Cutis laxa (CL) syndromes are a group of disorders characterized by abnormal elastic fibers resulting in the clinical appearance of redundant, loose, inelastic skin. Congenital CL results from errors in elastic fiber assembly, often with skin manifestations present from birth, while acquired CL often results from elastic fiber degradation. In this case, we describe a case of cutis laxa in a 47-year-old female patient, specifically acquired cutis laxa (ACL) given the late onset and lack of family history.

CASE REPORT
A 47-year-old female patient presented to a private practice clinic for a cosmetic consult. Her chief complaint was increased laxity of the skin on the abdomen, middle back, bilateral axillae, and bilateral medial thigh that had been present for four months. The patient was otherwise asymptomatic, and past medical history and family history were unremarkable. Aside from vitamin B12 supplementation, the patient denied medication use.

On physical exam, erythematous patches with dermal atrophy were noted on the chest, abdomen, and back (Images 1-3). Redundant and wrinkled skin was noted on the abdomen, bilateral axillae, lower back, and bilateral inner thighs (Images 4-6).
Abnormal laboratory results a month after initial visit included a low Vitamin D level of 23 and urinalysis and complete blood count (CBC) supportive of a urinary tract infection diagnosis. B-type natriuretic peptide (BNP), thyroid-stimulating hormone (TSH)/thyroxine (T4), uric acid, Anti-nuclear antibodies (ANA) screen, C-reactive protein (CRP), rheumatoid factor, and erythrocyte sedimentation rate (ESR) were unremarkable. The patient was referred to an academic facility where further workup, including protein electrophoresis, lactic dehydrogenase (LD), Rapid Plasma Reagin (RPR), Lyme Ab, and gamma-glutamyl transferase (GGT), T-spot for tuberculosis, HSV type 1 and 2, transglutaminase IgA antibody, celiac disease antibodies, repeat CBC, prolactin, and ultrasound of abdomen were found unremarkable.

Two 4mm punch biopsies from the right upper thigh (A) and right lower abdomen (B) were performed. On histology, both biopsies showed similar features with interstitial giant cells located in the mid to deep dermis exhibiting elastophagocytosis. A Verhoeff-Van Gieson (VVG) special stain for elastic fibers showed significant loss of elastic fibers from the papillary dermis to the mid-to-deep dermis. The areas of multinucleate giant cells highlighted the areas of elastophagocytosis and some areas of short, thick, and fragmented forms (Images 9-13).

A third biopsy performed two months later revealed increased density of dermal mast cells with retention of elastic fibers. Due to the presence of increased mast cells, the patient underwent a workup to exclude underlying systemic mastocytosis. Pertinent laboratory findings of this workup included an elevated tryptase level of 41.9. Bone marrow exam, chromosome analysis, flow cytometry, and KIT (D816 V) mutation found no evidence of systemic disease.

Since initial diagnosis a year ago, the patient’s skin has continued to increase in laxity and surface area involvement, with corresponding elevation in tryptase level to 43 (Image 7, 8). The disease remains cutaneous as of now, but systemic involvement will be continually monitored. In terms of treatment, the patient has opted to schedule future surgery as a means of improving everyday functionality.

**DISCUSSION**

ACL is a rare disorder that typically occurs in adulthood and may be associated with various conditions and drugs. Some known triggers include medications, toxins, infections, cutaneous mastocytosis, and hematologic neoplasms. The exact etiology of ACL remains unknown. Low lysyl oxidase activity, high cathepsin G levels, and reduced alpha-1-antitrypsin are noted in some patients, likely contributing to decreased cutaneous elastin. Diagnosis is primarily clinical, but all subtypes of CL exhibit fragmented, reduced, and disordered elastic fibers, especially in the papillary dermis, on light microscopy.

![Figure 7. Anterior image depicting clinical progression (2021).](image)

![Figure 8. Posterior image depicting clinical progression (2021).](image)
Furthermore, a pertinent laboratory finding amongst an extensive workup at the time of diagnosis was an elevated tryptase level of 41.9, which has continued to elevate further in correlation with worsening clinical condition. We speculate that elevated tryptase levels are linked to the pathogenesis of our patients’ condition and conceivably a primary cause of her elastin breakdown. Tryptase is a neutral protease found in the secretory granules of human mast cells and has been found in vitro to be a marker of mast cell degranulation. In previous mechanistic studies that reviewed tryptase expression in relation to abdominal aortic aneurysm formation, it was concluded that tryptase contributed to elastin degradation. Elevated serum tryptase level is a minor diagnostic criterion for mastocytosis, a rare disease characterized by excessive production of mast cells that accumulate in the skin, bone marrow, and other visceral organs. The disorder is often due to a somatic mutation in the KIT gene and the preferred method of diagnosis is a bone marrow biopsy. Although both bone marrow biopsy and genetic testing did not reveal definitive evidence of systemic mastocytosis in our patient, it has been reported as a concurrent condition in patients with ACL and will continue to be a consideration. Further studies of ACL are necessary to determine targeted options to treat, or at least halt the progression, of this disease.

The authors have no conflicts of interest to report.

Helena Drolshagen, BS, is student University of Arkansas for Medical Sciences in Little Rock, AR.

Sara Shalin, MD, PhD, is chair of the department of dermatology and an associate professor in the departments of pathology and dermatology at University of Arkansas for Medical Sciences in Little Rock, AR.

Sandy Marchese Johnson, MD, FAAD is a dermatologist at Johnson Dermatology in Fort Smith, AR.