Pseudoxanthoma elasticum-like papillary dermal elastolysis: A large case series with clinicopathological correlation.

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Source

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Abstract

BACKGROUND:

Pseudoxanthoma elasticum (PXE)-like papillary dermal elastolysis (PDE) is a rare acquired elastic tissue disorder. To date, less than 20 cases have been reported.

OBJECTIVE:

We report a case series of 17 patients presenting with PXE-like PDE and discuss the clinicopathological correlation.

METHODS:

Seventeen cases of PXE-like PDE were collected prospectively and evaluated for common demographic, clinical, and histopathological features.

RESULTS:

All patients were women with a mean age of 61.8 years. The lateral sides and back of neck were the most common sites of involvement (100%), followed by the supraclavicular region (41.2%) and the axilla (35.3%). Systemic involvement was absent in all cases, and in 7 patients the discovery of PXE-like PDE was an incidental finding. The main histopathologic features included complete loss (82.4%) or marked
reduction (17.6%) of elastic fibers in the papillary dermis and the presence of melanophages in the same zone (88.2%).

LIMITATIONS:
Our results require validation with a larger series.

CONCLUSIONS:
Our findings help to differentiate PXE-like PDE from similar elastic tissue disorders based on the selective elastic tissue elimination in the papillary dermis and the presence of melanophages in the same zone as a possible consequence of subclinical junctional photodamage. PXE-like PDE is likely underdiagnosed rather than rare, and dermatologists should be aware of its similarity to inherited PXE to spare unnecessary investigations because of the lack of systemic involvement. Clinicopathologic correlation is critical as hematoxylin-eosin staining is nonspecific and elastic tissue stains are necessary to make the correct diagnosis.