a rare autosomal dominant disorder of obscure etiology (10). YNS is a rare clinical condition. During the last 35 years only about a hundred cases have been reported. We present the clinical findings of a man who suffered from undiagnosed bilateral pleural effusions and lymphedema for 27 years and was admitted to pulmonary department with pneumothorax.

Case Report

Three months ago, a 68–year-old patient was admitted to the emergency department with dyspnea, thoracic pain and chronic lower extremity edema. Clinical examination revealed bilateral effusions, left pneumothorax and yellow discoloration of the nails of the feet (Fig. 1). He told us that the yellow color of the nails and the peripheral edema and edema in eyelids first appeared 27 years ago. He did not ask for medical advice for 13 years, until he developed progressive dyspnea with gradual worsening of his exercise tolerance and bilateral pleural effusions in x-ray. Laboratory examinations in many general (Greek and German) hospitals revealed in addition to bilateral pleural effusions, a latent hypothyroidism thirteen years ago. Pleuroscopy revealed no abnormal findings. He was released with medicaments thyroxin and spironolactone. Due to normal thyroxin blood levels, he stopped ten months later using thyroxin. Thoracocentisis was used when dyspnea was worsening. He submitted during the last six years in three thoracocentisis and the last thoracocentisis took place nine months ago during his last hospitalization.

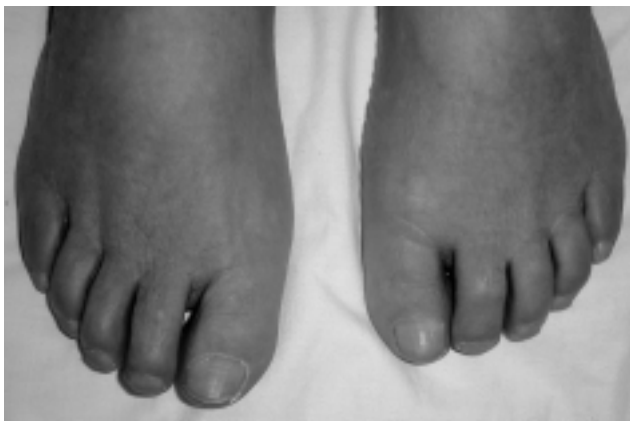

Fig. 1: Yellow nails in the lower limbs.

Our examinations showed normal hemoglobin level of 14.1 g/dl, normal leukocytes (8800/mm^3, 66% neutrophils) and ESR 45mm in the first hour. Albumin was reduced at 2.9 g/dl. All other biochemical agents were at normal levels. Immunoglobulins and thyroid function test were also at normal levels. Chest x-ray examination revealed left pneumothorax besides the bilateral pleural effusion. Billau system was used to control the pneumothorax successfully. Three paracentheses in the right hemithorax gave together about 4 l of fluid. The type of the liquid in both sides was exudative. Also gastroscopy and biopsy from jejunum were performed during his hospitalization. In some sections there were lymphangiectasis. When liquid stopped reproduction, we took off the Billau system. He was released without dyspnea and with advice to use diuretics.

Discussion

YNS was first described in 1964 by Samman and White (10). The full syndrome is characterized by rhino-sinusitis,

pleural effusion, bronchiectasis, lymphedema and dystrophic yellow nails (8,9). Individual manifestations of the syndrome may appear at different ages beginning from birth up to late adult life (2,11,12). Most patients develop YNS in early middle age and the overall male: female ratio seems to be 1:1.6 (9). YNS result from slow growth, possibly secondary to defective lymphatic drainage (7). Historically, nail changes were the first to be recognized. The slow rate of nail growth may be accompanied by color changes (pale yellow/green), onycholysis, and occasionally a distinct hump of the nail (10). Moreover, spontaneous clearing of the nail changes has been reported without resolution of the respiratory involvement (3). Pleural effusion appears to be a later manifestation. Our patient presented dyspnea due to pleural effusion eleven years later after the YNS onset. Pleural and pericardial effusions, chylous ascites and persistent hypoalbuminaemia can be explained by possible microvascullar permeability (1). Parietal lymphatic is the major route of pleural fluid exit. Fluid comes in the pleural cavity from the microvessels of systemic circulation (parietal pleura) and bronchial network (visceral pleura) (4,5). Dysfunction of lymphatic stomata is the reason of the pleural effusions in the YNS. Jejunal and duodenal biopsies may show the typical histological findings of intestinal lymphangiectasis, which may contribute to the pathogenesis of this syndrome (6).

YNS manifestations from respiratory system include bronchiectasis (14), which can be detected by high – resolution computed tomography (13). The cause of bronchiectasis is unclear, but again dysfunctional lymphatics are thought to play an important role with compromised drainage of secretions and local immune function (13). The reason of pneumothorax may be the persistent infection in bronchiectasis or the bad condition of pleura disturbance of the lymphatic system or to repeating thoracocentisis of the patients.

Chronic bilateral pleural effusion may be an index of suspicion of YNS. Dystrophic yellow nails especially in the lower limbs may appear many years before. Pneumothorax is an unusual and unclear sign of the syndrome. Disorder of the lymphatic system is the reason which could explain all the signs of YNS.

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