

Unusual manifestation of the yellow nail syndrome - Case report*

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Abstract: The yellow nail syndrome is a rare disorder characterized by the classic triad of yellow and dystrophic nails, lymphedema and pleural effusion. We report in this paper a case of yellow nail syndrome, presenting the classic triad of the disease, associated with an unusual lymph accumulation in the abdomen region.

Keywords: Lymphedema; Nail diseases; Yellow nail syndrome

INTRODUCTION

The yellow nail syndrome (YNS) is a rare disorder characterized by the classic triad of yellow and dystrophic nails, lymphedema and pleural effusion, resulting from malformations of the lymphatic system. It can come accompanied by other manifestations of the respiratory tract, such as bronchiectases and rhinosinusitis.^{1,2}

It was primarily described by Samman & White in 1964. In 1972, Hitler *et al* described that the presence of only two of three classical symptoms would be enough to establish the diagnosis, which in practice is much more frequent than the presence of the triad (only 27% of carrying patients).^{3,4,5} Positive familial history is an exception.

CASE REPORT

Male patient, 56 years old, referred bulging in the umbilical regions for 3 days, with low intensity pain. He denied vomiting or changes in bowel habits. He reported having already been diagnosed as a YNS carrier. He underwent bilateral pleurodesis some years ago and two umbilical herniorrhaphies. He was in good general state, eupneic and afebrile. Pulmonary auscultation with sparse snoring noises, bilaterally.

The abdomen was flaccid, with large bulge in the umbilical area, somewhat painful to palpation, with no signs of peritonitis (Figure 1).

He presented swollen lower limbs 2+/4+ with soft edema and trophic skin alterations. The ten toenails were yellowed and dystrophic, hyperkeratotic, with no cuticles or lunula and with increase of curvature. Fingernails presented the same alterations (Figures 2 e 3).

The abdomen CT scan revealed a 0.9 cm hernia ring in the umbilical area, with intestinal content as a component of this hernia (Figures 4 e 5).



FIGURE 1: Marked bulging in the umbilical region

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FIGURE 2: Right foot nails yellowed and dystrophic. Notice the absence of cuticle and lunula



FIGURE 3: Nails yellowed and dystrophic, curved and without lunula and cuticle (right hand)



FIGURE 4: CT scan of abdomen, axial plane. Umbilical hernia with extrusion of small intestine loops to subcutaneous tissue; accumulation of fluid intermingling with handles



FIGURE 5: CT scan of abdomen, sagittal plane. Herniation of small intestine loops to subcutaneous tissue; accumulation of fluid intermingling with handles

The bulge was punctured and there was abundant lymph outflow, which had atypical aspect, not observed in common umbilical hernias, easily diagnosed merely by anamnesis and physical examination.

Some hours later there was a new pain episode, after which a herniorrhaphy was performed, with moderate amount of lymph outflow during the intraoperative period.

DISCUSSION

YNS occurs more frequently in female middle-aged patients, although it can occur from infancy to senescence.

According to study carried out by Nordkild in 1986, with a series of 97 patients who had the syndrome, the yellow nails finding was present in 99% of the cases and was the first symptom in 37% of them.⁶ In the case of the patient above reported, the first manifestation referred was dyspnea, related to the pleuropulmonary effusion and bronchiectases, which prompted him to seek medical assistance years ago and undergo relief pleurodeses.

The 20 nails may be involved (as in the case of the patient in question), and it may be the only sign in 10% of the cases.⁴ Nail changes seem to result from impairment of lymphatic drainage of the fingers and toes. The yellowish color is probably due to lipofuscin pigment, resulting from lipid oxidation of free radicals.⁶ Besides the anomalous pigmentation, diminished nail growth speed (0.1 to 0.25mm/week), onycholysis, hyperkeratosis, disappearance of the lunula and cuticle, and increased curvature are characteristic.

It is important to perform mycological tests and culture for fungi in all patients, since there are reports of cases treated for long periods with oral antifungal drugs without improvement, which were later diagnosed with the syndrome. Several times it is subdiagnosed due to its rarity.

Regarding respiratory involvement, symptoms can start as a recurrent bronchitis, sinusitis, pneumonia and pleural effusions. The pleural liquid is an exudate, rich in proteins and lymphocytes. Fluid restriction and diuretics are not useful and often measures such as pleural shunts, pleurodesis and pleurectomy are indicated. As described above, the patient had

already undergone bilateral pleurodesis and at the time did not present significant respiratory symptoms.

Other serous membranes can be affected by the syndrome, which is even more uncommon. There is one case of pericardial effusion described in Brazilian literature.⁷ Our patient presented accumulation of lymph in the abdominal cavity, rarely found in cases described in literature, arising from intestinal lymphangiectasia.

Regarding the lymphedema, it is reported as the first symptom to appear in 1/3 of the cases, more often on the lower limbs.³ Mechanical measures such as limb elevation and use of elastic stockings are mandatory. The prevention of bacterial and fungal skin infections is equally important. The use of diuretics is little effective.

One of the main differential diagnoses to be remembered is Milroy's disease, a rare dominant

autosomal genetic disease, characterized by edema of the lower limbs with onset right after birth. In rare cases, the lymphedema appears later.⁸ The edema in general is bilateral and symmetric, and can be accompanied by hydrocele, urethral abnormalities in men, papillomatosis and nail dystrophy, only on the toes.⁹

It is also important to remember that YNS can occur in association with other systemic diseases, such as autoimmune disorders and malignancies, as well as in patients with tuberculosis and AIDS.^{4,8,10} There are also secondary cases following use of penicillamine.¹⁰

It is important to investigate in detail the possible concomitance of these diseases. This type of association was not found in our patient. Life expectancy in these patients is slightly shorter when compared to healthy patients. There is no cure for the syndrome, but supportive measures can enhance patient quality of life.⁸ □

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