

## Case Report

# Chemical Pleurodesis Could Exacerbate Lymphedema of Yellow Nail Syndrome

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Chemical pleurodesis is sometimes performed for the management of intractable pleural effusion. We describe a woman with yellow nail syndrome (YNS), which is characterized by yellow discoloration of the nails, lymphedema, and pleural effusion. At the age of 43, she was hospitalized with edema of the lower limbs. Despite a number of medical treatments, massive lymphedema of lower limbs developed over a period of three years, resulting in skin cracks and subsequent infection, septicemia and multiple organ failure. At autopsy, abnormally dilated lymph and blood vessels were evident in soft tissue throughout the whole body. She had undergone chemical pleurodesis at 36 years of age for reduction of pleural effusion associated with YNS. Our case illustrates possible complication of chemical pleurodesis to YNS, which resulted in accumulation of lymph flow into the lower half of the body.

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**Key Words:** chemical pleurodesis, yellow nail syndrome, pleural effusion, lymphedema

## Introduction

Yellow nail syndrome (YNS) was first described by Samman and White (1) as a combination of yellow discoloration of nails and lymphedema. Subsequently, Emerson (2) defined the YNS as the presence of a triad of dystrophic yellow nails, lymphedema, and pleural effusion. Chemical pleurodesis (CP) is sometimes performed to manage refractory pleural effusion of YNS. We re-

port here a case of YNS caused an exacerbation of lymphedema after CP.

## Case Report

A 43-year-old woman was admitted to our hospital for exacerbation of edema of the lower limbs. Past history included increase in body weight, oligouria, edema of the lower limbs and exertional dyspnea since she was 35. At the age of 36, bilateral CP was successfully performed, following failure of treatment with diuretics and corticosteroid, to reduce pleural effusion. At that time, she had yellow nails.

On admission, massive edema of the lower limbs were noticed. A plain chest X-ray showed bilateral thickening of the pleura due to CP. Considered together with the history of bilateral pleural effusion and yellow nails, she was clinically diagnosed as YNS. Despite a number of therapies, the edema of the lower limbs worsened gradually with repeated attacks of bronchopneumonia, and ultimately became incapacitating making it difficult for her to walk. A crack appeared on the skin of the lower limbs, and lymph fluid discharge was noted to a maximum volume of approximately 10 L/day. The upper half of the body was emaciated and its severity was inversely proportional with the extent of edema of the lower limbs. Finally, skin wounds on the inguinal area became infected, resulting in extensive cellulitis, which was associated with agranulocytosis. Blood cultures showed growth of *Alcaligenes Dentrificans xylosoxydans*. The patient died of multiple organ failure.

At autopsy, fluid accumulation was noted in the subcutaneous area of the lower limbs and abdomen, i.e., state of elephantiasis (fig. 1. a). Although 4,000 ml of bloody ascites was collected from the abdominal cavity, no fluid was detected in the thoracic cavity due

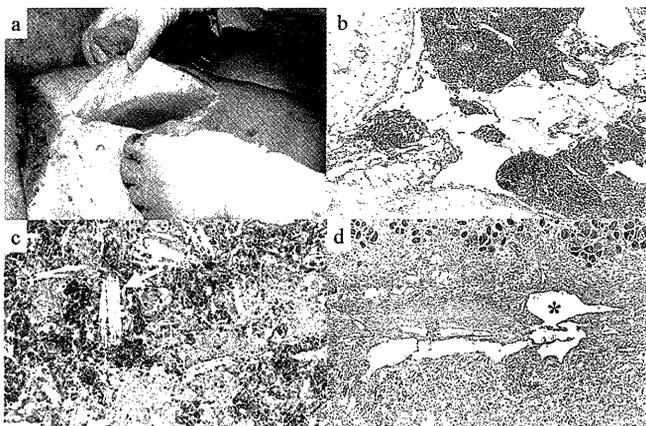
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to massive pleural adhesions caused by CP. Microscopic examination of the soft tissue revealed interstitial edema and the presence of abnormally dilated lymph and blood vessels throughout the body (fig. 1. b), although no clear abnormalities of such vessels were noted macroscopically. Lipid pneumonia consisting of foamy cells and cholesterol crystal was evident in both lungs (fig. 1. c) that weighed 285 g (right) and 550 g (left), and numerous abnormal vessels were evident in the boundary between the thickened pleura and lungs (fig 1. d). These findings suggested accumulation of edematous fluid in the lungs as a result of obstruction of the lymph flow caused by severe pleural fibrosis.



**Figure 1.** Pathological findings at autopsy. (a) Hematoxylin and eosin-stained specimens (b-d). Fluid accumulation under the thigh skin. (b) Marked dilatation of a lymph node sinusoid and lymph vessels in a soft tissue. (c) Lipid pneumonia with cholesterol crystals (arrow) in both lungs. (d) An unusual lymph vessel (asterisk) and blood vessels were evident in the boundary region between the pleura and lung. Original magnification: x8 (b), x50 (c), x16 (d).

## Discussion

YNS is characterized by yellow discoloration of the nails, lymphedema, and pleural effusion. Most patients develop YNS in early middle age, and the overall male: female ratio is 1: 1.6 (3, 4). Various degrees of lymphedema are described in about 80% of cases of YNS (2-4). The etiology is obscure, but it could be due to congenital anomalies of the lymph vessels. Various pathological processes including infections may stress the abnormalities in later years to cause frank edema (5, 6).

Pleural effusion of YNS is cryptogenic. Most cases show bilateral effusions, but unilateral cases are also reported (2-4, 6). Most such patients often have a history of recurrent exacerbations of chronic respiratory infections (2-4, 6). CP has been suggested for the

treatment of recurrent inveterate effusions of YNS, and Jiva et al. (7) reported its long-term effectiveness. Although they reported that the procedure was successful, it failed to control pleural effusion in 2 of 7 patients. CP was also performed in our case, which resulted in the control of pleural effusion, but subsequently resulted in progressive lymphedema of the lower extremities. In the present case, abnormally dilated lymph vessels were observed in almost every organ. It is likely that CP shifted the lymph flow to the lower part of the body, because the exacerbation of edema of the lower limbs and emaciation of the upper half of the body were progressive after CP. Thus, while CP is effective for the management of pleural effusions in YNS, it could potentially cause fatal exacerbation of lymphedema many years later.

Physician should be aware that CP may be effective for the treatment of pleural effusions in YNS, but it could cause severe and potential fatal lymphedema several years later.

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