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Management of Painful Segmental Cutaneous Leiomyoma in HLRCC Syndrome: Efficacy of Topical Calcium-Channel Blockers Validated by Cold-Provocation Test

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Introduction

Cutaneous leiomyomas are benign smooth-muscle tumors that frequently cause paroxysmal, difficult-to-manage pain triggered by cold, pressure, or emotion. The pain mechanism involves calcium-dependent smooth-muscle hypercontractility, providing a rationale for the use of calcium-channel blockers (CCBs). However, systemic CCB therapy is often limited by adverse effects such as hypotension and dizziness. We present a case of segmental cutaneous leiomyoma associated with Hereditary Leiomyomatosis and Renal Cell Cancer (HLRCC) syndrome, successfully managed with a novel approach using topical diltiazem. This report highlights the diagnostic importance of segmental presentation and validates the therapeutic efficacy through a controlled cold-provocation test.

Materials and Methods

A 61-year-old man presented with a 38-year history of multiple painful papulonodules in a segmental distribution on the left abdomen (Figure 1). The pain was severe (Visual Analogue Scale [VAS] 9/10) and impaired daily functioning. Diagnosis was confirmed via histopathological examination (Figure 2). Given the segmental pattern, genetic testing for the fumarate hydratase (FH) gene was performed. To objectively assess the efficacy of topical treatment, a standardized cold-provocation test was implemented. This involved controlled ice application with time-series pain recording (VAS) to measure onset, peak intensity, and recovery time before and after the initiation of topical therapy.



Figure 1. Clinical presentation showing multiple smooth, firm, pink dermal papules and nodules coalescing into plaque- like clusters in a segmental (zosteriform/blaschkoid) distribution on the left anterolateral abdomen, approximating T8–T10.

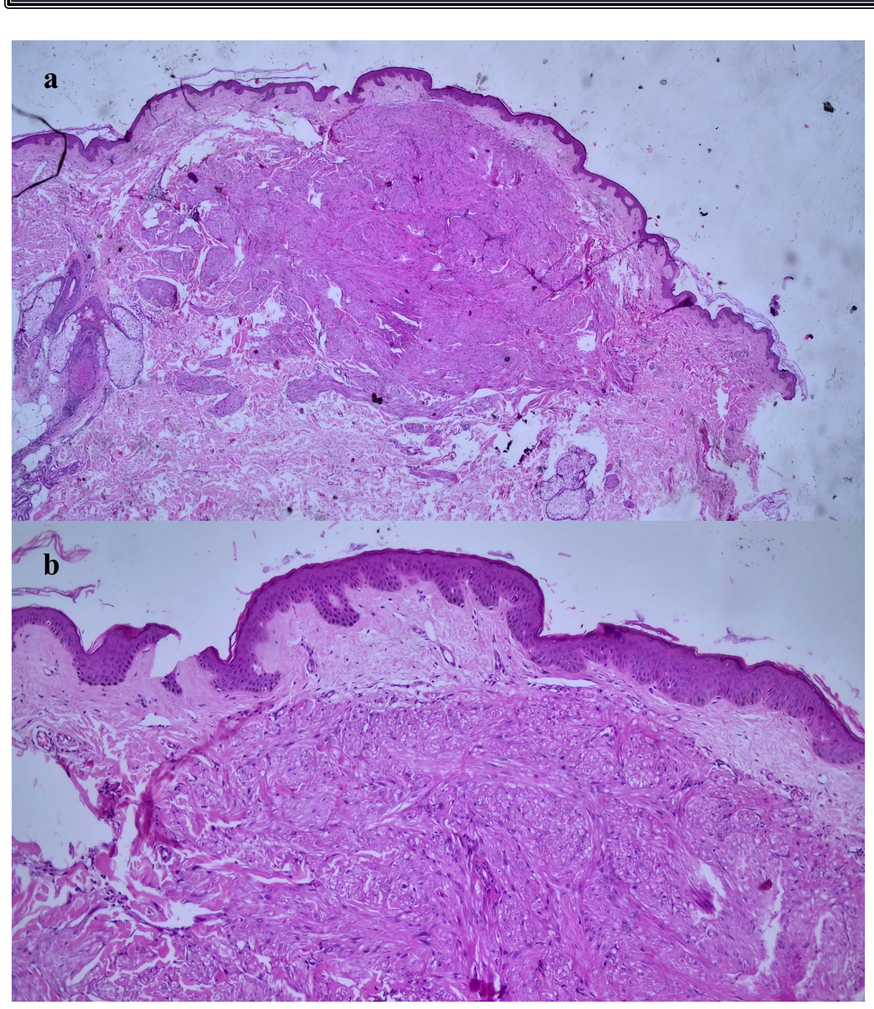


Figure 2. (a) Low- power histopathologic view showing myoid fascicular spindle cell arrangement forming a nodular structure (H&E stain, ×4). (b) High- power view demonstrating leiomyoma bundles located in the papillary dermis, composed of intersecting smooth muscle cells with characteristic oval, blunt- ended nuclei and eosinophilic cytoplasm, without atypia or mitosis (H&E stain, ×20).

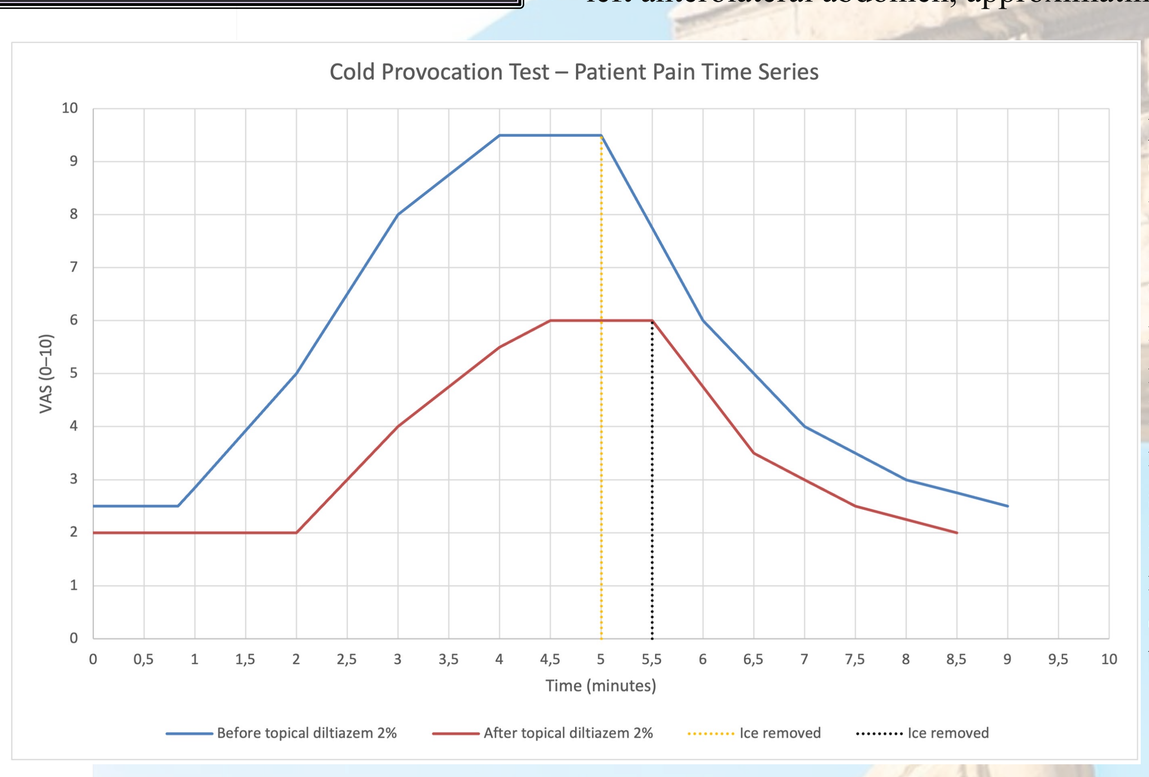


Figure 3. Cold provocation pain curves before and after topical diltiazem 2%: Ice was applied to a fixed area until a 1-min plateau, then removed (yellow/black lines). VAS (0–10) recorded every 30 s shows later onset, lower peak, and faster recovery after diltiazem (red) vs. before (blue).

Results

Genetic analysis identified a pathogenic heterozygous splice-site variant (c.1391-2A>G) in the FH gene, confirming the diagnosis of HLRCC. Initial treatment with oral amlodipine (5 mg/day) reduced resting pain to VAS 5/10 but dose escalation was prevented by dizziness. The addition of topical diltiazem 2% cream twice daily resulted in a further reduction of resting pain to VAS 2/10 and significant improvement in Brief Pain Inventory (BPI) scores. The cold-provocation test objectively demonstrated that topical diltiazem delayed pain onset, lowered peak pain intensity (from VAS 9.5 to 6.0), and accelerated recovery time compared to baseline (Figure 3). No local irritation or systemic adverse effects were observed during the follow-up.

Conclusion

This case highlights two critical clinical points. First, segmental cutaneous leiomyomas should prompt genetic evaluation for HLRCC, even in the absence of family history, to enable early renal cancer surveillance. Second, topical diltiazem serves as a mechanistically rational, non-invasive, and well-tolerated treatment for painful leiomyomas in HLRCC patients, particularly when systemic options are limited by adverse effects. Furthermore, the use of a standardized cold-provocation test provided objective evidence of its analgesic efficacy, offering a valuable assessment tool for future studies.

Encore declaration: This ePoster is declared as an encore presentation because the related manuscript was published after submission to EADV and before the EADV embargo lifting date.