



Letter to the Editor

Sensorineural Hearing Loss in Congenital Erosive and Vesicular Dermatitis

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Dear Editor,

We would like to present a case of congenital erosive and vesicular dermatitis associated with hearing impairment. To our knowledge, there are only few described cases of congenital erosive and vesicular dermatitis in the literature, and this is the only case presenting sensorineural hearing loss also.

Congenital erosive and vesicular dermatitis is a rare idiopathic and congenital (but apparently not hereditary) condition typically affecting preterm newborns and, more rarely, full-term newborns ^[1-4]. It was first reported by Cohen et al. ^[5] in 1985; the trunk and limbs are the most common areas affected by erosive dermatitis, which can involve more than 75% of the body. Less frequently, the tongue and scalp can be affected too, with the face and volar surfaces (palms and soles) relatively spared. Erosions, vesicles, and, later, crusts can be observed at birth. The prognosis is usually good as the lesions usually heal within the first months of life; however, typical stellate/reticulate scars usually persist. No other erosions usually occur after the neonatal period. To date, the pathogenesis of congenital erosive and vesicular dermatitis is uncertain; intrauterine traumas or infections have been considered as hypothetical causes ^[1-13].

We report the case of a 5-year-old boy. He was born at term by cesarean section (due to previous myomectomy) and presented symmetrical erosive, vesicular, and crusting lesions on the extremities and knees that healed within a few months but retained reticulated scars (Figure 1a). Informed consent was obtained from the patients' family. The diagnosis of congenital erosive and vesicular dermatitis was based on clinical features after excluding similar forms of perinatal dermatitis by cutaneous, blood, and laboratory tests. The newborn also presented a right hydrocele testis and a left testicular torsion that was treated by left orchiectomy in the early days of life. The child was hyposomic due to the slow speed of growth. Psychomotor and language development were otherwise normal.

The otoacoustic emissions (Accu-Screen; GN Otometrics, Taastrup, Denmark) were not detectable at birth. The auditory brainstem responses (ICS CHARTR; GN Otometrics, Taastrup, Denmark) tested at 1 month of life recorded a replicable wave V until 50 dB HL in both ears (Figure 1b). The results of tympanometry were bilaterally normal (Amplaid 724 Tympanometer; Amplimedical, Milan, Italy). The first conditioned orienting response audiometry was obtained at 8 months of age (Figure 1c), with a behavioral threshold at 50 dB from 250 to 4000 Hz. At the age of 4 years, the results of the first pure tone audiometry (Figure 1d) and vocal audiometry (Figure 1e) showed a moderate, bilateral, and symmetrical sensorineural hearing loss at mid-high frequencies (Amplaid 315 Audiometer; Amplimedical, Milan, Italy). Behind the ear aids have been prescribed at the age of 4 years.

A genetic assessment for the most common non-syndromic cases of genetic hearing loss was performed, with a negative result for GJB2 (connexin 26) and GJB6 (connexin 30) mutations.

The present case is interesting because (i) congenital erosive and vesicular dermatitis occurred in a full-term newborn and (ii) it was associated with sensorineural hearing loss. The most frequently associated findings described in the literature in congenital erosive and vesicular dermatitis are hyperthermia, ocular involvement, and onychia or hypoplastic nails. ^[1-13] Sensorineural hearing impairment could then be added as an associated clinical feature to this rare congenital condition, and an audiological evaluation should be considered in the diagnostic work-up of such patients.

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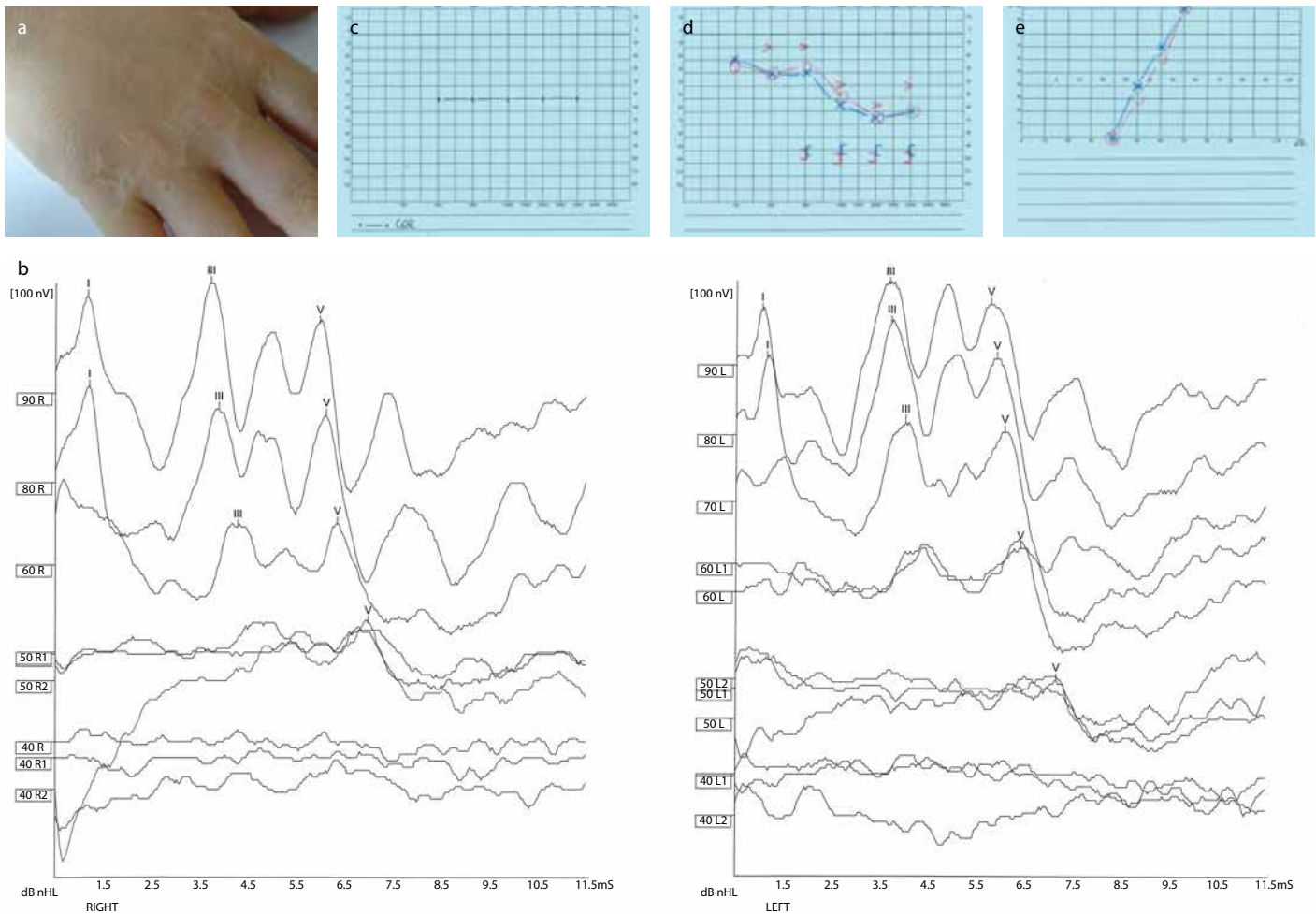


Figure 1. a-e. Typical reticulated scarring on the right hand (a), ABR showing a replicable wave V until 50 dB HL in both ears (b), COR audiometry depicted a behavioral threshold at 50 dB from 250 Hz to 4000 Hz at 8 months (c), Pure tone audiometry (d) and vocal audiometry showed a moderate, bilateral, and symmetrical sensorineural hearing loss at mid-high frequencies (e).

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