According to the literature, hospitalization is required in some cases, which did not happen in our case because of the early intervention (Table 2).[5]

Considering a reduced risk of agranulocytosis development and in accordance with Carneiro *et al.* (2011),[5] our aim was not to question DDS therapy for leprosy, but to stimulate clinical awareness of its risks by showing non-specific symptoms of agranulocytosis. We also highlight the need for laboratory test monitoring patients treated with DDS in order to favor the early treatment of this adverse effect, thus enhancing patient prognosis. ❑

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Pseudoxanthoma elasticum-like papillary dermal elastolysis*

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Dear editor,

Elastic fibers are important components of the extracellular matrix of the connective tissue. The elastic system comprises oxytalan and eulanin fibers. Various acquired and hereditary conditions are associated to changes of these fibers, among which we highlight pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE).

PXE-PDE is an acquired condition, that is manifested by multiple asymptomatic, sometimes itchy, non-follicular yellow or normochromic papules, with a diameter of 2-3mm, symmetrically distributed, that can coalesce into "cobblestone" plaques with predilection for the neck area, but also supraclavicular, axillary, flexor aspect of forearms, inframammary and lower abdomen regions. The lesions develop slowly, in months to years, and are clinically similar to pseudoxanthoma elasticum. However, they are differentiated by histopathology and for the lack of systemic involvement.[1]

Histopathology features include loss or marked reduction in the papillary dermis elastic fibers.

Prevalence is believed to be underestimated for this rare condition, what reinforces the importance of better clinical and histological identification by dermatologists and pathologists, to avoid mistaking it for pseudoxanthoma elasticum. [2]

The patient was a 71-year-old woman who presented with

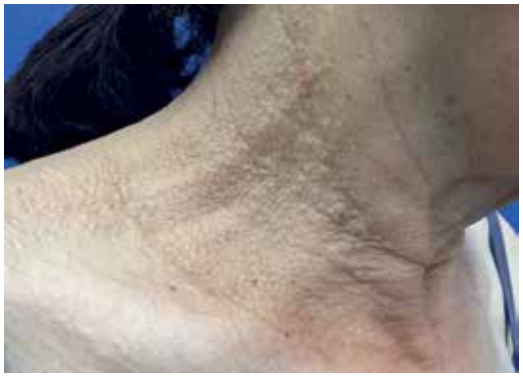

Figure 1: Normochromic non-follicular papules in the cervical region

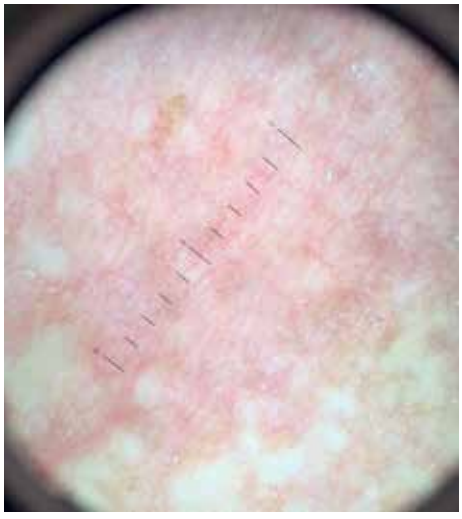

Figure 2: Linear vessels on dermoscopy

multiple asymptomatic non-follicular normochromic papules, symmetrically distributed on the cervical, supraclavicular and antecubital regions, that were progressively enlarging over a 10-year period (Figure 1).

On dermoscopy, there were multiple normochromic papules and linear vessels (Figure 2).

Considering the diagnosis of pseudoxanthoma elasticum, the patient was referred for ophthalmologic evaluation, when no typical angioid streaks were seen. A biopsy was taken from the cervical region, and histopathology with Verhoeff stain revealed loss of elastic fibers in the papillary dermis (Figure 3).

Described in 1992 by Rongioletti and Rebora, PXE-PDE is an acquired, rare disease that affects exclusively elderly women, usually after the fifth decade of life.[1]

Etiology is unknown, but is believed to be due to aging, exposure to ultraviolet radiation and abnormal elastogenesis.[2,3] There is a reported case of familial occurrence of the condition, suggesting a genetic component in the etiopathogenesis.

The diagnosis is confirmed by histopathology with special stains specific for elastic fibers, such as orcein, Verhoeff-Van Gieson or Weigert. The most common change is loss or marked reduction of eulanin and oxytalan elastic fibers in the papillary dermis, and the presence of melanophages. Epidermis can be normal or slightly thinned, with few cases of unspecific perivascular lymphocytic infiltrate in the papillary dermis. No elastic fiber calcification was seen with von Kossa stain, as in pseudoxanthoma elasticum.[2] With immunochemistry, loss of fibrillin 1 and 2 and microfibril-associated glycoprotein 1 and 24 can be seen. Monoclonal anti-amyloid P component antibody can also be used to demonstrate loss of elastic fibers in the papillary dermis. There are no relevant findings on H&E.

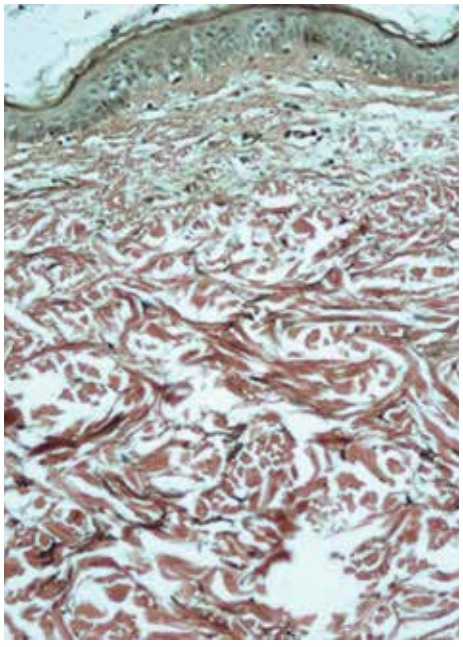

Figure 3: Loss of elastic fibers in the papillary dermis (Verhoeff, X200)

On dermoscopy, besides non-follicular papules, linear and arborizing vessels were described, corresponding to telangiectases seen in the papillary dermis on histology.[4,5]

The differential diagnosis should be made with pseudoxanthoma elasticum, white fibrous papulosis of the neck and mid-dermal elastolysis.[2]

Pseudoxanthoma elasticum is an inherited disorder characterized by calcification and fragmentation of elastic fibers that affects the skin, retina (with the appearance of angioid streaks) and cardiovascular system. Unlike pseudoxanthoma elasticum, PXE-PDE has no systemic manifestations.

The coexistence of PXE-PDE with white fibrous papulosis of the neck is known as fibroelastolytic papulosis.[3] There are no relevant data regarding treatment.

Based on the presented case and the reviewed literature, we highlight the relevance of being familiar with PXE-PDE and of differentiating it from pseudoxanthoma elasticum, to avoid extensive, unnecessary systemic investigations. It is also of importance to look for effective treatments. ❑

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