

## Rare Otologic Manifestation of Darier's Disease: A Case Report

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### ABSTRACT

This article presents a rare case of pinnae involvement in a 73-year-old female with Darier's disease, an autosomal dominant genodermatosis characterized by keratotic papules in a seborrheic distribution. The patient, diagnosed at age 16, presented with bilateral aural fullness and hypoacusis, alongside recurrent external ear crusting and debris accumulation, necessitating ongoing management with aural cleaning and topical treatments. Audiometric evaluation confirmed normal hearing thresholds. This case highlights the otologic manifestations of Darier's disease, emphasizing the need for interdisciplinary care between dermatology and otorhinolaryngology to manage chronic symptoms and maintain patients' quality of life.

**Keywords:** Darier's disease; Pinnae involvement; External otitis; Keratosis follicularis

### CASE REPORT

This article presents a rare case of affection in the pinnae of a 73-year-old female patient with Darier's disease. She was referred to our otorhinolaryngology outpatient clinic due to bilateral aural fullness and hypoacusis. The patient was initially diagnosed with this rare condition when she was 16 years old and has been under our care, as well as that of the dermatology outpatient clinic, for more than 5 years. During this period she has experienced frequent external ear crusting and the accumulation of debris, necessitating regular aural cleaning and the application of antibiotic ear drops due to recurrent external otitis. Additionally, she has been utilizing alternate-day acitretin along with antibiotic and corticosteroid cream during exacerbations to alleviate her clinical condition.

Images of the right ear (Figure 1) and left ear (Figure 2) are provided, illustrating diffuse dryness and crusting affecting both pinnae.



**Figure 1:** Right ear



**Figure 2:** Left ear

Darier's disease, alternatively referred to as keratosis follicularis or dyskeratosis follicularis, is an uncommon autosomal dominant genodermatosis. Typically, the disease manifests during puberty and progresses as a chronic condition, with flare-ups often induced by factors such as sun exposure, friction and heat. It is distinguished by the persistent eruption of red-brown, keratotic papules distributed in a seborrheic pattern, among other associated manifestations.

A right (red) and left ear (blue) audiogram was performed (Figure 3), verifying the presence of normal hearing with a Speech Reception Threshold of 15dB bilaterally.

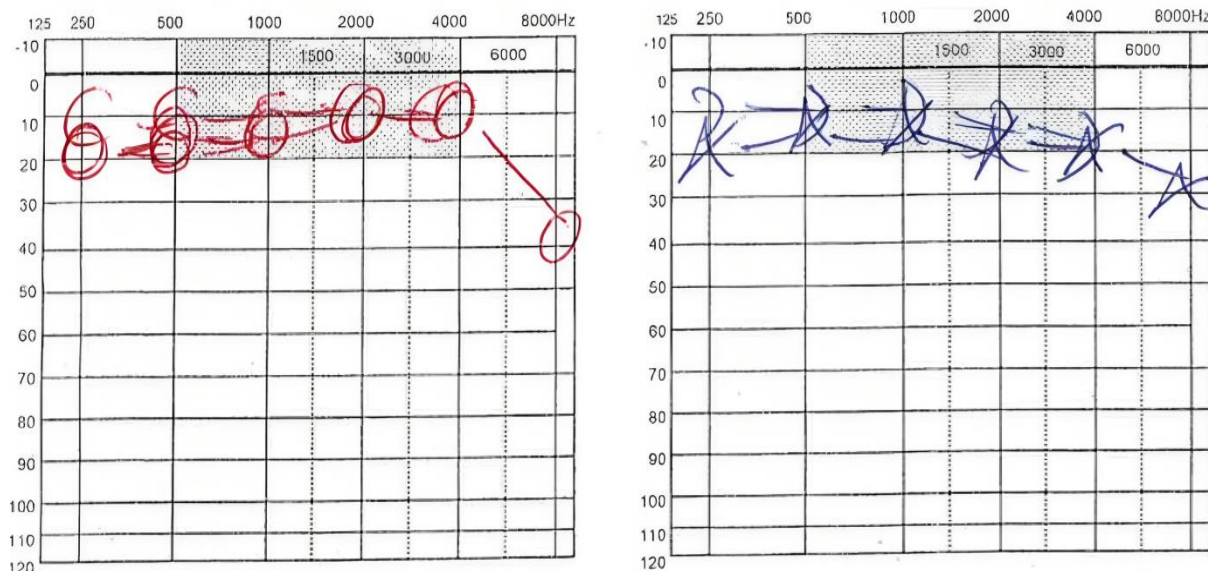


Figure 3: A right (red) and left ear (blue) audiogram

While this condition does not typically result in permanent hearing impairment, it significantly affects the quality of life for patients and requires ongoing outpatient and emergency unit management due to frequent exacerbations.

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