Yellow Nails, Lymphedema and Pleural Effusion*

Treatment of Chronic Pleural Effusion with Pleuroperitoneal Shunting

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Pleural effusion secondary to lymphedema may be chronic, symptomatic and refractory to treatment, occasionally requiring invasive and painful procedures such as chemical pleurodesis, open pleural abrasion or pleurectomy to achieve control of the effusion and gain symptomatic relief. We report a patient with yellow nail syndrome and chronic pleural effusion successfully treated with pleuroperitoneal shunting.

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FPD = purified protein derivative; LDH = lactic dehydrogenase

The triad of yellow nails, lymphedema and pleural effusion was first described by Emerson in 1966 and was labelled the yellow nail syndrome. Since then, more than 100 patients with the classic triad or variants thereof have been described. Pleural effusions in this syndrome range from small, unilateral and asymptomatic to large, bilateral and debilitating. We describe a patient with yellow nail syndrome and symptomatic chronic pleural effusion inadequately controlled with frequent thoracentesis but successfully treated with pleuroperitoneal shunting.

CASE REPORT

A 65-year-old white woman was referred to the Pulmonary Clinic of the University of Chicago Hospitals and Clinics in November 1986 for evaluation of a left pleural effusion that was found on routine physical examination. The patient admitted to a several month history of mild fatigue and dyspnea upon walking several blocks. She noted pretilial and ankle edema bilaterally for several years, which was stable, and did not respond to leg elevation, support stockings or a trial of diuretics. She stated that since 1980, her nails had been thick, yellow and would break easily, and she always wore nail polish. She denied cough, fevers or night sweats. She had never smoked and had no known occupational exposure. There was no family history of lymphedema, bronchiectasis, sinusitis or immune deficiency.

Physical examination was remarkable for dullness and decreased breath sounds at the left lung base. There was 1+ pretilial and ankle edema, which pitted slightly and was not brawny. The nails were yellow, thickened, onycholytic and had transverse ridging. The lunulae were absent (Fig 1).

Chest radiograph revealed a large left pleural effusion with a normal heart and pulmonary vasculature (Fig 2). A 5-TU PPD was read as positive with 10 mm of induration. An echocardiogram was normal. Thoracentesis revealed straw-colored fluid with 150 red blood cells/μL; 2,800 white blood cells/μL; 93 percent lymphocytes; 5 percent macrophages; 1 percent eosinophils; 1 percent mesothelial cells. The protein was 5.5 g/dl; LDH, 150 IU/L; pH 7.44. The AFB smear and culture were negative. Pleural biopsy revealed chronic inflammation without granulomata or giant cells. The AFB stain and culture of the biopsy specimen were negative. Fungal cultures of nail clippings were negative.

A diagnosis of pleural effusion secondary to yellow nail syndrome was made. Over the following eight months, the patient required thoracentesis and removal of 1,500 ml of pleural fluid every four weeks for relief of severe fatigue, dyspnea on exertion and orthopnea. The symptom-free interval between taps was approximately three weeks. Pleural fluid analysis of all subsequent therapeutic taps was similar to the initial values. The AFB cultures and cytologic findings on all specimens were negative. In March 1987, a small right pleural effusion was noted on chest radiograph.

Because of declining efficacy of thoracentesis in relieving her symptoms, obliteration of the pleural space was considered but the patient refused both chemical pleurodesis and open pleural abrasion or pleurectomy. In July 1987, the patient underwent placement of a left pleuroperitoneal shunt (Denver Biomaterial Inc, Denver) with a 1.5 ml pumping chamber. Thoracoscopy performed at that time revealed thickening of the diaphragm, pericardium and posterior parietal pleura consistent with chronic pleural inflammation.

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FIGURE 1. Patient's fingers. Note yellow color, thickening, transverse ridging, onycholysis and absence of lunulae.
time was unremarkable. The patient was instructed to compress the pumping chamber 100 cycles four times per day. Despite good compliance with this regimen, three weeks after placement of the shunt the patient was severely fatigued and dyspeic, and chest radiograph showed reaccumulation of the left pleural effusion. It was assumed that the shunt had become obstructed.

The patient underwent thoracentesis every four weeks until November 1987, when she underwent placement of a second pleuroperitoneal shunt. The patient was again instructed to compress the pumping chamber 100 cycles four times per day. Since the placement of this second shunt the patient has been followed up for one year and has not required thoracentesis, nor has she had any recurrence of her symptoms. Chest radiographs have shown a stable left pleural effusion, decreased in size compared with preoperative films, and a small, stable right pleural effusion (Fig 3).

**Discussion**

Pleural effusions in the yellow nail syndrome are thought to be secondary to dysfunction of pleural lymphatics. Once present, they tend to persist and may recur rapidly after therapeutic thoracentesis, making this an impractical option in many symptomatic patients. Chemical pleurodesis using quinacrine (Atabrine) and tetracycline has been successful in two patients. Nine patients have been treated successfully with open pleural abrasion, pleurectomy or decortication. Unfortunately, chemical pleurodesis causes pain and discomfort and has an appreciable failure rate, and open pleural abrasion and pleurectomy, while very effective, are major surgical procedures associated with a perioperative morbidity of up to 10 percent.

The pleuroperitoneal shunt is a modification of the Denver peritoneovenous shunt and may be inserted under local or general anesthesia in a procedure taking approximately 30 min. After insertion, the patient is required to compress the pumping chamber, which rests on the chest wall, for approximately 5 min four times per day, a frequency which results in fluid transfer of up to 1 L/24 h.

Pleuroperitoneal shunting is an effective mode of palliation of symptoms in patients with malignant pleural effusion. Little et al reported 17 patients with malignant effusion treated with pleuroperitoneal shunting. Twelve achieved effective palliation, with subjective relief of dyspnea in all, and decrease in the volume of pleural effusion on chest x-ray by greater than 50 percent in eight. Only one shunt became obstructed. The five patients who did not achieve palliation were either moribund or unable to compress the pumping chamber effectively. Cimochowski et al reported successful palliative use of pleuroperitoneal shunting in five patients with malignant effusion. The use of the pleuroperitoneal shunt in benign effusion in adults has been described in only two instances: an effusion secondary to mediastinal fibrosis and an effusion associated with amyloidosis. The temporary (less than three months) use of the pleuroperitoneal shunt also has been reported in the management of chylothorax in infants.

Our patient represents the first report of the use of the pleuroperitoneal shunt in the treatment of pleural effusion associated with the yellow nail syndrome. This procedure avoids the need for lengthy hospitalization and is associated with lower morbidity than chemical pleurodesis or open pleural procedures. It may also ameliorate the negative nitrogen balance which may result from repeated large volume thoracentesis in chronically ill patients. Pleuroperitoneal shunting should be considered as an option in the management of any symptomatic patient with chronic pleural effusion, benign or malignant.

**References**

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**Successful Closure of Chronic BPF by Thoracoscopy after Failure of Endoscopic Fibrin Glue Application and Thoracoplasty**

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We report a case of chronic debilitating BPF following right upper lobe resection. Despite several endobronchial applications of fibrin glue, we could not close it. Since the patient was extremely debilitated by symptoms due to the BPF, a thoracoplasty was attempted but was not successful. Finally, the BPF was definitely closed by instillation of talc into the pleura through thoracoscopy. To our knowledge, this is the first reported case of chemical closure of a recalcitrant BPF by the route of thoracoscopy. It also shows the failure of endoscopic fibrin glue application in such a condition.

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**BPF = bronchopleural fistula; AFB = acid-fast bacilli**

Bronchopleural fistula is a rare but difficult complication following pulmonary resection. Adequate and prolonged pleural drainage combined with antibiotics continues to be its basic treatment. When this fails, several surgical approaches have been applied with variable rates of success.3,4 Successful closure of BPF by endobronchial fibrin glue application through a flexible bronchoscope has recently been reported.3,4

**CASE REPORT**

A routine chest x-ray film was performed on a 64-year-old farmer before right hip replacement. A cavitating lesion with an air-fluid level was seen in the right upper lobe. He had received nine months of chemotherapy, two years previously for culture-positive bilateral upper lobe tuberculosis. Lung carcinoma or reactivation of tuberculosis was suspected. Repeated gastric and bronchial aspirates and urine collections showed no AFB on Ziehl-Neelsen staining and cultures. Fiberoptic bronchoscopy and bronchial cytologic studies were normal. Through fluoroscopy, a transthoracic needle aspiration was performed using a Nordenström needle. Cytologic studies, AFB smear and culture of the aspirate were negative.

A two-month trial of INH and rifampicin followed by six weeks of amoxicillin produced no change in the chest x-ray film. A second bronchoscopy was normal.

A right upper lobe resection was performed. Surprisingly, histologic examination of the resected lobe revealed extensive bronchiectasis of the right upper lobe colonized by *Aspergillus fumigatus*. One week later, the patient developed high fever, cough and

![Figure 1. Chest radiograph showing the persistent air-fluid level after right upper lobe resection.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=data/journals/chest/21608/ on 06/27/2017)