

Lichen Amyloidosis

Amyloidosis refers to a group of disorders characterized by the deposition of abnormal proteins, forming insoluble fibrils that accumulate in various tissues, disrupting normal function.

Amyloidoses are categorized into three major clinical subtypes: primary systemic amyloidosis, secondary systemic amyloidosis, and organ-limited types. Both systemic and localized forms of amyloidosis become more prevalent with advancing age, and the presentation of the disease before the age of 30 is rare. These disorders are caused by the accumulation of amyloid fibrils composed of at least 26 distinct proteins, all of which are associated with amyloid deposits in tissues.

Lichen Amyloidosis

Lichen amyloidosis is a cutaneous variant of amyloidosis and accounts for approximately 10% of cases of skin-based amyloidosis. It is distinct in that it does not typically involve systemic amyloidosis, although it may occasionally be associated with other systemic conditions, such as primary biliary cirrhosis, systemic lupus erythematosus, and Sjögren's syndrome. The cause of lichen amyloidosis is often idiopathic, but chronic friction or mechanical trauma to the skin is thought to play a role in its development, potentially leading to the formation of amyloid deposits at sites of repeated injury.

Lichen amyloidosis is particularly prevalent among Chinese individuals and typically affects adults. The hallmark clinical feature of lichen amyloidosis is intensely pruritic (itchy) hyperkeratotic papules that can coalesce to form gray to brown plaques. These lesions are most commonly found on the anterior tibiae, but they can also appear on the extensor surfaces of the upper extremities and trunk.

Diagnosis and Workup

The diagnosis of amyloidosis generally involves clinical evaluation supported by laboratory testing. For all patients suspected of having amyloidosis, a comprehensive workup should include kidney and liver function tests, blood counts, creatinine clearance, and measurement of protein levels in the urine. These tests help assess the extent of systemic involvement in cases of primary systemic or secondary systemic amyloidosis. In cases of systemic amyloidosis, additional diagnostic procedures, including bone marrow evaluation, may be necessary to confirm the diagnosis and determine the subtype of amyloidosis present.

Clinical Course and Complications

The primary complications of lichen amyloidosis are related to pruritus, which can lead to skin damage due to excessive scratching. Bleeding from scratched lesions is also common, as is the potential for secondary infection. Although lichen amyloidosis is primarily confined to the skin, it can occasionally result in painful lesions. However, skin-limited lichen amyloidosis typically does not affect overall mortality.

Treatment Options

The management of lichen amyloidosis is focused on relieving pruritus and improving the appearance of the affected skin. Treatment options include:

- **Sedating antihistamines:** These medications do not directly alleviate pruritus via their antihistaminic effects but are effective due to their sedative properties, which may help break the itch-scratch cycle.
- **Topical treatments:** Intralesional steroids are commonly used, either alone or in combination with topical corticosteroids or topical tacrolimus. These treatments help to reduce inflammation and manage itching at the site of amyloid deposition.
- **UVB phototherapy:** This treatment has shown promise in alleviating pruritus and improving skin appearance in some patients with lichen amyloidosis.
- **Systemic acitretin:** Acitretin, a systemic retinoid, can be effective in reducing the severity of skin lesions. However, treatment must be continued to maintain the effects, as the condition may relapse upon discontinuation of therapy.
- **Surgical interventions:** In cases of isolated or particularly troublesome lesions, surgical excision or dermabrasion may be performed to remove the affected tissue and alleviate symptoms.

Conclusion

Lichen amyloidosis, a cutaneous form of amyloidosis, is most commonly observed in adults and is characterized by intensely pruritic, hyperkeratotic papules that may form plaques, often in response to chronic friction or trauma. Although systemic involvement is rare, associated systemic conditions may be present. Management primarily focuses on relieving symptoms, particularly pruritus, through topical treatments, systemic therapies, and sometimes surgical intervention. Early recognition and treatment can significantly improve quality of life, though further research into long-term management and the underlying pathophysiology of lichen amyloidosis is necessary.

References

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