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Cutaneous involvement of light chain deposition disease: A case report

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Abstract

Light chain deposition disease is a rare condition associated with plasma cell dyscrasia and other lymphoproliferative disorders in which there is overproduction and deposition of non-amyloid light chains in various organs, leading to organ dysfunction. It is well-established that the majority of patients with light chain deposition disease exhibit renal involvement. Although awareness of extrarenal manifestations is increasing, cutaneous involvement has rarely been reported. Herein, we present a case of light chain deposition disease with cutaneous manifestations in the absence of any renal disease. A biopsy of the skin revealed amorphous eosinophilic material within the superficial dermis. Using special stains, immunohistochemistry, and direct immunofluorescence, the deposits were confirmed to be kappa light chains.

Keywords

Light chain deposition disease, LCDD, monoclonal immunoglobulin deposition disease, non-amyloid deposition disease

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Introduction

Light chain deposition disease (LCDD) is a rare condition characterized by the deposition of non-amyloid monoclonal immunoglobulins in various organs, often resulting in impaired organ function. Most notably, renal dysfunction, including renal failure and nephrotic syndrome, has been observed in up to 96% of cases. Though less common, extrarenal deposits can clinically affect other organs including the heart, liver, and peripheral nervous system.[1] Cutaneous manifestations are rare and poorly characterized, as only a select few cases have been presented in the literature previously.[2–4] Even more uncommon are cases, such as the current one, of LCDD with biopsy-proven cutaneous involvement in the absence of renal disease in the setting of a monoclonal gammopathy.[2]

Case

A 59-year-old man presented with a 5-week history of progressive diffuse myalgias, arthralgias, weakness, and polyneuropathy. His medical history is extensive, but most notable for IgG kappa monoclonal gammopathy of unknown significance (MGUS), axonal polyneuropathy, treatment-refractory rheumatologic disease of unclear etiology, and rheumatic heart disease. His extended stay in the hospital was marked with extensive workup for his presentation, surrounded by an interdisciplinary team of specialists which included, but was not limited to, dermatology, hematology, rheumatology, and neurology.

Initial dermatologic evaluation revealed multiple lesions including bilateral xanthelasmas, lipomas on the dorsal forearms, and a recent hemorrhagic blister on the left fourth digit. Additionally, there were several long-standing asymptomatic violaceous hyperpigmented, well-circumscribed atrophic patches on the elbows bilaterally. Similar lesions were noted on the chest and back. Punch biopsies were taken from the lesions on his right elbow and upper back. The biopsy taken from the latter demonstrated neutrophilic dermatosis, however non-specific for the disease process. Furthermore, the lesion in this area spontaneously resolved a few days post-biopsy. The biopsy taken from the right elbow was more informative, revealing extensive deposition of amorphous, eosinophilic material within the superficial dermis as well as

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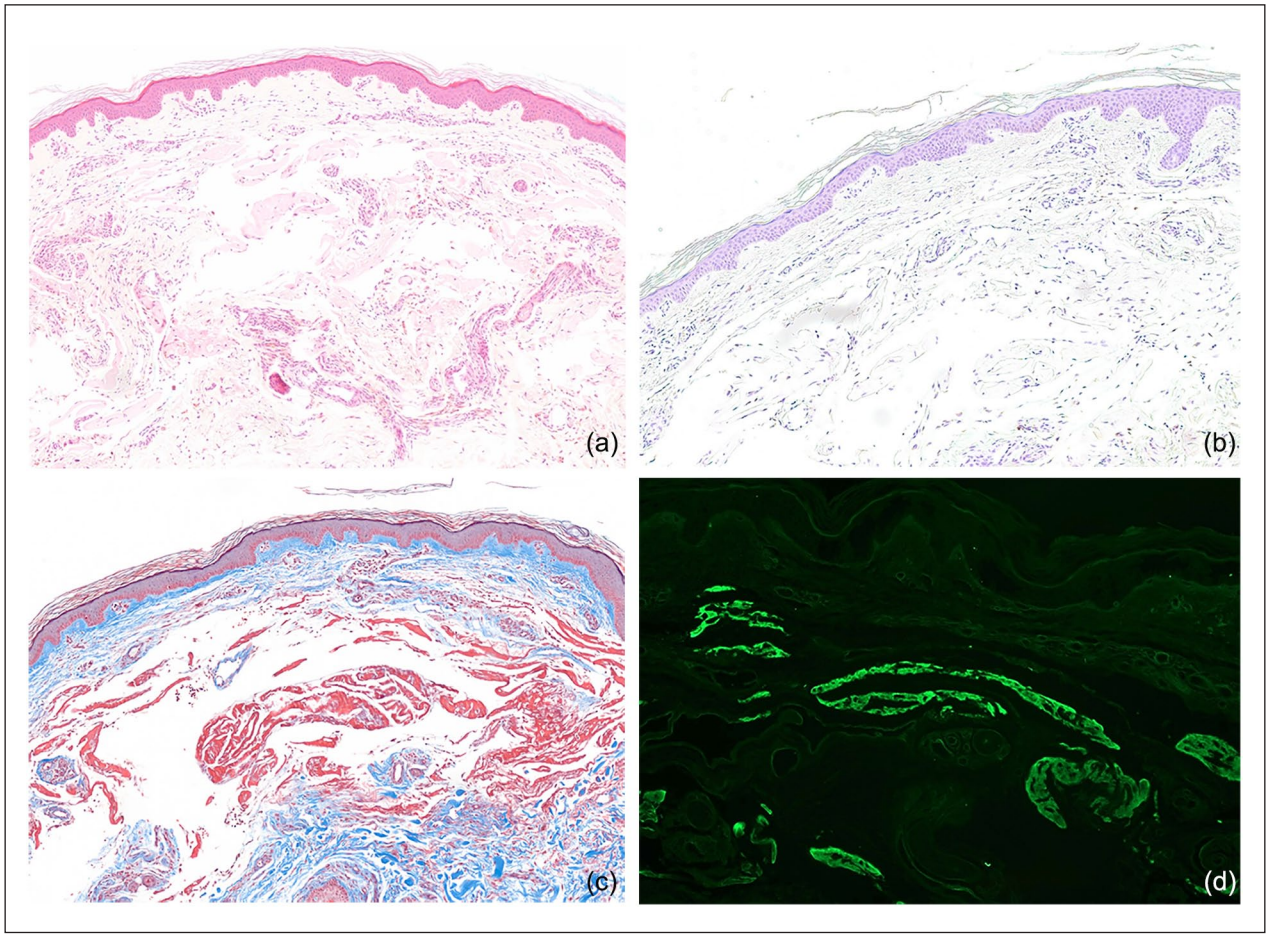

Figure 1. Light chain deposition disease in skin. (a) Extensive deposition of amorphous eosinophilic material in the dermis (hematoxylin and eosin ×200). (b) Deposits are Congo-red negative (Congo-red stain ×200). (c) Deposits display a red reaction (Masson's trichrome stain ×200). (d) Dermal deposition of kappa light chains (direct immunofluorescence ×400).

reactive vascular proliferation at the level of the superficial vascular plexus with abundant dermal hemosiderin deposition, suggestive of occlusive vasculopathy (Figure 1(a)). Using special stains, the deposits were negative for Congo-red and displayed a red reaction with Masson-trichrome (Figure 1(b) and (c)). Kappa and lambda immunostains were difficult to interpret due to increased background staining. Overall, the histochemical findings were consistent with LCDD. Moreover, direct immunofluorescence was performed on the formalin-fixed paraffin-embedded tissue and confirmed a strong fluorescence within the papillary dermis for kappa light chains (Figure 1(d)).

Subsequently, the patient had a bone marrow aspirate and biopsy which revealed kappa-restricted plasma cells (<10%) consistent with a plasma cell neoplasm, without evidence of amyloid deposition. Other notable investigations included a cerebrospinal fluid analysis, which was negative for malignancy, but showed an elevated IgG synthesis rate. Given the patient's polyneuropathy, a sural nerve biopsy was performed, showing chronic axonopathy without evidence of light chain or amyloid deposition. A final periorbital punch biopsy of the under eye was taken which demonstrated benign xanthelasma. A cardiac MRI was also performed to rule out infiltrative disease. Further diagnostic investigations precluded any abnormalities with any other organs associated with LCDD.

Given the patient's skin lesions were asymptomatic, no further management was pursued. However, a series of treatments were initiated with the goal to abate his polyneuropathy and underlying IgG kappa MGUS, though none were particularly effective. Ultimately, the patient's hospital stay was complicated by progressive polyneuropathy and fluctuating weakness, dysphagia, and trivalvular heart failure. He was eventually admitted to ICU with mixed hypoxemic respiratory failure and septic shock presumed secondary to aspiration pneumonia. Unfortunately, he had persistent and worsening shock despite full medical therapy and died approximately 4 months post-admission.

Discussion

LCDD is characterized by the overproduction of monoclonal light chains and subsequent deposition in various organs. Under

light microscopy, these deposits appear as amorphous eosinophilic material that stain negative for Congo-red. Electron microscopy demonstrates a granular pattern and an absence of fibrillar structures. In addition, immunofluorescent studies reveal kappa restriction in 80% of cases.[1] LCDD is strongly associated with plasma cell dyscrasias and less commonly other lymphoproliferative disorders. The median age of diagnosis is 58 years and approximately 60% of cases are seen in men.[1,5]

Deposition of light chains is primarily known to cause organ dysfunction in the kidneys, but recognition of extrarenal involvement is gradually improving.[5] Recently, Joly et al. conducted a large nationwide cohort study from the French National Reference Center database which highlights the extrarenal manifestations seen in LCDD. Of the 212 patients with this condition, only 8 (i.e., 3.8%) had associated skin findings.[4] Prior to this study, Hendricks et al. reviewed six cases of LCDD with cutaneous involvement, only one of which was associated with a monoclonal gammopathy without evidence of renal disease. This case seems to correspond to our patient's case, except for the difference in the light chain restriction pattern.[2]

While ongoing research is valuable for determining the true prevalence of cutaneous manifestations in LCDD based on histopathological diagnosis, there is no consistent dermatologic clinical picture. For instance, while Hendricks et al. corresponds upper dermis light chain involvement with clinical findings of erythematous lesions and/or purpura, in our case the involvement of the upper dermis yielded a similar, yet distinct set of findings, including violaceous hyperpigmented, well-circumscribed atrophic patches.[2]

Given the absence of an established, cohesive clinical profile, LCDD is often misdiagnosed or underreported. Clinicians should be highly vigilant for the disease when encountering new or persistent unexplained cutaneous lesions, particularly in patients with a known monoclonal gammopathy or complex medical history. As has been well documented, there are a vast number of monoclonal gammopathies with associated cutaneous findings, such as multiple myeloma, Schnitzler Syndrome, and POEMS syndrome, amyloid-light chain amyloidosis, scleromyxedema, and Waldenström macroglobulinemia.[6] In our case, the patient was extensively investigated for each of these conditions in hopes of more targeted management; however, he did not meet the criteria for any of the above. For LCDD, a larger case series may be informative in elucidating its clinical features that correspond to its histopathological pattern of involvement, potentially aiding in earlier detection and guiding treatment.

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Author contributions

S.V.-N.: data curation, visualization, writing (original draft preparation). H.M.: investigation, resources, supervision, writing (review and editing).

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Consent to participate

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References

1. Leung N, Landgren O, Rakumar SV, et al. Haematolymphoid tumors. In: *WHO classification of tumours*. 5th ed., Vol. 11. Lyon: International Agency for Research on Cancer, 2024.
2. Hendricks C, Fernández Figueras MT, Liersch J, et al. Cutaneous light chain deposition disease: a report of 2 cases and review of the literature. *Am J Dermatopathol* 2018; 40(5): 337–341.
3. Topalidis C, Boulogeorgou K, Lazaridis A, et al. Light chain deposition disease: spotlight on a rare cutaneous disease. *Eur J Dermatol* 2023; 33(3): 297–299.
4. Joly F, Cohen C, Javaugue V, et al. Randall-type monoclonal immunoglobulin deposition disease: novel insights from a nationwide cohort study. *Blood* 2019; 133(6): 576–587.
5. Popov H, Stoyanov GS and Ghenev P. Light chain deposition disease: a morphological case report. *Cureus* 2022; 14(6): e26357.
6. Claveau J-S, Wetter DA and Kumar S. Cutaneous manifestations of monoclonal gammopathy. *Blood Cancer J* 2022; 12(4): 58.