Lipoid proteinosis. *A report of 2 siblings and a brief review of the literature.*

To the Editor

Lipoid proteinosis (LP), also known as hyalinosis cutis et mucosae or Urbach-Wiethe disease (OMIM 247100) is a rare, autosomal recessive disorder typified by generalized thickening of skin, mucosae and certain viscera. I read with great interest the recent report of 2 Saudi sibling affected with this disorder.[1] However, I would like to mention that we have noted 10 Saudis reported with this disease in our earlier review on genetic skin disorders in the gulf area,[2] 6 of them have been described by a report of Uthman et al[3] and 4 by Nanda et al.[4] Using DNA from 3 affected siblings in a consanguineous Saudi Arabian family, a genome-wide linkage was performed,[5] and the disorder was mapped to 1q21. It is clear from all of the above reports that LP is not rate in the Arabian ancestry from the eastern province of Saudi Arabia, and the occurrence of close-relative intermarriages in Saudi Arabia may increase likelyhood of the occurrence of this disease in this geographical area. And the pre-marital counseling program, in this province, should take this into consideration, in order to provide optimal genetic advice to future couples.

Khalid M. Al-Aboud
Dermatology Department
King Faisal Hospital, Makkah
Kingdom of Saudi Arabia

Reply from the Author

I would like to thank Dr. Khalid M. Al Aboud for his interest in my paper and his valid comments. I agree fully with him as to the need for genetic counseling in our population due to the high degree of family intermarriage.

Sahar Al-Natour
Department of Dermatology
King Faisal University
College of Medicine, Dammam
Kingdom of Saudi Arabia

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