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A Clinical Case of Recurrent Pleural Effusions, Culminating in Empyema-thoraces in a Sudanese Woman with Yellow Nail Syndrome

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Abstract: A 50-years-old Sudanese woman was admitted to Elshaab Medical Teaching Hospital, Khartoum, Sudan. She had a history of repeated pleural effusions and a family history of yellow nail syndrome. The patient presented with productive cough, dyspnea and chest pain. On clinical examination, her nails were yellowish-green in color and dystrophic in appearance, thickened and excessively curved. X-ray and CT scan confirmed the presence of fluid in the right side of the chest. On administration of intercostals tube a purulent exudate was collected in the drain from the right side of the chest. The purulent exudate (empyema) was attributed to invasion of the pleural effusion by secondary bacterial organism. Microbiological examination revealed isolation and identification of *Pseudomonas stutzeri* from the infected pleural fluid. As severe dyspnea and chest pain continued, thoracotomy was decided and the patient was referred to thoracic surgery unit. The removal of pus and decortication of the pleura resulted in improved respiratory condition of the patient. The present study describes the first report of a clinical case of recurrent pleural effusions in association with yellow nail syndrome in a Sudanese patient.

Key words: Pleural Effusions, yellow nail, syndrome, chestpain

INTRODUCTION

The yellow nail syndrome is a rare disorder which is characterized by yellow nails, lymphedema and pleural effusion. The etiology of the syndrome is unknown, but impaired lymphatic drainage may play a role in its pathogenesis. The yellow nail syndrome has been reported in association with various conditions such as thyroid disease, hypogamma-globulinemia, nephrotic syndrome, rheumatoid arthritis, obstructive sleep apnea and keratosis obturans in the external ear, carcinoma of the gall bladder, xanthogranulomatous pyelonephritis, onychomycosis and protein-losing enteropathy[1-3]. A case of the yellow nail syndrome associated with diffuse lymphangiectasia, diagnosed by jejunal and duodenal biopsies, involving the whole small bowel, was described by Malek *et al.*[4]. While the etiology of the syndrome is obscure, the pathogenesis seems to involve impaired lymphatic drainage. When this syndrome is suspected on the basis of the examination of the nails, extensive investigations including dermatologic, pneumologic and lymphographic parameters have to be launched to exclude underlying neoplasma or autoimmune disease, frequently associated with this syndrome.

The treatment is controversial and poorly effective. Vitamins A and E, Zn sulfate and itraconazole are drugs the most often advised by dermatologist. However, Spontaneous healing is reported in about 30% of the study. In this study, we present the first report of a clinical case of yellow nail syndrome in a Sudanese patient associated with recurrent pleural effusions, which became invaded by secondary bacteria and subsequently developed in chest empyema.

Case report: A 50-year-old Sudanese woman who had productive cough, dyspnea and chest pain was admitted to our hospital. On examination, her nails were yellowish-green in color and dystrophic in appearance, thickened and excessively curved. Our hospital records showed that the patient's sister had a history of the same syndrome which recovered spontaneously. Thus, the patient had a documented family history of the yellow nail syndrome. She has no features indicative of cardiac problems such as edema of her lower extremities or clinical signs on cardiac examination. Chest radiograph including X-ray and CT scan, confirmed the presence of pleural effusion in the right side of the chest. Biochemical tests including liver function tests and

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liver enzymes analysis were normal. However, the patient had hypoproteinaemia (1.4 g dL^{-1}) and urine analysis showed relatively high number of puss cells. In addition, the patient had a high titer of rheumatoid factor (Rh>8 IU mL^{-1}). Cytopathological examination of pleural biopsy showed thickened fibrous wall with numerous scattered chronic inflammatory cells consisting of infiltrations of lymphocytes and plasma cells but no definite granuloma or malignancy was seen. Pleural fluid demonstrated predominantly pus cells.

DISCUSSION

The yellow nail syndrome is a rare clinical entity, which combines three main features yellowish dystrophic nails, chronic lymphedema and pleural effusion. The yellow nail syndrome consists of yellowish, dystrophic nails, lymphedema and pleural effusion. In the present study, the clinical findings of a 50-years old Sudanese woman, who suffered from this rare syndrome, are described. We believe that the present case is the first documented report of the syndrome in association with infected repeated pleural effusions (empyema) in a Sudanese patient. On thoracotomy, empyema was taken care of by pleurectomy (decortication) and removal of pus. The cause of empyema was attributed to contamination of the pleural fluid by secondary bacterial invasion. Microbiological examination revealed that empyema was a result of invasion of the pleural fluid by gram-negative bacilli identified as *Pseudomonas stutzeri*. In a recent study, we have isolated *P. stutzeri* from an infected ruptured pulmonary hydatid cyst[5]. This bacterial organism was reported to be a natural inhabitant of the respiratory system. However, under certain circumstances such as trauma to the chest wall and lungs, or repeated insertion of the intercostals tubes, opportunistic infections may develop, leading to development of chest empyema. Antibiotic sensitivity test showed that *P. stutzeri* was sensitive to a range of antibiotics with highest sensitivity being against tetracycline. There is no specific treatment for yellow nail syndrome but improvement in the health condition of the patient was evident by combined surgical and medicinal treatment with oral tetracycline.

Pleural effusion secondary to lymphedema may be chronic, symptomatic and refractory to treatment, occasionally requiring invasive and painful procedures such as chemical pleurodesis, pleural abrasion or pleurectomy and pleuroperitoneal shunting to achieve control of the effusion and gain symptomatic relief[6].

The literature review reveals that the yellow nail syndrome may be associated with a variety of pathologies. The mechanism involved in pathogenesis of this syndrome consists of a dysfunction of the lymphatic drainage system, but details remain unknown. Currently, definitive treatment does not exist but topical application of vitamin E and zinc compounds may improve the aspect of involved nails. Treatment with oral zinc supplementation has also been reported by Arroyo and Cohen[7]. Evaluation of ninety seven cases of yellow nail syndrome patients was previously studied[2]. The study showed that the disorder appears to be congenital and may have a genetic component which has yet to be defined Govaert *et al.*[8]. This assumption is also supported by the fact that our patient had a family history of yellow nail syndrome.

The recurrent pleural effusions are most likely to be due to hypoplastic lymphatics. Cytopathology showed dilatation of both visceral and parietal pleural lymphatics and the pleural fluid characteristically contains a high percentage of lymphocytes[9]. Patients often give history of recurrent attacks of bronchitis and may have chronic sinusitis, bronchiectasis and recurrent pneumonia. It is worth mentioning that the literature on this syndrome is scanty in Africa and no information is available about the syndrome in the Sudan. This study demonstrated that yellow nail syndrome is not as rare as it appears from the number of published reports and a greater awareness of its various forms will probably lead to more frequent diagnosis. Since spontaneous recovery may occur, the majority of clinical cases of yellow nail syndrome are likely to be missed in remote areas.

To the best of our knowledge, the presented clinical case described the first report of yellow nail syndrome in a Sudanese patient associated with recurrent pleural effusions. The pleural effusion became invaded by secondary bacteria and subsequently developed in chest empyema. Thus, thoracotomy was decided to remove chest empyema and decortication was performed as a procedure of choice due to remove thickening of the pleura.

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