Yellow nail syndrome associated with sjögren's syndrome

Case Report

Sjögren sendromu ile ilişkili sarı tırnak sendromu

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SUMMARY

Yellow Nail Syndrome (YNS) consists of the triad of deformed yellow nails, lymphedema and pleuraleffusion . The syndrome is common among middle aged individuals. This is an infrequently reported clinical entity with only few cases reported from our country.

Key words: *Yellow nail syndrome, Sjögren's syndrome, lymphedema, pleural effusion.*

OZET

Sarı tırnak sendromu plevral efüzyon, lenfödem ve distrofik sarı tırnaklarla karakterize bir hastalıktır. Hastaların çoğunluğunu orta yaş bayanlar oluşturur. Oldukça nadir görülen bu sendrom ülkemizde birkaç vaka raporunda tanımlanmıştır.

Anahtar Kelimeler: *Sarı tırnak sendromu, Sjögren Sendromu, lenfödem, plevral efüzyon.*

INTRODUCTION

Yellow nail syndrome is a rare disorder of the nail, which is usually accompanied by lymphedema (swelling of parts of the body caused by blockage or damage to the drainage of the lymphatic system). It may also be associated with recurrent pleural effusions (fluid collection in space surrounding the lungs) and less commonly bronchiectasis (chronic, abnormal dilation of the bronchi in the lungs), chronic bronchitis and sinus infections (1-5).

It has been reported that congenital malformations and a secondary dysfunction of lymphatic vessels may be responsible for the syndrome including changes of nail, but its exact mechanism is still not known (6). It was found FOXC2 gene in a family with Meige lymphedema, and also in a family with yellow nail syndrome. The authors observed 4

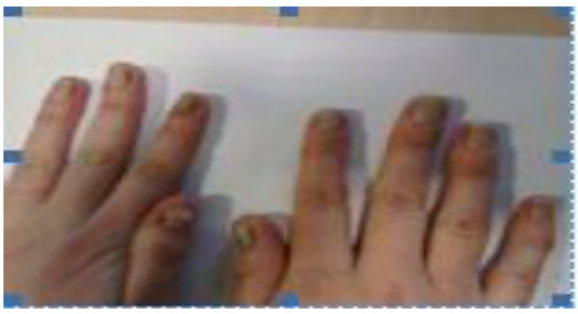
overlapped phenotypically defined lymphedema syndromes: Meige lymphedema, lymphedema-distichiasis syndrome, lymphedema and ptosis, and yellow nail syndrome (7).The syndrome is most often seen in middle-aged individuals. Here we present the first case of a 42 year-old woman with the classic triad of the syndrome associated with Sjögren syndrome.

CASE PRESENTATION

A 42 year- old woman presented with yellow discoloration and thickening of hand and feet nails for two years. Her past medical history was remarkable with recurrent sinusitis and pulmonary infection attacks in addition to these dry eyes and mouth for four years. Due to the recurrent sinusitis, surgery performed a year ago. Physical examination revealed moderate lympheadema on hands and legs. While expirium was prolonged; bilateral, partial, rhoncus were heard over the lung on inspirium.

On dermatologic examination, all of her nails showed a yellow-greenish discoloration with thickening of the nail plates, excessively curving a long both axes and loss of the cuticles (Fig. 1). Dry mouth, eyes and photosensitivity were present on the otorhinolaryngology and ophtalmologic examination. Laboratory studies were normal including complete blood count, CRP, HIV testing, thyroid functional tests, protein electrophoresis, tumor markers, routine biochemistry and urinalysis. However ANA, anti- la and anti – ro positivity detected. Direct microscopic examination and culture of the nail clippings were negative for fungi. The patient was consulted by a pulmonologist. Chest radiograph revealed some patchy non-homogeneous densities in the right paracardiac and perihilar areas and left paracardiac areas. Pulmonary function tests were found to be normal and showed that the obstruction was not developed yet. Lymphedema was observed clinically in our case but lymphocintigraphy was not performed . Furthermore, the patient's abdominal ultrasonography was normal.

Figure 1. *Yellow-greenish discoloration, thickening of the nail plate, and disappearance of the cuticle on both hands in the 42 year-old-women.*



DISCUSSION

Yellow nail syndrome known as "Primary lymphedema associated with yellow nails and pleural effusion" is a very rare medical syndrome thatincludes pleura leffusions, lymphoedema andyellow dystro phic nails. Approximately 40% will also have bronchiectasis. It is also associated with chronic sinusitis and persistent coughing. It usually affects adults (1-4). YNS may be associated with a number of systemic diseases such as rheumatoid arthritis, thyroiditis, acquired immunodeficiency syndrome, tuberculosis, immunologic disorders, malignancies and mycosis fungoides (8). YNS is a rare disease and its exact pathogenesis is still not known. Most of the RA patients with YNS had been treated with the anti-rheumatic agents D-penicillamine and bucillamine. In non-treated patients, spontaneous YNS was very rare. However, pulmonary diseases, edema and other systemic complications were frequently observed in both drug-induced and spontaneous YNS associated with RA. Although the nail changes and systemic complications are probably due to different causes in drug-induced YNS, a careful search for systemic complications are necessary in patients who develop nail changes (9). Since There was no arthritis

component in our case she had not been treated with any anti rheumatic drugs. The disease is believed to start with congenital lymphatic blood vessel hypoplasia and inadequate lymphatic drainage system (6) and it is responsible for all the findings including nails. Lymphedema may begin in the legs at the late ages. Edema may also affect the genitalia, hands, face, and vocal cords (10). The disease is also considered to have a genetic component, although it has not been proved yet (5). There were not similar findings on the other people in the family of our case, and late onset was expressed to be in assosiation with systemic diseases such as sjögren's syndrome.

The mean age at onset is 40 years. However, the age of onset varies widely; for example lymphedema may be present even from the birth until 60's and it rarely affects children. The yellow nails are found in 89%, lympedema in 80% and pleural effusion in 36% of the cases. These three findings are concurrently seen totally only in one, third of patients (11). In 1972 it was stated that existence of two findings are enough to establish the diagnosis (12). Moreover, only typical nail changes are enough for the diagnosis of YNS (13). In most cases, all of the 20 nails are affected. The nails are typically yellowish green in color, very slow growing, thickened and excessively curved a long both axes. The cuticle and lanula are usually absent and onycholysis is frequently evident (1,8,11). Our case was a 42 year-old woman and findings of the nails was said to have been present from the birth onwards.

In our case there was history of having former respiratory infection but have not bronchiectasis. Pleural effusion is expected to occur later. In 1994 rhinosinusitis may be recognised as part of the syndrome and rhynosinusitis frequency has been reported 83% (4). She had similar nail findings, pansinusitis and lymphedema but had not pleural effusion and bronchiectasy.

There is no treatment or cure specifically for the resolution of the condition. Nutritional supplementation with vitamin E appears to be effective in controlling yellow nail syndrome, for unknown reasons. It has been noted that Itraconazole and fluconazole are oral antifungal agents appear to speed up the rate of growth of nails, which may be of benefit in yellow nail syndrome even though it is not caused by fungal infection. The patient was given vitamin E, and hydroxychloracin 200 mg /day po for treatment of the sjögren's syndrome.

CONSENT

Written informed consent was obtained from the patient's mother for publication of this case report and accompanying images.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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