photophobia due to the heterozygous missense mutation p.Val37Glu in GJB6 and the homozygous mutations Val27Ile in GJB2 (Figure 2).

The present patient with CS carrying the GJB6 missense mutation p.Ala88Val and the GJB2 variant p.Val27Ile showed mild SNHL and photophobia. The identified GJB6 mutation p.Ala88Val was previously reported in a patient with CS without evidence of hearing disturbance or photophobia. Often, GJB2 mutations lead to SNHL in heterozygotes and homozygotes. Patients with GJB2 mutations are also known to show ocular involvement. Thus, in the present case, the additional symptoms of mild SNHL and photophobia might be attributable to p.Val27Ile in GJB2, although p.Val27Ile in GJB2 is a common SNP in the Japanese population.

In conclusion, the present case suggests that the coexistence of a GJB6 mutation and a heterozygous GJB2 variant in CS may lead to SNHL and photophobia in addition to the triad that typifies CS, even though the GJB2 variant is an SNP when it presents without any GJB6 mutation.

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Correction: This article was corrected on August 22, 2013, to correct an author’s name spelling.


CORRECTION

Incorrect Spelling of Author’s Name: In the article titled “Clouston Syndrome With Heterozygous GJB6 Mutation p.Ala88Val and GJB2 Variant p.Val27Ile Revealing Mild Sensorineural Hearing Loss and Photophobia,” posted online first in JAMA Dermatology on July 17, 2013 (doi:10.1001/jamadermatol.2013.4766), the first author’s name was spelled incorrectly throughout. The correct spelling is Kazumitsu Sugiuara, MD, PhD.