Darier´s disease as an unusual risk factor for otitis externa: A case report

Original Article

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ABSTRACT

Background: Darier´s disease is known for affecting seborrheic areas of the body, including the ear, but no previous case reports describe recurrent otitis externa as a manifestation of this rare disease.

Case presentation: We describe the case of a 70-year-old woman presenting to the Otolaryngology department with bilateral otalgia and pruritus with a 4-day evolution. She complained of recurrent similar episodes for many years requiring treatment. Scaly, pruritic, greasy lesions of the pinna were present and bilateral otitis externa was diagnosed. Inframammary and lower leg involuting lesions were also present. She had been diagnosed with Darier´s disease by the age of 16 years reporting frequent flares. Targeted treatment resolved the otologic complaints after 1 week and Dermatology evaluation was advised to pursue systemic medical optimization.

Conclusion: Patients showing chronic otitis externa along with concurrent dermatologic lesions should be referred to the Dermatology clinic so that diagnostic investigation and targeted therapies may be offered.

Key Words: Darier´s disease, Recurrent otitis externa, skin condition, Ear, Otology, Dermatology.

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BACKGROUND

The ear is a reported site of affection in Darier´s disease. The condition may elicit recurrent otologic complaints such as chronic pruritus or otalgia and otorrhea when concurrent otitis externa occurs, compelling patients to attend an Otolaryngologist. Awareness for this specific diagnosis may be vital for the appropriate management and follow-up of this entity. Darier´s disease is a rare autosomal dominant genetic skin disorder that manifests predominantly by scaly, pruritic, greasy and/or crusted papules throughout the body.[1-3] It is characterized as a hereditary acantholytic dermatosis with a typical onset during adolescence[2]. The ear is a reported site of affection, since the disease is known for affecting Seborrheic areas of the face[2,4,5]. Other typically affected areas are the forehead, nostrils, eyebrows, the scalp, neck, central chest, back and fold areas.[2] Patients may therefore be misdiagnosed with acne, eczema, and seborrheic dermatitis.[3,6]

CASE PRESENTATION:

We describe the case of a 70-year-old woman presenting to the Otolaryngology department with bilateral otalgia and pruritus with a 4-day evolution. She had been diagnosed with Darier´s disease by the age of 16 years by means of a skin biopsy revealing a typical acantholytic dyskeratosis histopathological pattern. She had been medicated with acitretin 25 mg in alternate days, despite reporting frequent flares. She described having recurrent similar episodes of otalgia and pruritus for many years requiring repeated topical treatment. External physical aspect of the pinna is shown in (Figure 1 and 2) (informed consent was obtained). The otoscopic examination showed an extremely desquamative external auditory canal with concurrent edema and scarce otorrhea bilaterally. The tympanic membrane was slightly erythematous yet intact without signs of middle ear involvement. After careful inspection, visible involuting skin lesions were also seen at the leg (Figure 3) and inframammary region. A diagnosis of bilateral otitis externa was assumed and a scheme of oral ciprofloxacin, ciprofloxacin + fluocinolone acetonide solution and topical ointment of hydrocortisone + Natamycin + Neomycin were prescribed, with resolution of the complaints after 1 week. The patient was also recommended to attend the Dermatologist to pursue systemic medical optimization. Three months after evaluation no other flares were registered.

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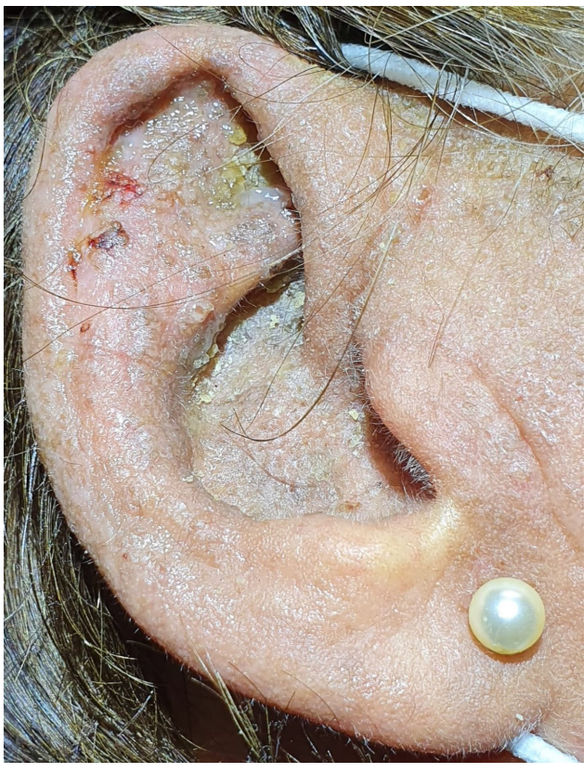

Fig. 1: Visual aspect of the right pinna. Note the greasy and scaly papules.

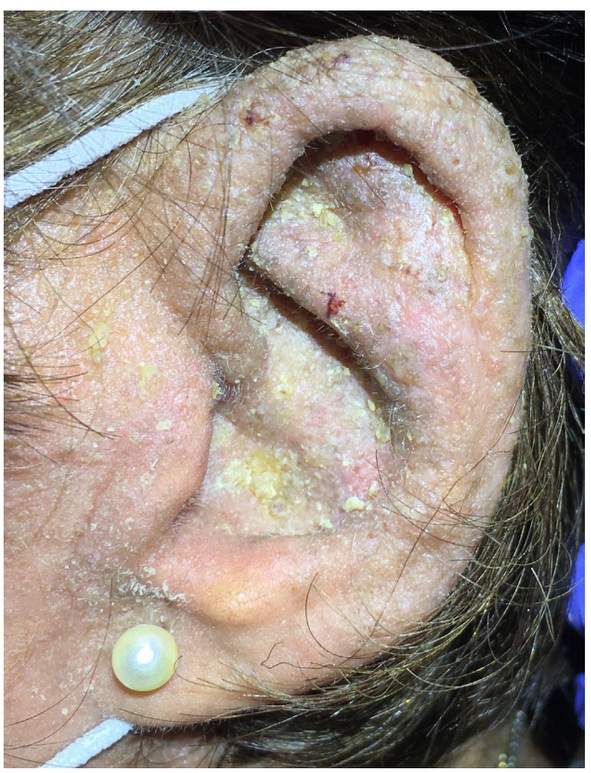

Fig. 2: Visual aspect of the left pinna. A darkish "elephant skin" is present at inspection of the helix.

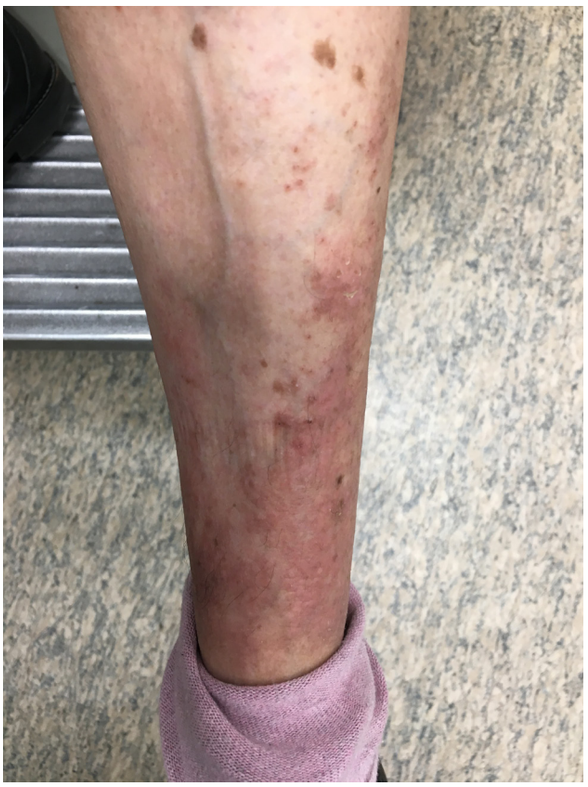

Fig. 3: Visual aspect of the leg. Note the violet blemishes.

DISCUSSION

The abnormal gene behind Darier´s disease has been identified as ATP2A2, found on chromosome 12q23-24.1. This gene codes for the SERCA enzyme or pump (SarcoEndoplasmic Reticulum Calcium-ATPase) that is required to transport calcium within the cell.[1,7–10] The exact mechanism by which this abnormal gene causes the disease is still under investigation but may be related to the disruption of skin cells junction mediated by desmosomes.[4,11] The severity of the disease varies over time, with affected patients experiencing flare-ups alternating with periods of lower disease activity.[4] Our patient clearly described this chronology. The diagnosis may require a skin biopsy, with a typical histopathological pattern.[5,12] Treatment is largely directed for reliving symptoms, but oral retinoids may be used for severe cases (such acitretin in this patient) at the expense of increasing side effects.[12] This case highlights the importance of an holistic approach in cases of chronic scaly lesions of the ear and external acoustic canal with an associated history of recurrent otitis externa.

CONCLUSION

Patients showing chronic otitis externa along with concurrent dermatologic lesions should be referred to Dermatology so that diagnostic investigation and targeted therapies may be offered.

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CONFLICT OF INTEREST

There are no conflicts of interest.

RESEARCH ETHICS AND PATIENT CONSENT

Authors declare that Informed consent was obtained for the clinical case description, so as for the images provided in the manuscript. The submission complies with local and International ethical standards.

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