Actas Dermosifiliogr. 2007;98:372-6

LETTERS TO THE EDITORS

Yellow Nail Syndrome

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To the Editor:
We describe a 75-year-old woman with a history of hypertension, hypertensive heart disease, acute pulmonary edema, chronic bronchitis, recurrent pleural effusion, and chronic maxillary sinusitis who was being treated with furosemide, spironolactone, acetylcysteine, omeprazole, budesonide, and salbutamol. She was hospitalized for an increase in dyspnea caused by left pleural effusion and attended by our department for a 15-year history of nail abnormalities that had been treated with oral and topical antifungals and vitamin complexes without improvement. The examination showed that all 20 nails had a dark greenish-yellow discoloration with transverse and longitudinal thickening of the curvature, distal onycholysis, transverse ridges, and absence of cuticle and lunula (see figure). The patient had had lymphedema without dimpling in the lower limbs for more than 20 years, as well as chronic diarrhea of unknown cause for the past 15 years. Based on these findings, a diagnosis of yellow nail syndrome was made. The patient declined treatment for the onychopathy.

Yellow nail syndrome is a chronic disease characterized by the triad of nail abnormalities, respiratory symptoms, and lymphedema. It is usually sporadic and is more common among middle-aged women.[1,2] The diagnosis requires 2 of the 3 criteria, with nail manifestations essential.

The pathogenesis is not clear. Drainage abnormalities observed on lymphography and morphological alterations observed in biopsies suggest a congenital abnormality of the lymphatic vessels. In contrast, the results of lymphoscintigraphy and the clinical reversibility of the syndrome may indicate a functional abnormality.[3,4] Lymphatic involvement explains the lymphedema, pleural effusion, and intestinal lymphangiectasis. Free radicals are released in the nails, delaying growth and keratinization. Additionally, lipofuscin is released and then deposited in the nail layers, hence the name of the syndrome.

Nail abnormalities tend to be the first symptom, with the condition characterized by outbreaks and involvement of all nails to greater or lesser extent. The nail is totally or partially affected, with a variable yellowish tone. Other alterations include distal greenish or gray discoloration, delayed growth, increase in transverse and longitudinal curvature,[5] thickening, transverse ridges, onycholysis, onychoptosis, and pseudo-paronychia.

The differential diagnosis should consider the following: onychomycosis, traumatic pachyonychia, lichen planus, exogenous substances, medications, and systemic diseases.[6]

The condition leads to pulmonary disease, mainly bilateral recurrent pleural effusion as well as bronchiectasis, chronic bronchitis, and sinusitis. It also causes chronic diarrhea or ascites in the digestive system and chronic lymphedema in the lower limbs.

Yellow nail syndrome may be associated with systemic diseases such as rheumatoid arthritis, thyroiditis, dysglobulinemia, Guillain-Barré syndrome, immunodeficiency, and nephrotic syndrome. Moreover, its association with neoplasms may lead to the suspicion of a paraneoplastic syndrome.[7,8]

Treatment of the systemic manifestations and associated diseases improves the nail symptoms.

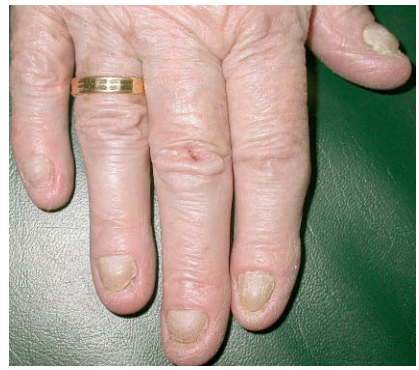

Figure. Yellowish discoloration, nail shortening and thickening, and increased curvature of the nail layers.

Spontaneous remission may occur, and while treatment of the nails is usually ineffective, it should be initiated in the case of pain or for esthetic or functional reasons. Vitamin E achieves the best results at a dose of 1200 IU a day for 3 to 6 months.[9] Other treatments are topical vitamin E, zinc sulfate,[10] systemic antifungals in pulse therapy,[3,11,12] and intralesional triamcinolone acetonide.[13]

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Radiotherapy-Induced Scalp Angiosarcoma

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To the Editor:

We describe a 57-year-old man with multiple basal cell carcinomas on the scalp, where he had undergone radiation therapy in childhood. He consulted for a pearly mass adjacent to a violaceous mass of 5 cm that was poorly defined, indurated, asymptomatic, and with several peripheral nodules (Figure 1), but no local or regional lymph node enlargement. The biopsy showed a basal cell carcinoma and a highly malignant angiosarcoma, with mixed epithelioid and fusiform cell type, high mitotic index, and considerable vascular invasion (Figure 2). Immunohistochemistry showed strong positivity for CD31 (Figure 3), partial for cytokeratins, and low for CD34 and VIII antigen. The computed tomography scans and magnetic resonance imaging revealed infiltration of the subcutaneous cellular tissue and a lesion in the left frontoparietal lobe, the nature of which could not be determined, as the patient declined angiography. Biopsies of the underlying bone and cerebral parenchyma showed no evidence of infiltration. Surgery and local radiation therapy were performed. However, at 2 months, new nodules appeared on the scalp as well as enlarged retroauricular lymph nodes; these were treated by local radiation therapy. Pulmonary metastases have appeared recently, and the patient is undergoing systemic chemotherapy at the time of writing.

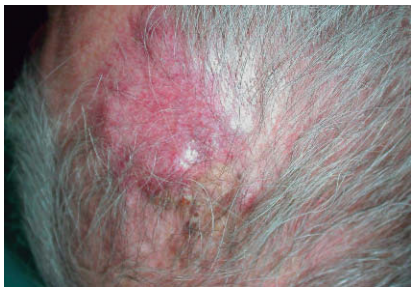

Figure 1. Violaceous, indurated, and poorly delimited lesion, with peripheral nodules, in the left parietal region of the scalp.

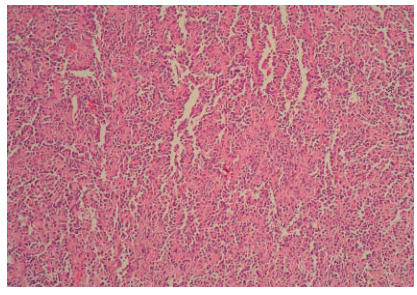

Figure 2. Hematoxylin-eosin stain. Anastomosed, dilated vascular channels with proliferation of atypical endothelial cells.

Angiosarcoma is a rare malignant tumor. A third of these tumors occur in the skin, with a predisposition for superficial soft tissues.[1] The condition is more common in older white men. Lymphedema, chronic radiodermatitis, and immunosuppression are related factors.

The pathogenesis is unknown, although it appears to have a multifocal origin in the lymphatic vessels. As occurs with basal cell carcinomas, gene mutations have been found in the gene for p53, which would induce overexpression of vascular endothelial growth factor.[2,3]

Angiosarcoma can display 4 clinical presentations: idiopathic angiosarcoma of the scalp and face, angiosarcoma associated with chronic lymphedema, angiosarcoma secondary to radiation therapy, and primary angiosarcoma of the breast. Initially, all of these types