SKIN MANIFESTATIONS OF INTERNAL DISEASE

RARE, CONCURRENT PRESENTATION OF SCLEROMYXEDEMA AND ERYTHEMA ELEVATUM DIUTINUM ASSOCIATED WITH PARAPROTEINEMIA

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Background: Scleromyxedema is a rare cutaneous mucinosis characterized by generalized popular/sclerodermoid skin findings associated with underlying monoclonal gammopathy. Erythema elevatum diutinum (EED) is a rare, chronic form of leukocytoclastic vasculitis (LCV) also associated with paraproteinemia. This is the first reported case of EED and scleromyxedema manifesting in the same patient.

Observation: Patient is a 72-year-old man with a one-year history of pruritic eruption that started on his face then spread to his body/arms. He complained of associated joint pain and swelling in his hands and elbows. Upon presentation, patient had confluent firm, skin-colored papules on his face, neck, trunk, and bilateral upper extremities with positive Sharpei-sign. His elbows, MCP and PIP joints were edematous and tender to palpation. A skin biopsy showed dermal fibroblasts in an edematous stroma with abundant mucin on Alcian blue. Pathology finding was consistent with scleromyxedema. He underwent extensive work-up and was found to have IgG-Kappa monoclonal gammopathy with elevated kappa-lambda ratio of 1.38 on SPEP. His CRP was elevated (58mg/L) and his other laboratory results were unremarkable. The patient was started on IVIG infusions (2mg/kg) at monthly intervals. His skin disease and joint pain improved drastically. Four-months into IVIG-treatments, he developed pink, firm plaques on bilateral elbows and dorsal feet. Biopsy from his left elbow lesion showed LCV consistent with EED. He was promptly started on hydroxychloroquine 200mg twice daily and his new skin lesions improved. He will undergo a bone-marrow biopsy to evaluate for bone-marrow dyscrasia.

Key-message: Scleromyxedema is diagnosed based on characteristic clinical and histological findings. Disease course is progressive and may lead to death. IVIG is considered an effective first-line treatment. The current patient responded drastically to IVIG, but then developed typical EED lesions that responded well to hydroxychloroquine. Presentation of both conditions in the same patient is exceedingly rare.